Fall 2021

KHF Hemosphere

2021 KHF Unite for Bleeding Disorders Walk

A sunny fall day greeted our Walk teams early on Saturday, October 23, 2021, at E. P. “Tom” Sawyer State Park on the eastern edge of Jefferson County. Thirteen teams from the greater Louisville area, Bowling Green, Cynthiana, and Somerset checked in for our 8th Unite for Bleeding Disorders Walk and accompanying festivities, which reflected a Halloween theme. Everyone’s spirits were high as Venus Marcum, our Walk Committee Chair, led the teams on their Walk route after the pinwheel ceremony, which set the stage for unity, hope, and resilience.

Afterwards, Walkers had the opportunity to visit with national and local sponsors, while the children enjoyed face painting, balloon animals, photo booth snapshots, pumpkin painting, games, and trick or treat. The costume contest featured three winners: Trey Poynter, a bloody-scary chef, Jack Meadows, a pirate; and Kayden Cockriel as Hermione Granger from “Harry Potter.” Leading up to the awards ceremony, adults enjoyed the drawing for door prizes.

The fundraising competition had been fierce among the top teams during recent weeks. Winners were determined by their leaderboard standing right before the Walk. The 3rd place Walk team winner was Team XL from Louisville with Dianne Hardman as Team Captain. They raised $1,430. The 2nd place Walk team winner was Team Tag and Trey’s Turtles from Bowling Green with Monica, Tag, and Trey Poynter as Team Captains. They raised $1,875. The 1st place Team winner was Team LEVI with Hunter Griffith Hill and Levi Hill from Cynthiana as Team Captains. They raised $2,134. We sincerely thank these teams and all the other teams for their fundraising accomplishments and their commitment to KHF’s mission and cause on behalf of the bleeding disorders community whom we serve. *Walk story continues on page 5*

Special News

MASAC Guidelines for Pregnancy and Perinatal Management of Women with Inherited Bleeding Disorders and Carriers of Hemophilia A or B

Revisions: 251; 175; 252

Women with bleeding disorders are at risk for bleeding in the peripartum period. Women with bleeding disorders and female carriers of hemophilia may also have affected newborns who are at risk for bleeding at birth and in the neonatal period. These guidelines provide recommendations for the diagnosis and management of women with bleeding disorders during pregnancy, labor and delivery, and in the postpartum period to minimize the risk of bleeding-related complications and facilitate early diagnosis of affected infants.

Preconception Diagnosis and Genetic Counseling

Adolescent females and women who may be carriers of hemophilia A or B, should be evaluated before they become pregnant. Factor assays should be completed during the non-pregnant state in a reference lab that specializes in coagulation testing. Normal factor activities do not exclude the diagnosis of genetic hemophilia carrier. Genetic testing should be done to establish hemophilia carrier status in potentially affected women. It is most effective to first test the male proband in the family and then test family members. (1)

It is also critically important to diagnose adolescent females and women with family history of von Willebrand disease and other inherited bleeding disorders prior as early as possible and to define risk of transmission to children prior to pregnancy.

Preconception genetic consultation should be offered to all women with bleeding disorders and possible carriers who plan to pursue a pregnancy. Women and their families should be acquainted with the various methods of diagnosing a potentially affected infant prior to delivery and the associated risks of each. Methods include preimplantation diagnosis, invasive prenatal diagnosis (chorionic villus sampling, amniocentesis, and cordocentesis), and ultrasound determination of fetal sex. (2-4) It regard to fetal sex, that female sex does not exclude potential for bleeding diathesis related to hemophilia carrier status especially in carriers with skewed x-inactivation or in females with compound heterozygosity for two factor gene variants.

Pregnancy Management

Pregnancy should be managed by a multidisciplinary team of specialists, including a coagulation disorders specialist, an obstetrician/gynecologist and an anesthesiologist, all of whom are knowledgeable in the management of women with bleeding disorders. Early pregnancy care should include anticipatory guidance about bleeding because spontaneous early pregnancy loss is associated with need for interventions in approximately 50%. (5) Women with bleeding disorders are not generally believed to be at increased risk of miscarriage unless they have either fibrinogen or factor XIII deficiency. (4,6, 7) Women who have fibrinogen deficiency with a bleeding phenotype are at a high risk of miscarriage if their fibrinogen levels are below 100 mg/dL. They should receive replacement therapy during pregnancy to maintain their levels above 100 mg/dL. (6) Women with severe factor XIII deficiency are also at high risk of miscarriage and should also receive replacement therapy monthly during pregnancy. (4)

Any early pregnancy procedures should be performed in settings where hemorrhage could be managed, including uterine evacuation for early pregnancy loss and procedural abortion. Bleeding prophylaxis for uterine evacuation can include uterotonics and anti-fibrinolytics (e.g. tranexamic acid). Prophylaxis with VWF concentrate, desmopressin, and/or anti-fibrinolytics is indicated prior to uterine evacuation and other invasive procedures during pregnancy, such as chorionic villous sampling, amniocentesis, and cerclage.

