

Factor Nine News

The Coalition for Hemophilia B

Summer 2016

Topics in Hemophilia



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WFH 2016 World, July 2016

NHF's 68th Annual Meeting



July was an amazing month as National Hemophilia Foundation's 68th Annual Meeting was held July 21st-July 23rd at the beautiful Gaylord Resort & Convention Center in Orlando, Florida.

We attended the Impact Award Ceremony hosted by Patrick Lynch and sponsored by Shire. One of the award recipients was John Jewell, a member of our Coalition family. We are proud of all honorees on their outstanding service to the community! Congratulations! 🎉



WFH 2016 World Congress

Following NHF, World Federation of Hemophilia 2016 World Congress was held at the Orange County Convention Center July 24th-July 28th.



It was wonderful to meet people with hemophilia from all over the world and hear their stories. It was an amazing opportunity having the two conferences held at the same time for National to meet International! A rare opportunity embraced by many! In total there were about 5,000 attendees! 🎉



In Sympathy, Matthew M. Stinger



We were very sad to hear of the passing of *Matthew Martin Stinger*, a much loved member of the bleeding disorder community. Matthew passed away suddenly on Monday, August 15, 2016 at the age of 33. The son of Susanne and the late Arthur Stinger, Matthew was born in Abington, Pennsylvania. Matt held bachelor degrees in both Psychology and Nursing from Seton Hall University. From 2006-2011, he worked as a registered nurse in the pediatric emergency room at Children's Hospital of Philadelphia. After leaving the hospital, Matt worked as an RN/Hemophilia Resource Specialist for a specialty pharmacy serving the needs of patients within the bleeding disorders community. Matt was very involved with The Double H Ranch Camp both as a camp attendee and a counselor. Donations in Matthew's name may be sent to *The Double H Ranch Camp*, 97 Hidden Valley Road; Lake Luzerne, New York 12846.

Rest in Peace, Matthew. You will be greatly missed.

CHARLIE

By: Melissa Franzen

My 11-year-old Charlie has had quite the summer this year. Charlie has severe hemophilia B. This summer was Charlie's third year of going to hemophilia camp at The Painted Turtle in Southern California. The past two years, he has attended camp and has done well infusing himself with the help of the wonderful doctors and nurses that volunteer there. However, after he got home from camp, he was never interested in infusing himself. Being the helicopter hemophilia mom that I am, I would of course always step in to do it for him.



This year, something was different. He came home really excited at how well he had done on his infusion and he told me that he wanted to do it himself. He asked me to stand across the counter from him so that I couldn't step in and do it for him if he didn't get it right away. I watched him carefully as he mixed

the factor and prepped all of his supplies. He put on the tourniquet and went in on the back of his hand. He went in a little high so I encouraged him to go down a little bit and motioned to him across the counter. Surprisingly, I didn't swoop in and take it out of his hand. It was all worth it, as I saw his face light up when he got the return and he knew he was in. Since then, he has infused himself every time. He informed me that he has set a goal to try to infuse himself every time before he goes back to camp next year! This has been so huge for us!



He's about to enter into sixth grade, and our school, sixth grade is the year you get to go to a weeklong science camp. Even though it is only about 20 minutes from our house, they are gone for five days camping up in the mountains. It is a big rite of passage for these kids. They hike and learn about nature, geology and astronomy. When I met with his sixth grade teacher to discuss hemophilia, I mentioned this had been a big summer for him. I asked if we came into her classroom and showed her how he infuses from beginning to end, and showed her how confident he was in the process, would she allow him to self-infuse at science camp? To my surprise, she said yes! I am confident he will do great. Science camp is coming up in November and we can't wait!

Even though a little bit of me is sad that he doesn't need me for everything, I couldn't be more proud of Charlie. I can't wait to see what the future holds for this strong, self-empowered boy. Thank you to The Painted Turtle camp for giving Charlie the confidence he needed to take the leap into taking care of himself and his hemophilia. 🐢



He's free to infuse only once every 14 days. **Are you?**

The only FDA-approved treatment for hemophilia B with up to 14-day dosing.* **Visit us at IDELVION.com.**



Dosing schedule that fits into your lifestyle



High and sustained Factor IX levels



A median annualized spontaneous bleeding rate of zero in 7- and 14-day prophylaxis

*In appropriate people 12 years and older. Talk with your doctor.

Important Safety Information

IDELVION is used to control and prevent bleeding episodes in people with hemophilia B. Your doctor might also give you IDELVION before surgical procedures. Used regularly as prophylaxis, IDELVION can reduce number of bleeding episodes.

IDELVION is administered by intravenous injection into the bloodstream, and can be self-administered or administered by a caregiver. Do not inject IDELVION without training and approval from your healthcare provider or hemophilia treatment center.

Tell your healthcare provider of any medical condition you might have, including allergies and pregnancy, as well as all medications you are taking. Do not use IDELVION if you know you are allergic to any of its ingredients, including hamster proteins. Tell your doctor if you previously had an allergic reaction to any FIX product.

Stop treatment and immediately 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 Factor IX, which could stop IDELVION from working properly. You might need to be tested for inhibitors from time to time. IDELVION might also increase the risk of abnormal blood clots in your body, especially if you have risk factors. Call your healthcare provider if you have chest pain, difficulty breathing, or leg tenderness or swelling.

In clinical trials for IDELVION, headache was the only side effect occurring in more than 1% of patients (1.8%), but is not the only side effect possible. Tell your healthcare provider about any side effect that bothers you or does not go away, or if bleeding is not controlled with IDELVION.

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**.

Please see brief summary of prescribing information for IDELVION on next page.

IDELVION®, Coagulation Factor IX (Recombinant), Albumin Fusion Protein
Initial U.S. Approval: 2016

BRIEF SUMMARY OF PRESCRIBING INFORMATION

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

What is IDELVION?

IDELVION is an injectable medicine used to replace clotting Factor IX that is absent or insufficient in people with hemophilia B. Hemophilia B, also called congenital Factor IX deficiency or Christmas disease, is an inherited bleeding disorder that prevents blood from clotting normally.

IDELVION is used to control and prevent bleeding episodes. Your healthcare provider may give you IDELVION when you have surgery. IDELVION can reduce the number of bleeding episodes when used regularly (prophylaxis).

Who should not use IDELVION?

You should not use IDELVION if you have had life-threatening hypersensitivity reactions to IDELVION or are allergic to:

- hamster proteins
- any ingredients in IDELVION

Tell your healthcare provider if you have had an allergic reaction to any Factor IX product prior to using IDELVION.

What should I tell my healthcare provider before using IDELVION?

Discuss the following with your healthcare provider:

- Your general health, including any medical condition you have or have had, including pregnancy, and any medical problems you may be having
- Any medicines you are taking, both prescription and non-prescription, and including any vitamins, supplements, or herbal remedies
- Allergies you might have, including allergies to hamster proteins

- Known inhibitors to Factor IX that you've experienced or been told you have (because IDELVION might not work for you)

What must I know about administering IDELVION?

- IDELVION is administered intravenously, directly into the bloodstream.
- IDELVION can be self-administered or administered by a caregiver with training and approval from your healthcare provider or hemophilia treatment center. **(For directions on reconstituting and administering IDELVION, see the Instructions for Use in the FDA-Approved Patient Labeling section of the full prescribing information.)**
- Your healthcare provider will tell you how much IDELVION to use based on your weight, the severity of your hemophilia B, your age, and other factors. Call your healthcare provider right away if your bleeding does not stop after taking IDELVION.
- Blood tests may be needed after you start IDELVION to ensure that your blood level of Factor IX is high enough to properly clot your blood.

What are the possible side effects of IDELVION?