MASAC Guidelines for Pregnancy and Perinatal Management of Women with Inherited Bleeding Disorders and Carriers of Hemophilia A or B

Labor and Delivery Management

Women with bleeding disorders are at risk of bleeding complications during pregnancy and in the postpartum period. A plan for the management of childbirth should be in place well before delivery. Women should give birth in a facility that has available consultation with a bleeding disorders specialist, neonatology, and the appropriate laboratory, pharmacy and transfusion services support. Clotting factor replacement and other hemostatic agents and blood products must be available on-site.

Hemophilia A carriers may have congenitally low factor levels that increase their risk of bleeding. Even though their factor VIII levels rise during pregnancy, they may not rise to the levels achieved by women without a bleeding disorder. Similarly, women with von Willebrand disease have von Willebrand factor levels and factor VIII levels that rise during pregnancy, but possibly not to the levels achieved by women without a bleeding disorder. Factor IX levels do not rise during pregnancy. Factor VIII, IX, or VWF levels should be assessed once or twice in the third trimester of pregnancy in order to plan for possible administration of clotting factor concentrates or other clotting factor replacement therapy at the time of delivery.

Hemophilia carriers and women with VWD should receive factor replacement prophylaxis at the time of delivery if their factor levels are less than 50% (50 IU/dL). (2-4) When available, recombinant or virally inactivated clotting factor concentrate should be used as opposed to fresh frozen plasma or cryoprecipitate. (2-4) Women with factor levels above 50% (50 IU/dL) should be given the option of neuraxial anesthesia. (8) Targeting a VWF activity level of 0.50 to 1.50 IU/mL over targeting an activity level of 1.50 IU/mL is advisable. (9) VWF activity levels should be maintained at 0.50 IU/mL while the epidural is in place and for at least 6 hours after removal. The assessment of whether neuraxial anesthesia is appropriate for an individual patient is a complex decision that includes assessment of factors outside the scope of these guidelines. The ultimate decision about whether it is appropriate for an individual patient to undergo these procedures lies with the obstetric anesthesiologist or other clinician performing the procedure. Decisions regarding anesthesia and delivery should be made in the context of a multidisciplinary discussion with input from anesthesia, hematology, and obstetrics and shared decision making with the patient. These discussions should take place well in advance of the patient's due date. Patients should also be assessed for thrombotic risk post-delivery, and prophylaxis (e.g., compression stockings or low-molecular-weight heparin) should be provided when needed.

(See MASAC Document #250 for specific treatment product recommendations for each of these bleeding disorders.)

DDAVP (1-desamino-8-D-arginine vasopressin) is a synthetic vasopressin that can be used to raise both von Willebrand factor levels and factor VIII levels in patients who respond and can be used during pregnancy for prophylaxis prior to invasive procedures such as amniocentesis. (2-4,10) Sánchez-Luceros et al noted no adverse events in 34 women with VWD who received a single dose of DDAVP immediately prior to epidural catheter placement. (11) However, the use of DDAVP at the time of childbirth can cause fluid retention. The United Kingdom guidelines for the management of women with bleeding disorders recommend that fluids be restricted to 1000 mL for the 24-hour period after DDAVP administration.(5) Since women routinely receive two to three times that amount of fluid at the time of delivery, routinely receive oxytocin which also causes fluid retention, and invariably undergo redistribution of fluid from the extravascular to the intravascular space after delivery, DDAVP must be used with caution, and electrolytes and urine output must be monitored closely for 24 hours.(3)

Continued on pg. 6

Event News

2021 Summer Family Event – Annual Education Meeting

The Summer Family Event was such a joyful time to be able to re-connect with our bleeding disorders community in person again after transitioning to a number of virtual programs last year.

First thing, attendees visited with exhibitors and subsequently enjoyed Jim Romano's riveting key note advocacy address during breakfast, entitled "If I Can Do It, You Can Do It." Jim is a nationally known advocate for the bleeding disorders community whose wealth of experience, knowledge, and enthusiasm was exhilarating and inspiring to all. Breakout sessions addressed "Raising Resilience" and "Reframing Your Mindset" and were presented respectively by Julie Thomas from Genentech and John Martinez from Sanofi Genzyme. They found an active listening audience at a time when everyone is trying to cope with the negative fallout of living through a pandemic and the pain, loss, and grief it has brought to many. A wholesome lunch and a drawing for several door prizes followed the educational programming. The children's program was facilitated by Connie Thacker of Christian Fellowship. She was assisted by our board member, Travis Price, who knows how to entertain kids with his fabulous, hand crafted games.

Outgoing board members after six years of service were our treasurer, Brad Comer, who presented the financial report; and Barbara Bitter, who had been a member of the Vegasville and Walk Committees. We greatly appreciate their contributions of time, talent, and resources. New board members are Jennifer Dunegan of Louisville who already serves on the Camp and Walk Committees and Mason Stout of Somerset who is a counselor at our "Camp Discovery" summer camp program and a member of the Camp Committee. New officers are Laura Webb, president; Eric Marcum, vice president, Kristin Taylor, treasurer and Andrew Hartmans, secretary. The event concluded by awarding gift certificates to all interested families to visit Muth's Candies on Market Street, Louisville's oldest and most famous candy store.