Allergic reactions can occur with IDELVION. Call your healthcare provider right away and stop treatment 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 can make antibodies, called inhibitors, against Factor IX, which could stop IDELVION from working properly. Your healthcare provider may need to test your blood for inhibitors from time to time.

IDELVION might increase the risk of abnormal blood clots forming in your body, especially if you have risk factors for such clots. Call your healthcare provider if you experience chest pain, difficulty breathing, or leg tenderness or swelling while being treated with IDELVION.

A common side effect of IDELVION is headache. This is not the only side effect possible. Tell your healthcare provider about any side effect that bothers you or does not go away.

Please see full prescribing information, including FDA-approved patient labeling.

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



As I See It

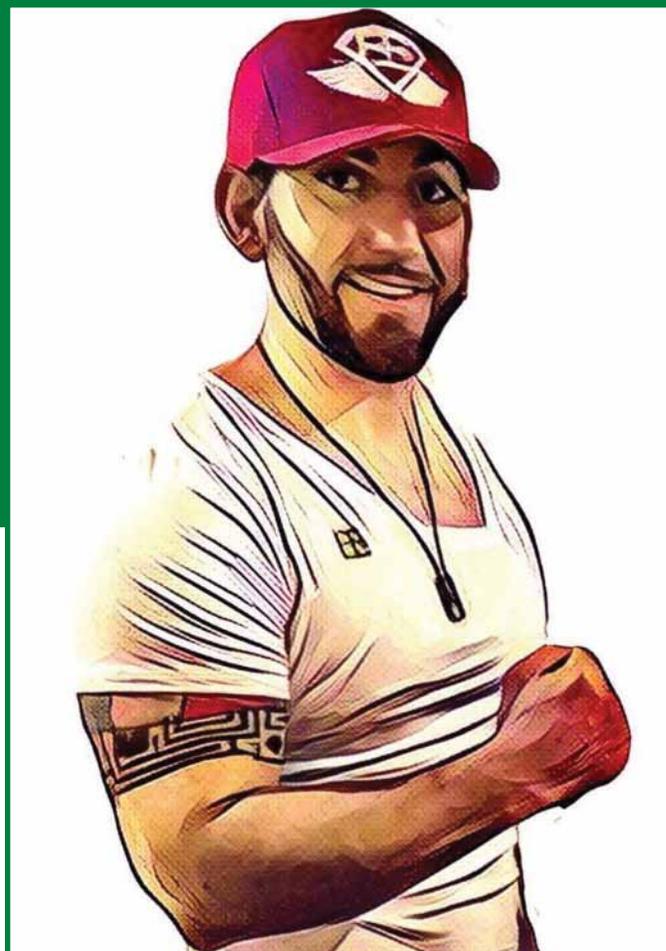
My Story— Rewritten

By L.A. Aguayo

Severe hemophilia is a badge of honor that I display loud and proud. At every opportunity, I spread awareness about bleeding disorders and try to paint a realistic picture that will give others a glimpse into our lives. We face so many obstacles, but until we spread awareness and educate others, we will always be living in our own shadows and continue to be misunderstood. Having hemophilia is like a roller-coaster ride, with lots of ups and downs. Managing these almost made me throw in the towel, but I got some news that would change my life forever.

When I grew up in the late 1980s, information on hemophilia was scarce, and the only time my family learned anything new was when I went in for a yearly HTC checkup. To my parents, hemophilia was scary: it meant that I couldn't get hurt, or else I would bleed internally. If I did get a bleed, then I needed medication and I would eventually get better. When I was 15, my parents put the responsibility into my hands to either tell them when I had a bleed or just infuse myself. But while I was trying to accept this responsibility, my parents separated. My dad started a new family and my mom was busy working, trying to support her two boys. This left little time to focus on my hemophilia and my ever-changing needs.

Over the years I went from job to job and relationship to relationship, because nobody understood me. My employers wanted to know why I was always calling in sick, and my girlfriends never understood my anger and pain. I was an emotionally unstable person to be around. The only consistent thing about me was my desire to be a professional athlete. Though I dreamed of being a football player, basketball player, or professional wrestler, golf seemed most feasible. Year after year, I invested all my money to compete in US Amateur, Gateway PGA, and Metropolitan Golf Association events, where I played against the best. I always told myself that I would never give up on my dreams.



Eventually, I was competing at a high level in professional golf events like the US Open Qualifier. Although outwardly, my golf career seemed to be looking up, internally I knew that I was getting worse. Having some pretty severe symptoms, like memory loss, lack of energy, sleepiness, and feeling very edgy, prompted me to make an emergency visit to my HTC.

This visit would change my life forever. Only 26, I was told that my liver was in the final stage of cirrhosis and that I needed treatment [for HCV] immediately. Treatment would consist of getting weekly injections of interferon for three months, along with taking two other pills (Sovaldi® and Ribavirin). I was told that I would be very sick throughout the process. I did research online, and after reading what people had to say, of course I was under the impression that my life was practically over. I read about liver transplants, and how most people don't live much longer after a transplant. I spent most of the next few weeks randomly crying and feeling scared. I told myself I would just file for disability because I couldn't keep a job with hemophilia, and now with this, I would be as good as dead.

I started the treatment, and just as the doctors said, I was very sick. I mostly slept all day and hid in my dark room, where I would occasionally play video games, and then



I'd come out for a quick snack. I tossed and turned in my bed, always feeling like my head would explode or that I needed to vomit. I didn't shave or get a haircut, and I was as pale as the walls in my room. Simply said, I felt hopeless.

Halfway through the treatment, something started to change for me mentally. Although I wasn't dying, I really did experience the feeling of someone telling me that my life was at risk and that my time was limited. Something very powerful happens when you feel like you haven't much time. You start to think of your legacy. How would my two young sons look at my life if I died today? How do I want to be remembered? Do I have a purpose?

I woke up one day feeling better than average, and I had time to soul search. It was like a light bulb went on in my head. I finally realized my purpose was to use my life experiences and struggles to help as many people as possible in the bleeding disorder community. It was time for the world to listen to me, and I was going to create awareness about what we go through. I knew this wouldn't be easy, but I was dedicated and full of purpose.

I knew if I wanted to be a leader in my community, then I had to start living my life by example. From that day forward, I dedicated myself to being compliant to my infusion regimen, becoming extremely fit, and volunteering in the community. I started surrounding myself with people who would help me grow, and

segregated myself from people who tried to bring me down or didn't believe in me. After dealing for so long with failed relationships, panic attacks, random crying, and even contemplated suicides, it was my opportunity to rewrite my book—and this time, I want the world to read it.

I'm still on my journey to becoming the best me possible, but I'm definitely on the right path. I'm growing every day, and I will never make excuses for why I can't do something. I've accepted my past as a blessing that allows me to reach out and relate to others. To me, hemophilia isn't a handicap, it's my reason why. My purpose. 🐾

Luis Andres ("L.A.") Aguayo is 28 and has severe hemophilia B. He lives in St. Louis, Missouri, with his fiancé Amanda and four children. L.A. works full time as a patient service representative in the bleeding disorder community and enjoys volunteering for the Gateway Hemophilia Association. L.A. is currently in a competition to make the cover of Men's Health Magazine and represent the bleeding disorder community as the 2016 Men's Health "Ultimate Guy." His motto is "See you at the top!"

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Living with Hemophilia and Depression

By Maria Iannone

Climbing the Seven Summits has been the dream of many adventurers. To date only 231 people in history have completed the Seven Summit list. As of March 2015, I have summited five ... with only Mt. Vinson (Antarctica) and Mt. Everest remaining. Completing the Seven Summits is already an overwhelming endeavor; now imagine accomplishing this with a bleed[ing] disorder. I am excited to take on this task and educate others on hemophilia and overcome my own personal obstacles in the process! ~ Chris Bombardier

Adventures of a Hemophiliac Blog

Chris Bombardier is a 29-year-old with severe factor IX deficiency. He tells me he was not always so self-assured and inspired as his Seven Summits blog suggests. He has a phobia of needles and infusing, which he was only able to overcome because of his desire to play baseball in high school. He recalls feeling frustration and anger and, when he was younger, never really knowing why. After college, when his baseball career ended, Chris felt lost. He stopped infusing, started drinking, and was not active in the community. Chris was severely depressed. He did not know how to reach out to his hemophilia treatment center (HTC). When the HTC social worker asked, "How's it going?" in clinic, Chris would give the socially acceptable answer, "Fine."

Chris says he has always felt different, missing extra time from baseball because of his hemophilia. This spring he had a bleed that stalled his training for climbing Mt. Rainier. Chris knows how lucky he is to have access to factor, good medical care, and loving family and friends. But he continues to cope with feeling angry about having to overcome the pain and setbacks associated with living with hemophilia. He's not alone.

Depression Is Normal

Depression is a normal part of the range of human experience. We are drawn together as a species by our need to be engaged and connected with each other, and in the activities that make up our daily living. We would not have cohesive communities, families, and work environments if we did not feel deeply about each other and what we are doing. Because of this, when we feel a loss, we appropriately experience difficult emotional reactions like depression.

According to the Diagnostic and Statistical Manual of Mental Disorders (DSMV), sad feelings become a problem when they last longer than two months after a major loss, continue for at least two weeks, and cause problems in important areas of your life. For some, everything may



look good from the outside, but it takes a monumental effort to keep up appearances and stay functional. Fortunately, depression almost always gets better with proper focused attention.

Unfortunately, depression is an illness that still carries with it a negative stigma for many people. Our cultural norms and societal values often leave little room for depression. Success means "living happily ever after." Depression is associated with immature coping skills, exemplified in common phrases like "Big boys don't cry" and "Put your big-girl pants on and get over it."

Depression strikes people of all ages, races, creeds, educational levels, and socioeconomic levels. The symptoms that depressed people experience are affected by the traits they inherit, and what they have learned about how to cope with stress. Stressful, long-

term, difficult life challenges such as financial problems, physical limitations, pain, health issues, access to adequate healthcare, and the high cost of medications can all contribute to the emotional burden of stress. Depression is one of our most common problems. It is also a complex issue.

Depression and Chronic Illness

The Hemophilia Experiences, Results and Opportunities (HERO) study, conducted by the HERO Advisory Board,¹ found that 57% of parents of children with hemophilia feel the need for psychosocial support in coping with their child's hemophilia. In 2007 the Arizona Hemophilia and Thrombosis Center (AzHTC), where I work, found that a little more than 1 in 3 men with hemophilia experienced depression. We discovered that people with hemophilia who also reported a lack of social support or problems with unemployment were more likely to be depressed. Sadly, we also found that almost a quarter of all the people surveyed said they were without social support and almost half were unemployed.

The HERO study found that poor mental health puts a burden on relationships and employment. Imagine the downward spiral generated by being depressed and feeling isolated. Of the 57% of parents who reported feeling the need for psychosocial support in the HERO study, 33% did not actually seek the help they needed. This is what makes depression so debilitating. As early as 2004, the Cleveland Clinic Health System reported that as many as 1 in 4 men with chronic illness are suffering from symptoms of depression. In the general population the same year, 1 in 25 men in the US suffered from symptoms of depression. That means that living with a chronic illness increases the rate of depression by 6 to 8 times that of the general population.

Unfortunately, experiencing depression affects the way a person feels and thinks about his or her health. Depression can increase perception of pain. Chronic depression can also cause gastrointestinal problems, change immune function, raise cholesterol, and contribute to heart disease. The ENIGMA study, authored at the Brain and Mind Research Institute in Australia, shows that depression promotes changes in the brain that can affect memory. Depression can contribute to changes in ability to focus and concentrate. It's not surprising that people suffering from depression often feel in desperate need of immediate relief.

Depression is associated with an increase in risky behaviors like drug and alcohol abuse, poor adherence to medical treatments, and decreases in quality of life. The HERO study found that mental health can be an underlying force in choosing treatment regimes. At the AzHTC, we found that 2 in 3 of the men surveyed reported difficulties in their normal activities due to their depressive symptoms. Choices in hemophilia care treatment plans,



as well as the ability to follow through with those plans, can be influenced by depression.

It is universally accepted within the bleeding disorder community that patients with hemophilia enjoy better health and quality of life when they engage in prophylactic treatment with factor. Straying from prescribed treatments is damaging and, at worst, life threatening. Unfortunately, factor replacement therapy often requires a demanding regimen of frequent intravenous administrations, usually 1 to 3 times per week. Taking factor as prescribed can be difficult because of the physical burden of venipuncture or central venous access, inconvenience, complications, fear of the risks of infections or inhibitors, and cost. Sometimes the emotional and practical toll of hemophilia care on patients or their parents can promote misunderstanding about when to treat or call a provider. For some, the experience can be so overwhelming that they stop paying attention and deny the negative consequences of failing to treat as prescribed. Adolescents are especially prone to feeling a lack of commitment to treat. Many are afraid of exposing their hemophilia diagnosis to family or friends, or in a public situation. It's not uncommon for people to limit their activities because they will take their medication only at home. Symptoms of depression and anxiety can further reduce one's ability to take proper care.

Fortunately, it is becoming widely accepted that mental health and the impact of living with a chronic illness are inseparably linked. Mental health screening and care are increasingly available in HTC's. If we pay attention to depression, we can reduce the severity of the symptoms and improve quality of life.

Signs and Symptoms of Depression

Eight basic categories of symptoms are part of a clinical assessment for depression: sleep, interest, guilt, energy, concentration, appetite, lethargy or restlessness, and thoughts of suicide. All eight categories are broad in scope, and are evaluated based on whether a person experiences significant changes from normal behavior patterns.

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BeneFix
Coagulation Factor IX (Recombinant)
Room Temperature Storage



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- Have fun and earn points
- Make charitable donations

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What is BeneFix?

BeneFix, Coagulation Factor IX (Recombinant), is an injectable medicine that is used to help control and prevent bleeding in people with hemophilia B. Hemophilia B is also called congenital factor IX deficiency or Christmas disease.

BeneFix is **NOT** used to treat hemophilia A.

Important Safety Information

- BeneFix is contraindicated in patients who have manifested life-threatening, immediate hypersensitivity reactions, including anaphylaxis, to the product or its components, including hamster protein.
- Call your health care provider right away if your bleeding is not controlled after using BeneFix.
- Allergic reactions may occur with BeneFix. Call your health care provider or get emergency treatment right away if you have any of the following symptoms: wheezing, difficulty breathing, chest tightness, your lips and gums turning blue, fast heartbeat, facial swelling, faintness, rash or hives.

- Your body can make antibodies, called "inhibitors," which may stop BeneFix from working properly.
- If you have risk factors for developing blood clots, such as a venous catheter through which BeneFix is given by continuous infusion, BeneFix may increase the risk of abnormal blood clots. The safety and efficacy of BeneFix administration by continuous infusion have not been established.
- Some common side effects of BeneFix are nausea, injection site reaction, injection site pain, headache, dizziness and rash.

Please see the Brief Summary for BeneFix on the next 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.

*BeneFix was approved February 11, 1997.

R_x only

Brief Summary

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.Pfizer.com or call our medical communications department toll-free at 1-800-934-5556.

Please read this Patient Information carefully before using BeneFix and each time you get a refill. There may be new information. This brief summary does not take the place of talking with your doctor about your medical problems or your treatment.