We thank all volunteers, speakers, and facilitators who made this event possible. We also want to express our appreciation to the industry exhibitors who supported and participated in this event: Accredo, Bayer HealthCare, BioMarin, CSL Behring, CVS Caremark, Genentech, HEMA Biologics, Heritage Biologics, Novo Nordisk, Optum Infusion Pharmacy, Paragon Hemophilia Solutions, Sanofi Genzyme, and Takeda.

Event News

Fall Scholarship Awards

We were thrilled to award three post-secondary scholarships for the 2021 fall semester. The Herb Schlaughenhoupt, Jr. Memorial Scholarship was awarded to Lori Lyons Hall of Richmond, Kentucky. Lori graduated summa cum laude from Eastern Kentucky University and is now pursuing a doctoral degree in Veterinary Science at Auburn University in Alabama. The Theodore (Ted) B. Forcht Memorial Scholarship was awarded to John Graham. John is the son of John and Leah Graham of Winchester. This fall, John is a sophomore at Centre College in Danville. His major is finance and economics with an emphasis on politics. The Betty Meadors Mattingly Memorial Scholarship award was bestowed on Lillian Omerso of New Albany, Indiana. Lillian is the daughter of Brian and Jennifer Omerso. A couple of years ago, this talented young lady participated in "Hemophilia, the Musical." She is now a freshman at Olivet Nazarene University. Lillian is majoring in zoology. We extend our heartfelt congratulations and best wishes for their educational endeavors to these most deserving scholarship recipients! The next deadline for submitting a scholarship application is January 15, 2022 for the 2022 spring semester. For an application form and guidelines, contact the KHF office at 502-456-3233 or 1-800-582-CURE (2873) or send an email to **info@kyhemo.org**.

2021 KHF Unite Walk cont. from pg. 1

Each team member of the top three teams received a handsome medal and a warm toboggan. The top three team captains received a lovely inscribed glass award to commemorate and show our appreciation for their leadership and hard work. The top three highest individual fundraisers were also recognized. In third place was Constance Wheat from Louisville with $880 whose team achieved fourth place and barely missed the 3rd place win; in second place was Dianne Hardman from Louisville with $1,380; and in first place were Hunter Griffith Hill and Levi Hill with $1,959. They also received a very nice glass award for their achievements. Walk t-shirts, tote bags, and incentive gifts for individuals who raised $250 or more added a special touch to everyone's participation in this important fundraiser and community unity event. We thank from the bottom of our hearts all walkers, team captains, donors, volunteers, and sponsors who made the 2021 Kentucky Unite for Bleeding Disorders Walk a success. Local Gold Sponsor was Novo Nordisk; Silver Sponsors were CSL Behring and HEMA Biologics; Bronze Sponsors were BioMarin, Octapharma, Pfizer, Sanofi Genzyme, Specialty Care Rx, Takeda; Kilometer Sponsors were Biomatrix and Republic Bank & Trust Company. The Walk raised $35,256 for KHF's programs and services for Kentucky's bleeding disorders community.

More News

MASAC Guidelines cont. from pg. 3

Method of Delivery

As of December 2008, the Centers for Disease Control (CDC) Uniform Data Collection (UDC) database contained data on 580 male infants with hemophilia A or B. In 569 out of the 580 infants, the mode of delivery and the outcome were known. 385 infants with hemophilia (68%) were delivered vaginally and 184 by caesarean delivery. There were 16 intracranial hemorrhages among the 385 infants delivered vaginally (twelve were delivered by vacuum extraction and four by forceps), giving a rate of intracranial hemorrhage of 4 percent among infants delivered vaginally. In contrast, there was one intracranial hemorrhage among 184 infants delivered by caesarean section, giving a rate of 0.5%. Among the 236 mothers who were known to be carriers, delivery plans were modified somewhat: 156 infants (66%) were delivered vaginally with 1 operative vaginal delivery and 2 intracranial hemorrhages (1.3%), while among the 80 infants who were delivered by caesarean section there were no intracranial hemorrhages. (12)

In 2019 the PedNET Haemophilia Research Foundation published results from the PedNET multicenter study including 926 neonates (786 with severe and 140 with moderate hemophilia). Six hundred thirty-three were delivered by vaginal delivery and 293 by Cesarean section. Fifteen (2.4%) of vaginal deliveries were complicated by intracranial hemorrhage compared to 5 (1.7%) of Cesarean sections (P=not significant). As demonstrated in other studies vaginal delivery with instrumentation was associated with high risk of intracranial and other major bleeds. (13).

While most infants of hemophilia carriers can be safely delivered vaginally, the outcome of labor cannot be predicted, and a spontaneous (non-operative) vaginal delivery cannot be guaranteed. A vaginal delivery may be associated with abnormal labor. Therefore, obstetricians caring for women who are carriers of hemophilia should discuss with the woman the maternal and fetal risks of a vaginal delivery versus a planned caesarean delivery; the option of a planned caesarean delivery should be considered when an affected or potentially affected infant is anticipated. (12) In women who elect vaginal delivery, forceps and vacuum extraction, interventions that triple the risk of intracranial hemorrhage in affected infants, should be avoided, as should fetal scalp electrode monitoring during labor.

Continued in the next Hemosphere edition

Source: National Hemophilia Foundation (NHF) Web Site - www.hemophilia.org

KHF Cares as Covid Continues

Kentucky Hemophilia Foundation continues to provide financial assistance to bleeding disorder families whose household income has decreased because of loss of job, lay off, furlough, or reduced hours during the ongoing COVID-19 health crisis or due to another type of emergency and who are unable to pay a specific household bill. Requesting families must reside in Kentucky, and the person seeking assistance must either have a bleeding disorder or be the parent of a minor child with a bleeding disorder. Assistance is contingent on the availability of funds.

Call **502-456-3233 or 800-582-CURE (2873)** or send an email to info@kyhemo.org to make a request.

More News

Kentucky Hemophilia Foundation Membership

We thank these current KHF members

Individual/Family Memberships, $20
Michael & Cathy Johnson
Nita Wayne-Zehnder

Supporting Memberships, $35
Judy Hayes
in memory of Jason Hayes
John L. Silletto

Sustaining Memberships, $100
Arthur Hackman

John & Leah Graham
Barbara W. Grayson
D. Spalding Grayson
Dr. David & Leslie Houvenagle

Benefactor Memberships, $250
Glen & Deborah Hitt
Laura & Glenn Webb

Champion/Corporate Member, $500
LTC (R) John & Pat Tharp

2021 Summer/Fall Donations

We thank the following individuals and companies for their generous support!

Donor, $7,000
VACO LLC

Donor, $500
LTC (R) John & Patricia Tharp

Donors, $100+
Dianne Hardman for KHF's Unite Walk
Greg Fiscus
Kroger Community Rewards

Donors, $50 - $99
Louise Hardaway for KHF's Summer Camp
Jennifer Hitt

Donors, Up to $49
Amazon Smiles
Fr. William L. Fichteman

In Memory

July 1, 2021 – November 1, 2021

Gone from our sight but never our memories; gone from our touch but never our hearts… May their memory be a blessing!

Leander "Lee" Goff Strand, Jr. & Evelyn Barr Kramer

Do The Five

Follow these steps to prevent or reduce complications of bleeding disorders

1. **Get an annual comprehensive checkup at a hemophilia treatment center.**
2. **Get vaccinated – Hepatitis A and B are preventable.**
3. **Treat bleeds early and adequately.**
4. **Exercise to protect your joints.**
5. **Get tested regularly for blood-borne infections.**

To find out more about the National Prevention Program developed by the National Hemophilia Foundation in collaboration with the Centers for Disease Control and Prevention (CDC), click on www.hemophilia.org or call toll-free 800-42-HANDI.

Like us on Facebook and keep up-to-date on all **KHF** activities and events.

KOSAIR CHARITIES

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KENTUCKY HEMOPHILIA FOUNDATION
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A **ONCE-WEEKLY** TREATMENT OPTION FOR HEMOPHILIA B.

To find out about a prescription option, talk to your doctor or visit

OnceWeeklyForHemophiliaB.com

Pfizer

When it comes to your hemophilia A treatment

Move beyond the threshold[a]

Esperoct® can give you high factor levels for longer.[b]

Extend half-life beyond the standard 22-hour average half-life in adults[c]

FOR ADULTS AND ADOLESCENTS

Switching made easy with a standard 50 IU/kg dose every 4 days

-50% fewer infusions if you previously infused every other day

-40% fewer infusions if you previously infused 3x a week

High factor levels

At or above 3% for 100% of the time[d,e]

At or above 5% for 90% of the time[d,f]

Flexible on the go

The ONLY extended half-life product that can be stored up to 104°F[g]

Please see Brief Summary for complete storage instructions.

Safety Proven across 5 studies, the largest and longest EHL clinical trial program

[a]Of 1% trough factor levels for standard half-life (SHL) products in adults and adolescents.
[b]Compared with SHL products.
[c]Data shown are from 42 adults who received a pharmacokinetic (PK) assessment around the first Esperoct® 50 IU/kg dose.
[d]Trough level goal is 1% for prophylaxis.
[e]Data shown are from a study where 175 previously treated adolescents and adults received routine prophylaxis with Esperoct® 50 IU/kg every 4 days. Pre-dose factor activity (trough) levels were evaluated at follow-up visits. Mean trough levels for adolescents (12-<18 years) were 2.7 IU/dL.
[f]Steady-state FVIII activity levels were estimated in 143 adults and adolescents using pharmacokinetic modeling.
[g]For up to 3 months.

What is Esperoct®?

Esperoct® [antihemophilic factor (recombinant), glycopegylated-exei] is an injectable medicine to treat and prevent or reduce the number of bleeding episodes in people with hemophilia A. Your healthcare provider may give you Esperoct® when you have surgery

- Esperoct® is not used to treat von Willebrand Disease

IMPORTANT SAFETY INFORMATION

Who should not use Esperoct®?

- You should not use Esperoct® if you are allergic to factor VIII or any of the other ingredients of Esperoct® or if you are allergic to hamster proteins

What is the most important information I need to know about Esperoct®?