What is BeneFix?

BeneFix is an injectable medicine that is used to help control and prevent bleeding in people with hemophilia B. Hemophilia B is also called congenital factor IX deficiency or Christmas disease.

BeneFix is **NOT** used to treat hemophilia A.

What should I tell my doctor before using BeneFix?

Tell your doctor and pharmacist about all of the medicines you take, including all prescription and non-prescription medicines, such as over-the-counter medicines, supplements, or herbal remedies.

Tell your doctor about all of your medical conditions, including if you:

- are pregnant or planning to become pregnant. It is not known if BeneFix may harm your unborn baby.
- are breastfeeding. It is not known if BeneFix passes into the milk and if it can harm your baby.

How should I infuse BeneFix?

The initial administrations of BeneFix should be administered under proper medical supervision, where proper medical care for severe allergic reactions could be provided.

See the step-by-step instructions for infusing in the complete patient labeling.

You should always follow the specific instructions given by your doctor. If you are unsure of the procedures, please call your doctor or pharmacist before using.

Call your doctor right away if bleeding is not controlled after using BeneFix.

Your doctor will prescribe the dose that you should take.

Your doctor may need to test your blood from time to time.

BeneFix should not be administered by continuous infusion.

What if I take too much BeneFix?

Call your doctor if you take too much BeneFix.

What are the possible side effects of BeneFix?

Allergic reactions may occur with BeneFix. Call your doctor or get emergency treatment right away if you have any of the following symptoms:

wheezing	fast heartbeat
difficulty breathing	swelling of the face
chest tightness	faintness
turning blue (look at lips and gums)	rash
	hives

Your body can also make antibodies, called "inhibitors," against BeneFix, which may stop BeneFix from working properly.

Some common side effects of BeneFix are nausea, injection site reaction, injection site pain, headache, dizziness and rash.

BeneFix may increase the risk of thromboembolism (abnormal blood clots) in your body if you have risk factors for developing blood clots, including an indwelling venous catheter through which BeneFix is given by continuous infusion. There have been reports of severe blood clotting events, including life-threatening blood clots in critically ill neonates, while receiving continuous-infusion BeneFix through a central venous catheter. The safety and efficacy of BeneFix administration by continuous infusion have not been established.

These are not all the possible side effects of BeneFix.

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

How should I store BeneFix?

DO NOT FREEZE BeneFix. BeneFix kit can be stored at room temperature (below 86°F) or under refrigeration. Throw away any unused BeneFix and diluent after the expiration date indicated on the label.

Freezing should be avoided to prevent damage to the pre-filled diluent syringe.

BeneFix does not contain a preservative. After reconstituting BeneFix, you can store it at room temperature for up to 3 hours. If you have not used it in 3 hours, throw it away.

Do not use BeneFix if the reconstituted solution is not clear and colorless.

What else should I know about BeneFix?

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

If you would like more information, talk to your doctor. You can ask your doctor for information about BeneFix that was written for healthcare professionals.

This brief summary is based on BeneFix® [Coagulation Factor IX (Recombinant)] Prescribing Information LAB-0464-9.0, revised August 2015.

AlphaNine® SD

Coagulation Factor IX (Human)



www.alphaninesd.com

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1. Problems with sleep, such as having a hard time staying awake in the evening, waking up in the morning, or extra daytime napping; or, conversely, having trouble falling asleep, staying asleep at night, or waking too early in the morning.
2. A recent loss of interest in doing things that would normally be pleasurable, such as hobbies, friends, sex, exercise, or work.
3. A tendency to focus on unrealistic negative thoughts, feel guilty or preoccupied with past failings, or misinterpret trivial day-to-day events.
4. A disabling experience of tiredness or fatigue without physical exertion, such as being exhausted for no clear reason.
5. A lack of ability to concentrate or make decisions; a tendency to become easily distracted or have problems with memory.
6. Significant changes in appetite, resulting in weight changes that require forced eating or constantly craving food.
7. Problems with slowed speech, thinking, or body movements; or, conversely, rapid speech, rapid changes in the content of a conversation, an inability to sit still, pacing, hand-wringing, pulling or rubbing of skin or clothing.
8. Thoughts of death or suicide, thinking about giving up, “ending the pain,” or a belief that “others would be better off without me.”



According to the Suicide Intervention Handbook,² most suicidal people are unsure about dying and are desperately looking for a way out. Talking about suicide does not create or increase the risk; it reduces the risk. Often, nonfatal harmful behaviors such as unsuccessful suicide attempts, taking pills, cutting, and

other high-risk activities are an invitation for others to help. Most people communicate how they are feeling by direct statement, physical signs (cutting themselves, losing weight, isolating), or emotional reactions (irritability, moodiness, sadness). Everyone thinking about suicide should be taken seriously. Feelings or events over a prolonged period of time are what typically contribute to a suicide attempt.

Depression and Families

The burden of raising a child with hemophilia can take a toll. According to the HERO study, parents may be coping with their own feelings of guilt, shock, disappointment, fear, or anxiety. Raising a child with hemophilia can place stress on parents' sense of self-esteem and personal relationships. This can lead to anger, resentment, guilt, and sometimes overcompensation in parenting style.

Children with hemophilia have the burden of coping with normal developmental milestones as well as frequent medical procedures and complications. Medical issues range from serious, requiring hospitalization, to routine, requiring frequent venipuncture. Medical treatment may be traumatizing, impact children's social lives, or limit their ability to fully participate in family, school, and community activities. Adolescence is complicated by the stress associated with developmental milestones in conjunction with changing hormones and great pressures to conform. Teens who are different at this stage can be bullied, feel ostracized, or have a sense of not being fully in control. Teens are trying to individuate from their parents. Some adolescents may have a “woe is me” attitude because of their bleeding disorder, feeling hopeless or overly cautious. Others may overcompensate for the restrictions imposed by living with a bleeding disorder, and may take unnecessary risks. The stress associated with growing up with hemophilia and raising a child with hemophilia increases the likelihood of symptoms of depression.

Recognizing depression in children presents unique challenges. *Zero to Three*, a national organization dedicated to the mental well-being of children, emphasizes that children often do not have the verbal or cognitive skills to describe their experience. This leaves the burden of awareness on their primary caretakers. Changes in established patterns of behavior—especially persistent depressed mood or irritability—are clues to inspire questioning about whether depression is at play. As any parent knows, this is easier said than done. In infants and toddlers, we look for marked changes in sleeping or eating habits, and feelings of anger, irritability, or restlessness. In older children, we look for changes in eating or sleeping habits; depressed or irritable mood; restlessness, lethargy, or fatigue; feeling sad, hopeless, worthless, or guilty; having little interest in doing things that are normally fun; trouble concentrating or problem solving; trouble responding to teachers or caregivers; increased irresponsible or defiant behavior; or thoughts of death or suicide. Usually, attentive parents know when something is up, and it doesn't hurt to err on the side of caution and check with your doctor, HTC, or mental healthcare provider.

Learn About Treatment Options

Assessing depression is a complex diagnostic process



CHALLENGES
SOLUTIONS
GRATEFUL
ALTERNATIVES

Jay lives with severe hemophilia B with inhibitors.

Change the way you picture living with a rare bleeding disorder

Novo Nordisk is helping people like Jay write his story.

That's why we are continuously seeking new ways to help support and educate the bleeding disorders community. Because at Novo Nordisk, we're always committed to helping you make your potential possible.

that requires professional attention to medical issues, a full review of symptoms, collecting information from all possible sources, and getting a detailed medical and social history. Visit your HTC team if you suspect depression in yourself or a loved one.

When assessing medical issues, it is vital to consider risk factors such as medications that can cause depressive symptoms and medical conditions known to cause depression. In the current healthcare climate, it can be difficult for a provider to devote time to a full review of symptoms. In 2002, Dr. Ronald Remick, at St. Paul's Hospital in Vancouver, BC,³ identified four common errors in recognizing depression. The first results from underdiagnosing depression because of incomplete assessments. The second results from a failure to collect information from family, friends, and caretakers, relying solely on the patient, even though the patient's view may be skewed because of depressive symptoms. Third, a provider may minimize a patient's depression because of the existence of other major health problems. This sounds something like, "Of course you're depressed; anyone with your disability would be depressed." Fourth, depression can be overdiagnosed when a provider bases the diagnosis on a "depressed mood" without a complete screening. It's important to learn how to be a smart consumer when it comes to depression care, increasing your chance for more effective care.

The Patient Protection and Affordable Care Act (ACA), enacted in 2013, has helped people with chronic illness by barring insurance coverage exclusions due to preexisting conditions. The ACA also expanded care to those in poverty. In 2014 many plans were required to cover 10 essential health benefits (EHB). Mental healthcare is among the EHB. Specifically, many plans require screening and assessing for depression, alcohol misuse, tobacco use, and domestic violence. Further, healthcare systems are being tasked with treating mental health similarly to physical health. Mental health assessment may be covered in your plan. Please don't let the fear of not having coverage keep you from asking your HTC for help.

Talk About It

If you suspect that you or a loved one may be depressed, first of all, talk about it. Talk to your doctor, family, friends, therapist, or clergy. Engage in support groups at your

HTC, spiritual community, or neighborhood center. Get screened by a mental healthcare professional. Educate yourself: read books about depression, or search the Internet for professional articles from reliable sources. The National Institute of Mental Health (NIMH)⁴ has many resources. Learn to advocate for yourself and your loved ones.

Overcome the stigma of depression. You have many choices when coping with depression, including making changes to your lifestyle, engaging in psychotherapy, or trying medication; even doing nothing is a choice.

Successful treatment usually includes a combination of psychotherapy, medication, and lifestyle changes. However, treating depression is a highly individual process.

Think About How You Are Thinking

Unfortunately, when you're depressed, you don't feel like doing anything or talking to anyone. We react to life events based on automatic, sometimes subconscious interpretations. At

times these automatic thoughts can promote depression, heartache, even physical pain in the present. Automatic thoughts can lead to a passive interpersonal style, where we go along with anything, whether we like it or not, to avoid conflict at all costs. Maybe as children, we learned to be very passive because we got a treat for being "such a good kid," but as adults, we're struggling with our jobs because we are not assertive enough. Sometimes automatic thoughts lead to an extremely aggressive interpersonal style where we push people to do or think as we do in order to control outcomes. Habits like this can reduce our success at home, work, and—especially important for those with a bleeding disorder—at the HTC. Learned habits can contribute to poor listening styles such as "spacing out," "pretend" listening, "selective" listening, inattention, or being self-centered. All of these strategies, if left unchecked, can affect our ability to maintain healthy relationships or be productive within our family, community, and workplace.

Fake It 'Til You Make It

One of the most effective strategies to combat depression is to engage in a pleasurable activity. But when you're depressed, this strategy is often one of the first things to go. When you're trying to reclaim your life from depression, you may have to "fake it 'til you make it." The bottom line is to begin engaging in activities that, over



"AFTER WORKING
WITH MY DOCTOR
AND BASED ON MY RESPONSE,
I NOW INFUSE PROPHYLACTICALLY
EVERY 14 DAYS!"

Casey, on ALPROLIX

Casey, a MyALPROLIX Peer,[™] started on a once-every-10-day prophylaxis infusion schedule and adjusted to once every 14 days.

Learn more at www.alprolix.com/findyourfit

Extended protection* from bleeds

ALPROLIX is the first factor IX offering prophylaxis infusion schedules starting every 7 or 10 days with the potential to extend based on your response.

*ALPROLIX has been proven to help patients prevent bleeding episodes using a prophylaxis regimen.

The recommended starting prophylaxis regimens are either 50 IU/kg once weekly, or 100 IU/kg once every 10 days. Dosing regimen can be adjusted based on individual response.

Children under 12 years of age may have higher Factor IX body weight-adjusted clearance, shorter half-life, and lower recovery. Higher dose per kilogram body weight or more frequent dosing may be needed in these children.

Indications and Important Safety Information

Indications

ALPROLIX [Coagulation Factor IX (Recombinant), Fc Fusion Protein] is a recombinant DNA derived, coagulation factor IX concentrate indicated in adults and children with hemophilia B for:

- On-demand treatment and control of bleeding episodes
- Perioperative management of bleeding
- Routine prophylaxis to reduce the frequency of bleeding episodes

ALPROLIX is not indicated for induction of immune tolerance in patients with hemophilia B.

Important Safety Information

Do not use ALPROLIX if you are allergic to ALPROLIX or any of the other ingredients in ALPROLIX.

Tell your healthcare provider if you have or have had any medical problems, take any medicines, including prescription and non-prescription medicines, supplements, or herbal medicines, have any allergies and all your medical conditions, including if you are pregnant or planning to become pregnant, are breastfeeding, or have been told you have inhibitors (antibodies) to factor IX.

Allergic reactions may occur with ALPROLIX. Call your healthcare provider or get emergency treatment right away if you have any of the following symptoms: difficulty breathing, chest tightness, swelling of the face, rash, or hives.

Your body can also make antibodies called "inhibitors" against ALPROLIX, which may stop ALPROLIX from working properly.

ALPROLIX may increase the risk of formation of abnormal blood clots in your body, especially if you have risk factors for developing clots.

Common side effects of ALPROLIX include headache and abnormal sensation of the mouth. These are not all the possible side effects of ALPROLIX. Talk to your healthcare provider right away about any side effect that bothers you or does not go away, and if bleeding is not controlled using ALPROLIX.

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.

Please see Brief Summary of full Prescribing Information on the next page. This information is not intended to replace discussions with your healthcare provider.

ALPROLIX [Coagulation Factor IX (Recombinant), Fc Fusion Protein], Lyophilized Powder for Solution For Intravenous Injection.

FDA Approved Patient Information

ALPROLIX® /all' pro liks / [Coagulation Factor IX (Recombinant), Fc Fusion Protein]

Please read this Patient Information carefully before using ALPROLIX and each time you get a refill, as there may be new information. This Patient Information does not take the place of talking with your healthcare provider about your medical condition or your treatment.

What is ALPROLIX?

ALPROLIX is an injectable medicine that is used to help control and prevent bleeding in people with hemophilia B. Hemophilia B is also called congenital Factor IX deficiency.

Your healthcare provider may give you ALPROLIX when you have surgery.

Who should not use ALPROLIX?

You should not use ALPROLIX if you are allergic to ALPROLIX or any of the other ingredients in ALPROLIX. Tell your healthcare provider if you have had an allergic reaction to any Factor IX product prior to using ALPROLIX.

What should I tell my healthcare provider before using ALPROLIX?

Tell your healthcare provider about all of the medicines you take, including all prescription and non-prescription medicines, such as over-the-counter medicines, supplements, or herbal medicines.

Tell your doctor about all of your medical conditions, including if you:

- are pregnant or planning to become pregnant. It is not known if ALPROLIX may harm your unborn baby.
- are breastfeeding. It is not known if ALPROLIX passes into breast milk or if it can harm your baby.
- have been told that you have inhibitors to Factor IX (because ALPROLIX may not work for you).

How should I use ALPROLIX?