- **Do not attempt to do an infusion yourself unless you have been taught how by your healthcare provider or hemophilia treatment center**
- **Call your healthcare provider right away or get emergency treatment right away if you get any signs of an allergic reaction, such as:** hives, chest tightness, wheezing, dizziness, difficulty breathing, and/or swelling of the face

What should I tell my healthcare provider before using Esperoct®?

- Before taking Esperoct®, you should tell your healthcare provider if you have or have had any medical conditions, take any medicines (including non-prescription medicines and dietary supplements), are nursing, pregnant or planning to become pregnant, or have been told that you have inhibitors to factor VIII
- Your body can make antibodies called "inhibitors" against Esperoct®, which may stop Esperoct® from working properly. **Call your healthcare provider right away if your bleeding does not stop after taking Esperoct®**

What are the possible side effects of Esperoct®?

- Common side effects of Esperoct® include rash or itching, and swelling, pain, rash or redness at the location of infusion

Please see Brief Summary of Prescribing Information on the following page.

Discover more at Esperoct.com.

novo nordisk®

Novo Nordisk Inc., 800 Scudders Mill Road, Plainsboro, New Jersey 08536 U.S.A.

esperoct®
antihemophilic factor (recombinant), glycopegylated-exei

esperoct®

antihemophilic factor (recombinant), glycopegylated-exei

Brief Summary information about ESPEROCT® [antihemophilic Factor (recombinant), glycopegylated-exei]

This information is not comprehensive.

- Talk to your healthcare provider or pharmacist
- Visit www.novo-pi.com/esperoct.pdf to obtain FDA-approved product labeling
- Call 1-800-727-6500

Patient Information

ESPEROCT®
[antihemophilic factor (recombinant), glycopegylated-exei]

Read the Patient Information and the Instructions For Use that come with ESPEROCT® before you start taking this medicine and each time you get a refill. There may be new information.

This Patient Information does not take the place of talking with your healthcare provider about your medical condition or treatment. If you have questions about ESPEROCT® after reading this information, ask your healthcare provider.

What is the most important information I need to know about ESPEROCT®?

Do not attempt to do an infusion yourself unless you have been taught how by your healthcare provider or hemophilia treatment center.

You must carefully follow your healthcare provider's instructions regarding the dose and schedule for infusing ESPEROCT® so that your treatment will work best for you.

What is ESPEROCT®?

ESPEROCT® is an injectable medicine used to replace clotting Factor VIII that is missing in patients with hemophilia A. Hemophilia A is an inherited bleeding disorder in all age groups that prevents blood from clotting normally.

ESPEROCT® is used to treat and prevent or reduce the number of bleeding episodes in people with hemophilia A.

Your healthcare provider may give you ESPEROCT® when you have surgery.

Who should not use ESPEROCT®?

You should not use ESPEROCT® if you

- are allergic to Factor VIII or any of the other ingredients of ESPEROCT®
- if you are allergic to hamster proteins

If you are not sure, talk to your healthcare provider before using this medicine.

Tell your healthcare provider if you are pregnant or nursing because ESPEROCT® might not be right for you.

What should I tell my healthcare provider before I use ESPEROCT®?

You should tell your healthcare provider if you:

- Have or have had any medical conditions.
- Take any medicines, including non-prescription medicines and dietary supplements.
- Are nursing.
- Are pregnant or planning to become pregnant.
- Have been told that you have inhibitors to Factor VIII.

How should I use ESPEROCT®?

Treatment with ESPEROCT® should be started by a healthcare provider who is experienced in the care of patients with hemophilia A.

ESPEROCT® is given as an infusion into the vein.

You may infuse ESPEROCT® at a hemophilia treatment center, at your healthcare provider's office or in your home. You should be trained on how to do infusions by your hemophilia treatment center or healthcare provider. Many people with hemophilia A learn to infuse the medicine by themselves or with the help of a family member.

Your healthcare provider will tell you how much ESPEROCT® to use based on your weight, the severity of your hemophilia A, and where you are bleeding. Your dose will be calculated in international units, IU.

Call your healthcare provider right away if your bleeding does not stop after taking ESPEROCT®.

If your bleeding is not adequately controlled, it could be due to the development of Factor VIII inhibitors. This should be checked by your healthcare provider. You might need a higher dose of ESPEROCT® or even a different product to control bleeding. Do not increase the total dose of ESPEROCT® to control your bleeding without consulting your healthcare provider.

Use in children

ESPEROCT® can be used in children. Your healthcare provider will decide the dose of ESPEROCT® you will receive.

If you forget to use ESPEROCT®

If you forget a dose, infuse the missed dose when you discover the mistake. Do not infuse a double dose to make up for a forgotten dose. Proceed with the next infusions as scheduled and continue as advised by your healthcare provider.

If you stop using ESPEROCT®

Do not stop using ESPEROCT® without consulting your healthcare provider.

If you have any further questions on the use of this product, ask your healthcare provider.

What if I take too much ESPEROCT®?

Always take ESPEROCT® exactly as your healthcare provider has told you. You should check with your healthcare provider if you are not sure. If you infuse more ESPEROCT® than recommended, tell your healthcare provider as soon as possible.

What are the possible side effects of ESPEROCT®?