ALPROLIX should be administered as ordered by your healthcare provider. You should be trained on how to do infusions by your healthcare provider. Many people with hemophilia B learn to infuse their ALPROLIX by themselves or with the help of a family member.

See the **Instructions for Use** for directions on infusing ALPROLIX. The steps in the **Instructions for Use** are general guidelines for using ALPROLIX. Always follow any specific instructions from your healthcare provider. If you are unsure of the procedure, please ask your healthcare provider. Do not use ALPROLIX as a continuous intravenous infusion.

Contact your healthcare provider immediately if bleeding is not controlled after using ALPROLIX.

What are the possible side effects of ALPROLIX?

Common side effects of ALPROLIX include headache and abnormal sensation in the mouth.

Allergic reactions may occur. Call your healthcare provider or get emergency treatment right away if you have any of the following symptoms: hives, chest tightness, wheezing, difficulty breathing, or swelling of the face.

ALPROLIX may increase the risk of forming abnormal blood clots in your body, especially if you have risk factors for developing blood clots.

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

These are not all the possible side effects of ALPROLIX. Talk to your healthcare provider about any side effect that bothers you or that does not go away.

How should I store ALPROLIX?

Store ALPROLIX vials at 2°C to 8°C (36°F to 46°F).

Do not freeze.

ALPROLIX vials may also be stored at room temperature up to 30°C (86°F) for a single 6 month period.

If you choose to store ALPROLIX at room temperature:

- Note on the carton the date on which the product was removed from refrigeration.
- Use the product before the end of this 6 month period or discard it.
- Do not return the product to the refrigerator.

Do not use product or diluent after the expiration date printed on the carton, vial or syringe.

After Reconstitution:

- Use the reconstituted product as soon as possible; however, you may store the reconstituted product at room temperature up to 30°C (86°F) for up to 3 hours. Protect the reconstituted product from direct sunlight. Discard any product not used within 3 hours after reconstitution.
- Do not use ALPROLIX if the reconstituted solution is cloudy, contains particles or is not colorless.

What else should I know about ALPROLIX?

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

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time, will make you feel good. These activities can include spending time in nature, eating well, exercising, pursuing spiritual goals, increasing creativity, getting involved in your community, and improving your sleeping patterns. The goal of these activities is to have fun, feed your spirit, relax, breathe deeply, stay connected, lighten up, and help others. Remember that change usually takes time. Give yourself time, be patient, be compassionate. Treat yourself as you would your best friend.

If you need to improve your sleep, try an improved “sleep hygiene” routine. Cognitive behavioral therapists and physicians may be skilled in coaching you through modifying your sleep hygiene. Healthy sleep hygiene includes having a regular bedtime and waking time; “smart” napping and fighting after-dinner drowsiness; increasing your daytime exposure to natural light; boosting melatonin at night by making the room dark (no phone, TV, computer, e-reader); keeping your bedroom quiet, cool, and comfortable; refraining from big late-night meals; avoiding alcohol before bed; eliminating caffeine 4 to 6 hours before bedtime; and refraining from smoking.

Make relaxing your goal, not sleep, and try to postpone worrying or brainstorming. See your doctor if you experience persistent problems such as daytime sleepiness or fatigue, loud snoring, difficulty falling or staying asleep, frequent headaches, crawling sensations in your legs or arms at night, inability to move while falling asleep, physically acting out dreams during sleep, or falling asleep at inappropriate times.

Medication versus No Medication?

Whether to treat your symptoms with an antidepressant medication is a topic about which many people are passionate. There are many reasons some folks don't believe in taking medication. Other folks just want a pill so it can make them feel better. Healing is a process in which we need to stay engaged and open-minded. It's fine to try the path of least resistance, but if that doesn't work, be willing to reassess and make adjustments. Be an active, open-minded partner in your recovery from depression. There are myriad antidepressants from which your healthcare provider can choose to tailor a medication protocol that works for you. Proper follow-up after an antidepressant is started is to see your healthcare provider after two weeks, four weeks, and then monthly until symptoms are improved and side effects are minimized. Then, regular follow-up annually.

Sometimes, after symptoms have been relieved for an extended period, you and your healthcare provider may choose to evaluate whether you can safely discontinue your antidepressant and try taking a “drug holiday.” Sometimes your symptoms can return, and you and

your provider may choose to change your medication or dose. As with any medication, it's important to have a responsive, attentive healthcare provider and to promptly communicate all medication problems and changes.

Psychotherapy

People sometimes struggle with choosing whether to treat depression by engaging in psychotherapy. Some folks are adverse to psychotherapy because they have had, or know someone who has had, a negative experience with it. Some don't believe psychotherapy will help, they don't want to talk, they're too busy, or they are too ashamed.

There are many types of psychotherapy. How to choose? Again, talk to people. You will probably be surprised to learn that someone you know has seen a therapist they've liked. Get recommendations from people you respect. Interview several therapists to find someone you think may be a good fit and you can afford. After you have selected a therapist, show up, and, when it gets rough—which it may—don't quit.

Social Engagement

Reaching out to people with depression is a top priority. The AzHTC study found that having social support decreases the risk for depression by 80% and being employed decreases the risk by 84%. Fortunately, the bleeding disorder community is a well-organized network of support organizations that promote social interaction, education, and self-reliance. Many HTCs currently implement effective screening, education, and interventions for depression care and management. Regionally and nationally, there is a focus on improving comprehensive care and expanding psychosocial support. The psychosocial program at the AzHTC in Tucson includes screening all patients for depression and anxiety at every comprehensive clinic visit. Our screening includes two questionnaires and a short clinical interview, scheduling a follow-up appointment for people who need and want further help, and providing education regarding treatment options both in and out of the HTC.

Many people with hemophilia have said summer camp changed their lives, giving them a supportive peer group and a sense of self-worth—much needed by young people with a bleeding disorder. One mother reported her son's perspective was changed by being a camper, and later a counselor, at *Dream Street* (a camp for kids with serious medical conditions, mostly cancer). Each year a few kids failed to return to camp because they had died. Rather than feel sorry for himself, her son felt lucky that he “only had hemophilia.” Volunteering at summer camp also contributes to feelings of self-worth. Service to others is a great way to enlarge our perspective and possibly alleviate feelings of depression.

Here's some good advice from patients who participated in the HERO study: live life as normally as possible, keep active, get involved in the community, and educate those around you.

Coping Skills

Our ability to cope with stress, anxiety, and depression starts with acknowledging and accepting our experience in each moment. More advice from patients who participated in the HERO study: enjoy life, communicate openly, “embrace” hemophilia, and “get into” treatment.

Parents have the enormous responsibility of raising healthy children. This is sometimes compounded by the existence of a chronic illness like hemophilia. They have the equally enormous responsibility of letting go of these same children—and helping them achieve successful independence. Children cope with developing into productive members in their families, schools and communities, and, finally, in the larger society. We all have to learn how to productively negotiate emotionally and socially from the time of infancy into adulthood. Learning how to transition through life stages and within varied roles is a skill that constantly needs to be honed. We don't always get it right, but with an open mind, we can always learn. Developing the skills necessary to make conscious choices and strategizing to make desired changes in our lives is a process that takes a willingness to tolerate hard times and emotional upheavals, as well as awareness and practice.

Depression care is a team effort with the individual as the leader. Get educated about depression. Get screened. Healthcare providers need to improve screening, follow up with findings and medication prescriptions, and collaborate with patients and other healthcare providers. Patients need to show up and challenge themselves to do the work. Remember, change can be difficult. Participate in proper treatment and don't give up until you get it right.

Chris's Outcome

Balancing recovery and pushing himself to reach his goals requires Chris Bombardier to process a broad range of positive and negative emotions. He attributes his initial and sustained recovery from depression to his wife's support, the resources at his HTC, and finding his passion for the outdoors. He finds his time in nature “good for



his soul.” He engaged in psychotherapy, became active in the hemophilia community, and challenged himself to set and achieve goals.