Common Side Effects Include:

- rash or itching
- swelling, pain, rash or redness at the location of infusion

Other Possible Side Effects:

You could have an allergic reaction to coagulation Factor VIII products. **Call your healthcare provider right away or get emergency treatment right away if you get any signs of an allergic reaction, such as:** hives, chest tightness, wheezing, dizziness, difficulty breathing, and/or swelling of the face.

Your body can also make antibodies called "inhibitors" against ESPEROCT®, which may stop ESPEROCT® from working properly. Your healthcare provider may need to test your blood for inhibitors from time to time.

These are not all of the possible side effects from ESPEROCT®. Ask your healthcare provider for more information. You are encouraged to report side effects to FDA at 1-800-FDA-1088.

Tell your healthcare provider about any side effect that bothers you or that does not go away.

What are the ESPEROCT® dosage strengths?

ESPEROCT® comes in five different dosage strengths. The actual number of international units (IU) of Factor VIII in the vial will be imprinted on the label and on the box. The five different strengths are as follows:

Cap Color Indicator	Nominal Strength
Red	500 IU per vial
Green	1000 IU per vial
Gray	1500 IU per vial
Yellow	2000 IU per vial
Black	3000 IU per vial

Always check the actual dosage strength printed on the label to make sure you are using the strength prescribed by your healthcare provider.

How should I store ESPEROCT®?

Prior to Reconstitution (mixing the dry powder in the vial with the diluent):

Protect from light. Do not freeze ESPEROCT®.

ESPEROCT® can be stored in refrigeration at 36°F to 46°F (2°C to 8°C) for up to 30 months until the expiration date stated on the label. During the 30 month shelf life, ESPEROCT® may be kept at room temperature (not to exceed 86°F/30°C) for up to 12 months, **or** up to 104°F (40°C) for no longer than 3 months.

If you choose to store ESPEROCT® at room temperature:

- Record the date when the product was removed from the refrigerator.
- Do not return the product to the refrigerator.
- Do not use after 12 months if stored up to 86°F (30°C) **or** after 3 months if stored up to 104°F (40°C) **or** the expiration date listed on the vial, whichever is earlier.

Do not use this medicine after the expiration date which is on the outer carton and the vial. The expiration date refers to the last day of that month.

After Reconstitution:

The reconstituted (the final product once the powder is mixed with the diluent) ESPEROCT® should appear clear and colorless without visible particles.

The reconstituted ESPEROCT® should be used immediately.

If you cannot use the reconstituted ESPEROCT® immediately, it must be used within 4 hours when stored at or below 86ºF (30°C) or within 24 hours when stored in a refrigerator at 36°F to 46°F (2°C to 8°C). Store the reconstituted product in the vial.

Keep this medicine out of the sight and out of reach of children.

What else should I know about ESPEROCT® and hemophilia A?

Medicines are sometimes prescribed for purposes other than those listed here. Do not use ESPEROCT® for a condition for which it is not prescribed. Do not share ESPEROCT® with other people, even if they have the same symptoms that you have.

Revised: 10/2019

ESPEROCT® is a trademark of Novo Nordisk Health Care AG.

For Patent Information, refer to: http://novonordisk-us.com/patients/products/product-patents.html

More detailed information is available upon request. Available by prescription only.

Manufactured by:
Novo Nordisk A/S
Novo Allé
DK-2880 Bagsværd, Denmark

For information about ESPEROCT® contact:
Novo Nordisk Inc.
800 Scudders Mill Road
Plainsboro, NJ 08536, USA
1-800-727-6500

US19ESP00168 December 2019

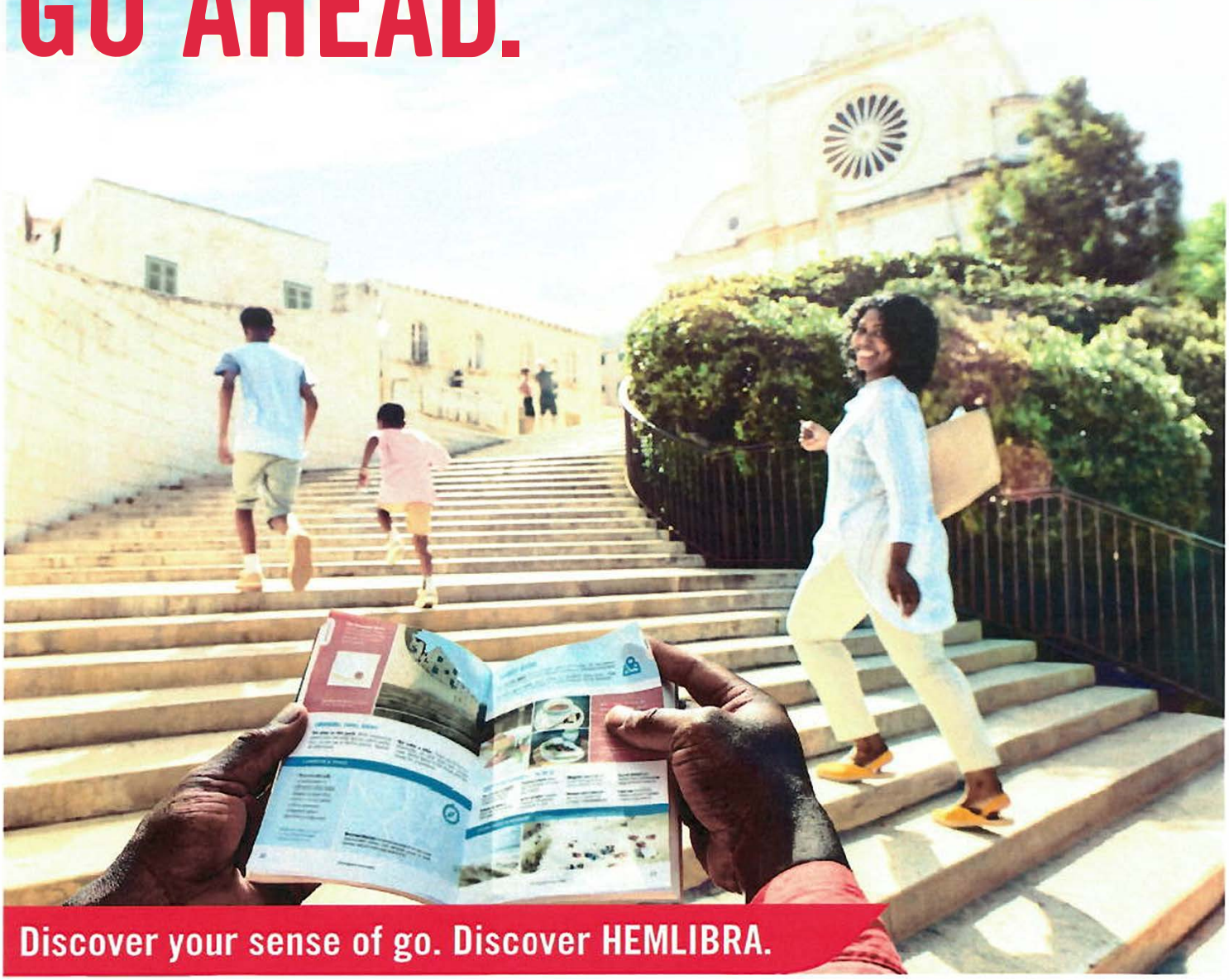
novo nordisk®

GO SEEK. GO EXPLORE.

GO AHEAD.

PEOPLE LIKE YOU. STORIES LIKE YOURS.

Explore more at HEMLIBRAjourney.com



What is HEMLIBRA?

HEMLIBRA is a prescription medicine used for routine prophylaxis to prevent or reduce the frequency of bleeding episodes in adults and children, ages newborn and older, with hemophilia A with or without factor VIII inhibitors.

What is the most important information I should know about HEMLIBRA?

HEMLIBRA increases the potential for your blood to clot. People who use activated prothrombin complex concentrate (aPCC; Feiba®) to treat breakthrough bleeds while taking HEMLIBRA may be at risk of serious side effects related to blood clots.

These serious side effects include:

- **Thrombotic microangiopathy (TMA),** a condition involving blood clots and injury to small blood vessels that may cause harm to your kidneys, brain, and other organs
- **Blood clots (thrombotic events),** which may form in blood vessels in your arm, leg, lung, or head

Please see Brief Summary of Medication Guide on following page for Important Safety Information, including **Serious Side Effects**.

HEMLIBRA®
emicizumab-kxwh
injection for subcutaneous use
150 mg/mL

Medication Guide
HEMLIBRA® (hem-lee-bruh)
(emicizumab-kxwh)
injection, for subcutaneous use

What is the most important information I should know about HEMLIBRA?

HEMLIBRA increases the potential for your blood to clot. Carefully follow your healthcare provider's instructions regarding when to use an on-demand bypassing agent or factor VIII (FVIII) and the recommended dose and schedule to use for breakthrough bleed treatment.

HEMLIBRA may cause the following serious side effects when used with activated prothrombin complex concentrate (aPCC; FEIBA®), including:

- **Thrombotic microangiopathy (TMA).** This is a condition involving blood clots and injury to small blood vessels that may cause harm to your kidneys, brain, and other organs. Get medical help right away if you have any of the following signs or symptoms during or after treatment with HEMLIBRA:
  - confusion
  - weakness
  - swelling of arms and legs
  - yellowing of skin and eyes
  - stomach (abdomen) or back pain
  - nausea or vomiting
  - feeling sick
  - decreased urination
- **Blood clots (thrombotic events).** Blood clots may form in blood vessels in your arm, leg, lung, or head. Get medical help right away if you have any of these signs or symptoms of blood clots during or after treatment with HEMLIBRA:
  - swelling in arms or legs
  - pain or redness in your arms or legs
  - shortness of breath
  - chest pain or tightness
  - fast heart rate
  - cough up blood
  - feel faint
  - headache
  - numbness in your face
  - eye pain or swelling
  - trouble seeing

If aPCC (FEIBA®) is needed, talk to your healthcare provider in case you feel you need more than 100 U/kg of aPCC (FEIBA®) total.

See "**What are the possible side effects of HEMLIBRA?**" for more information about side effects.

What is HEMLIBRA?