Chris hopes that other people with hemophilia will find relief from depression by talking about their experiences and finding the strength to acknowledge their condition and move on. Chris has learned that it's okay to struggle with his feelings of anger, frustration, and

guilt. With proper self-care, these feelings are no longer overwhelming. 🏹

Maria Iannone is a licensed professional counselor at the Arizona Hemophilia and Thrombosis Center (AzHTC) in Tucson, Arizona, where she has worked for nine years. She treats adults, children, and families. She has developed a comprehensive mental healthcare management program for patients with hemophilia at the AzHTC and engages in psychosocial research projects. She is the lead author of the peer-reviewed article “The Prevalence of Depression in Adults with Hemophilia,” and she wrote the nationally disseminated NHF Collaborating in Care module on depression and anxiety. Maria completed her graduate study in counseling psychology, an internship at the Blake Foundation in which she specialized in the assessment and treatment of children and the use of parent/child psychotherapy, and an internship in the Psychosocial Oncology Program at the University of Arizona Cancer Center, in which she specialized in assessment and treatment of breast cancer survivors. Maria draws from both her clinical experience and previous biomedical research experience.

1. <http://www.herostudy.org>.
2. Richard F. Ramsay, Brian A. Tanney, William A. Lang, and Tarie Kinzel, *Suicide Intervention Handbook* (Livingworks, 1999).
3. R. A. Remick, “Diagnosis and Management of Depression in Primary Care: A Clinical Update and Review,” *Canadian Medical Association Journal* 167 (2002): 1253–60.
4. <https://www.nimh.nih.gov>.

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NATHAN'S IXperience™



Nathan and his doctor discussed the **98% RECOVERY*** of IXINITY



HE'S INFUSING 4,000 IU* with IXINITY—less than in the past



He thinks the IXINITY reconstitution device is **EASY TO USE**

“When I heard the recovery rate was 98%, I thought, ‘Wow, that’s pretty close to the recovery of a plasma-derived product.’”

Watch Nathan's videos at PatientIXperiences.com

Nathan's experience with IXINITY may not be typical. Speak with your doctor to see if IXINITY may be a good option for you.

*Nathan uses 4000 IU per infusion. IXINITY recovery is an average based on lab tests of patients in the clinical study.¹ Your actual recovery and dose may be different. Speak with your healthcare professional about the right dose for you.

INDICATIONS AND IMPORTANT SAFETY INFORMATION

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.

Reference: 1. IXINITY [coagulation factor IX (recombinant)] prescribing information. Winnipeg, MB, Canada: Emergent BioSolutions Inc.; April 2015.

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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**

- Lightheadedness
- Dizziness
- Nausea
- Fainting

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?

Store IXINITY 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.

Do not use IXINITY if the reconstituted solution is not clear and colorless.

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.

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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Anylam Announces Positive Results in their Phase I Trial of Fitusiran



Anylam Pharmaceuticals has announced positive results from its Phase I clinical study of fitusiran, an RNA-interference drug for treatment of hemophilia A and B, including patients with inhibitors. Fitusiran reduces the amount of antithrombin, an inhibitor of clotting, in the circulation. This is expected to allow the blood to clot more easily, even in the absence of factor VIII or factor IX. In 17 patients without inhibitors receiving a fixed monthly, subcutaneous dose of 80 mg of fitusiran the results showed a mean antithrombin lowering of 87% and a median estimated annualized bleed rate (ABR) of zero. Six hemophilia A and B patients with inhibitors receiving a lower dose of fitusiran in another part of the study showed a mean antithrombin lowering of 81% and a significant reduction in ABR. The product was generally well tolerated and showed no evidence of thromboembolic complications (uncontrolled clotting). Anylam plans to initiate a Phase III study in early 2017.

Biogen to Spin Off Hemophilia Business as Bioverativ



Biogen has announced that it will spin off its hemophilia business unit, including Alprolix, its longer-acting factor IX product, as a new company named Bioverativ. The new company is expected to launch in early 2017. Bioverativ will continue R&D efforts into next-generation longer-acting products and gene therapy for hemophilia A and B. It will also continue Biogen's previous sponsorship of the My Life, Our Future genotyping program and their humanitarian aid program, which provides factor products to developing countries.

CSL Reports Additional Data from their Phase III Study of IDELVION



CSL Behring has reported additional data from their Phase III study of IDELVION, their new longer-acting factor IX product. IDELVION is a recombinant protein fusing factor IX to albumin to prolong its half life. The results suggest that treatment intervals of up to 21 days at a dose of 100 IU/kg are conceivable in older patients. In addition, IDELVION was studied in 19 patients undergoing 21 major or minor procedures. In all but

one case, a single pre operative dose of IDELVION was sufficient to maintain adequate factor IX levels throughout the procedure. There was no evidence of inhibitor development in any patient.

Emergent BioSolutions Announces Spin Off of Aptevo Therapeutics



Emergent Biosolutions has completed the spin off of its hematology and immunology businesses, including its factor IX product IXINITY, as a new company named Aptevo. Aptevo debuted as an independent company on 8/1/16.

Novo Nordisk Has Filed a U.S. License Application for their Longer-Acting Factor IX



Novo Nordisk has submitted a Biologics License Application (BLA) for their longer-acting pegylated factor IX product. The new product has been called N9-GP during development and has a proposed generic name of nonacog beta pegol. In a clinical study of 115 patients with severe or moderately severe hemophilia B, the product was efficacious in routine prophylaxis, treatment of bleeding episodes and surgery for adults. The product was well tolerated and showed no signs of inhibitor development. The half-life was found to be about five times longer than standard-acting factor IX products.

Shire Announces the Discontinuation of Baxalta's BAX 335 Gene Therapy Project



Shire, who acquired Baxalta earlier this year, has announced the discontinuation of Baxalta's BAX 335 gene therapy project. The reason is the inconsistency of the expression levels seen in their Phase I/II clinical study and the observation of a decrease in factor IX expression over time in some patients. Although one subject achieved factor IX levels of 20 - 25%, other patients showed much lower levels. Shire will continue R&D on gene therapy for hemophilia with hopes of initiating a next-generation gene therapy program in the future.

Girls Can Have Hemophilia B Too!

By: Lori Long

I am a woman with hemophilia B. I was also a little girl with hemophilia B, only we didn't know it. What we did know was that I had nosebleeds that lasted for days and joints that would swell up randomly. When I reached adolescence, my menstrual cycle was unbearable... the embarrassment and humiliation were sometimes overwhelming. My parents told every doctor throughout my childhood that my father had hemophilia B and that I exhibited some of the same symptoms. The doctors would in return tell my parents that girls just don't get hemophilia, and I was "just a bleeder." It's funny, I now identify myself as a "bleeder". The difference is that now I'm a bleeder with a formal diagnosis and access to medication. I never see it as being "just" a bleeder. I believe being a bleeder makes me a little tougher, a little more compassionate, a little more giving.

Since I wasn't diagnosed until I was 36 years-old, I have pretty significant joint damage. Am I angry about this? Sometimes. I also realize that having a bleeding disorder has given to me as much as it has taken from me. I recently had dinner with a friend who was told as a young girl that she was not a carrier based on a genetic test. She recently had her first child, a boy, and he has been diagnosed with hemophilia! Imagine her surprise! So even if you get checked early in life, things can still happen. During our conversation, I found myself feeling very emotional as I came to the realization (and shared with her) that having a bleeding disorder gives us extra angels.