HEMLIBRA is a prescription medicine used for routine prophylaxis to prevent or reduce the frequency of bleeding episodes in adults and children, ages newborn and older, with hemophilia A with or without factor VIII inhibitors.

Hemophilia A is a bleeding condition people can be born with where a missing or faulty blood clotting factor (factor VIII) prevents blood from clotting normally.

HEMLIBRA is a therapeutic antibody that bridges clotting factors to help your blood clot.

Before using HEMLIBRA, tell your healthcare provider about all of your medical conditions, including if you:

- are pregnant or plan to become pregnant. It is not known if HEMLIBRA may harm your unborn baby. Females who are able to become pregnant should use birth control (contraception) during treatment with HEMLIBRA.
- are breastfeeding or plan to breastfeed. It is not known if HEMLIBRA passes into your breast milk.

Tell your healthcare provider about all the medicines you take, including prescription medicines, over-the-counter medicines, vitamins, or herbal supplements. Keep a list of them to show your healthcare provider and pharmacist when you get a new medicine.

How should I use HEMLIBRA?

See the detailed "Instructions for Use" that comes with your HEMLIBRA for information on how to prepare and inject a dose of HEMLIBRA, and how to properly throw away (dispose of) used needles and syringes.

- Use HEMLIBRA exactly as prescribed by your healthcare provider.
- **Stop (discontinue) prophylactic use of bypassing agents the day before starting HEMLIBRA prophylaxis.**
- **You may continue prophylactic use of FVIII for the first week of HEMLIBRA prophylaxis.**
- HEMLIBRA is given as an injection under your skin (subcutaneous injection) by you or a caregiver.
- Your healthcare provider should show you or your caregiver how to prepare, measure, and inject your dose of HEMLIBRA before you inject yourself for the first time.
- Do not attempt to inject yourself or another person unless you have been taught how to do so by a healthcare provider.
- Your healthcare provider will prescribe your dose based on your weight. If your weight changes, tell your healthcare provider.
- You will receive HEMLIBRA 1 time a week for the first four weeks. Then you will receive a maintenance dose as prescribed by your healthcare provider.
- If you miss a dose of HEMLIBRA on your scheduled day, you should give the dose as soon as you remember. You must give the missed dose as soon as possible before the next scheduled dose, and then continue with your normal dosing schedule. **Do not** give two doses on the same day to make up for a missed dose.
- HEMLIBRA may interfere with laboratory tests that measure how well your blood is clotting and may cause a false reading. Talk to your healthcare provider about how this may affect your care.

What are the possible side effects of HEMLIBRA?

- See "**What is the most important information I should know about HEMLIBRA?**"

The most common side effects of HEMLIBRA include:

- redness, tenderness, warmth, or itching at the site of injection
- headache
- joint pain

These are not all of the possible side effects of HEMLIBRA.

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

How should I store HEMLIBRA?

- Store HEMLIBRA in the refrigerator at 36°F to 46°F (2°C to 8°C). Do not freeze.
- Store HEMLIBRA in the original carton to protect the vials from light.
- Do not shake HEMLIBRA.
- If needed, unopened vials of HEMLIBRA can be stored out of the refrigerator and then returned to the refrigerator. HEMLIBRA should not be stored out of the refrigerator for more than a total of 7 days or at a temperature greater than 86°F (30°C).
- After HEMLIBRA is transferred from the vial to the syringe, HEMLIBRA should be used right away.
- Throw away (dispose of) any unused HEMLIBRA left in the vial.

Keep HEMLIBRA and all medicines out of the reach of children.

General information about the safe and effective use of HEMLIBRA.

Medicines are sometimes prescribed for purposes other than those listed in a Medication Guide. Do not use HEMLIBRA for a condition for which it was not prescribed. Do not give HEMLIBRA to other people, even if they have the same symptoms that you have. It may harm them. You can ask your pharmacist or healthcare provider for information about HEMLIBRA that is written for health professionals.

What are the ingredients in HEMLIBRA?

Active ingredient: emicizumab-kxwh

Inactive ingredients: L-arginine, L-histidine, poloxamer 188, and L-aspartic acid.

Manufactured by: Genentech, Inc., A Member of the Roche Group,
1 DNA Way, South San Francisco, CA 94080-4990
U.S. License No. 1048

For more information, go to www.HEMLIBRA.com or call 1-866-HEMLIBRA.
This Medication Guide has been approved by the U.S. Food and Drug Administration
Revised: 10/2018

HEMLIBRA®
emicizumab-kxwh
injection for subcutaneous use
150 mg/mL

Genentech
A Member of the Roche Group

Natalie Kukla

Patient advocate

About Natalie

Natalie is a Novo Nordisk Hemophilia Community Liaison who has three family members living with bleeding disorders. She wants to make a difference and is excited to support people in the hemophilia community.

Connect with Natalie

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Hemophilia Community Liaison

MIDWEST (IL, KY, TN)

novo nordisk®

Thank You

To all our wonderful sponsors, donors, and members. This year, you and your support have allowed us to help over 1,000 families cope with the effects of a bleeding disorder. You can never truely know the effects your help has brought to the lives of so many.

For more information:
info@kyhemo.org or
502-456-3233
800-582-CURE (2873)

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