So when I'm angry, I remember my blood brothers and sisters. I remember those of you who have e-mailed,



Facebook messaged, texted, or even gotten on the phone with me when I'm on my fourth stick and staring down the fifth, and I just don't know if I can do it. I remember the love I feel when I walk into the lobby of the hotel at a national event, and my hemophilia family spots me and comes over to hug me. I remember those of you who stood with me while I cried because my son was receiving an infusion without me there for the first time because I had chosen to travel without him. I remember comparing notes with other bleeders on how our bleeds progress and what we all do about them. I remember having tea while laughing and crying with my blood sisters at the retreat for women with hemophilia B. And I really believe, as I'm thinking these thoughts, that all of you angels would not be in my life if I didn't have hemophilia B. I am grateful for all of you, every single day. 🍵

DON'T STAND ON THE SIDELINES

LEARN IF RIXUBIS
MAY BE RIGHT FOR YOU

Visit RIXUBIS.com to find your
local Baxalta sales representative

Indications for RIXUBIS [Coagulation Factor IX (Recombinant)]

RIXUBIS is an injectable medicine used to replace clotting factor IX that is missing in adults and children with hemophilia B (also called congenital factor IX deficiency or Christmas disease).

RIXUBIS is used to control and prevent bleeding in people with hemophilia B. Your healthcare provider may give you RIXUBIS when you have surgery. RIXUBIS can reduce the number of bleeding episodes when used regularly (prophylaxis).

Detailed Important Risk Information

You should not use RIXUBIS if you are allergic to hamsters or any ingredients in RIXUBIS.

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 nursing, are pregnant or planning to become pregnant, or have been told that you have inhibitors to factor IX.

Allergic reactions have been reported with RIXUBIS. Call your healthcare provider or get emergency treatment right away if you get 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 factor IX. An inhibitor is part of the body's defense system. If you form inhibitors, it may stop RIXUBIS from working properly. Consult with your healthcare provider to make sure you are carefully monitored with blood tests for development of inhibitors to factor IX.

If you have risk factors for developing blood clots, the use of factor IX products may increase the risk of abnormal blood clots.

Common side effects that have been reported with RIXUBIS include: unusual taste in the mouth, limb pain, and atypical blood test results.

Call your healthcare provider right away about any side effects that bother you or if your bleeding does not stop after taking RIXUBIS.

Please see following page for RIXUBIS Important Facts.

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.

Baxalta

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RIXUBIS
[COAGULATION FACTOR IX
(RECOMBINANT)]

MOVING FORWARD

RIXUBIS

[COAGULATION FACTOR IX (RECOMBINANT)]

FDA-approved Patient Labeling

RIXUBIS [Coagulation Factor IX (Recombinant)]

This leaflet summarizes important information about RIXUBIS. Please read it carefully before using this medicine. This information does not take the place of talking with your healthcare provider, and it does not include all of the important information about RIXUBIS. If you have any questions after reading this, ask your healthcare provider.

What is RIXUBIS?

RIXUBIS 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 blood from clotting normally. RIXUBIS is used to prevent and control bleeding in people with hemophilia B. Your healthcare provider may give you RIXUBIS when you have surgery. RIXUBIS can reduce the number of bleeding episodes when used regularly (prophylaxis).

Who should not use RIXUBIS?

You should not use RIXUBIS if you

- are allergic to hamsters
- are allergic to any ingredients in RIXUBIS.

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

What should I tell my healthcare provider before using RIXUBIS?

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 RIXUBIS passes into your milk and if it can harm your baby
- are pregnant or planning to become pregnant. It is not known if RIXUBIS may harm your unborn baby
- have been told that you have inhibitors to factor IX (because RIXUBIS may not work for you).

How should I infuse RIXUBIS?

RIXUBIS is given directly into the bloodstream. RIXUBIS 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 RIXUBIS by themselves or with the help of a family member.

Your healthcare provider will tell you how much RIXUBIS 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 RIXUBIS to be sure that your blood level of factor IX is high enough to clot your blood. Call your healthcare provider right away if your bleeding does not stop after taking RIXUBIS.

What are the possible side effects of RIXUBIS?

Allergic reactions may occur with RIXUBIS. Call your healthcare provider or get emergency treatment right away if you get a rash or hives, itching, tightness of the throat, chest pain or tightness, difficulty breathing, lightheadedness, dizziness, nausea or fainting. Some common side effects of RIXUBIS were unusual taste in the mouth and limb pain. Tell your healthcare provider about any side effects that bother you or do not go away. These are not all the side effects possible with RIXUBIS. You can ask your healthcare provider for information that is written for healthcare professionals.

What are the RIXUBIS dosage strengths?

RIXUBIS comes in five different dosage strengths: 250, 500, 1000, 2000 and 3000 international units. The actual strength will be imprinted on the label and on the box. The five different strengths are color coded, as follows:

Light Blue	Dosage strength of approximately 250 international units per vial
Pink	Dosage strength of approximately 500 international units per vial
Green	Dosage strength of approximately 1000 international units per vial
Orange	Dosage strength of approximately 2000 international units per vial
Silver	Dosage strength of approximately 3000 international units 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 RIXUBIS?

- Store at refrigerated temperature 2° to 8°C (36° to 46°F) for up to 24 months. Do not freeze.
- May store at room temperature not to exceed 30°C (86°F) for up to 12 months within the 24 month time period. Write on the carton the date RIXUBIS is removed from refrigeration. After storage at room temperature, do not return the product to the refrigerator.
- Do not use after the expiration date printed on the carton or vial.
- Reconstituted product (after mixing dry product with wet diluent) must be used within 3 hours and cannot be stored or refrigerated. Discard any RIXUBIS left in the vial at the end of your infusion.

What else should I know about RIXUBIS?

Your body may form inhibitors to factor IX. An inhibitor is part of the body's defense system. If you form inhibitors, it may stop RIXUBIS from working properly. Consult with your healthcare provider to make sure you are carefully monitored with blood tests for the development of inhibitors to factor IX.

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

Resources at Baxalta available to patients

For information on patient assistance programs that are available to you, including the Baxalta CARE Program, please contact the Baxalta Insurance Assistance Helpline at 1-888-229-8379.

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Baxalta

Coalition for Hemophilia B

Meetings on the Road

SAVE THE DATES!

Minneapolis, MN Family Meeting

Saturday, September 17, 2016

Hilton Minneapolis/Mall of America
3800 American Blvd. East
Bloomington, MN 55425
\$129 per night, plus tax
(952) 854-2100

St. Louis, MO Family Meeting

Saturday, October 22, 2016

Marriott St. Louis Airport
10700 Pear Tree Lane
St. Louis, MO 63134
\$110 per night, plus tax
(314) 423-9700



Austin, TX Family Meeting

Saturday, November 5, 2016

Austin Marriott North
2600 La Frontera Blvd.
Round Rock, TX 78681
\$105 per night, plus tax
(512) 733-6767

Chicago, IL Family Meeting

Saturday, November 12, 2016

Chicago Marriott Schaumburg
50 N. Martingale Road
Schaumburg, IL 60173
\$89 per night, plus tax
(847) 240-0100

Registration for these meetings is FREE and includes parking, breakfast, lunch, afternoon snack and dinner.

Gas and tolls reimbursed for families attending.

For more details and registration, please visit our website at www.hemob.org
or call Chris Villarreal at 347-949-9696

Please note that the Coalition for Hemophilia B does not cover airfare or hotel accommodations.

Pool on premises, don't forget your bathing suits!

Meetings are sponsored by a generous grant from



Fun in the Sun!

M C B E A C H W Y N X M Y J G
F A M I L Y B I K I N I T T Y
O M H A E Q B A S E B A L L S
C P L R B F C T F C H Z W V U
E I Y B J E B L G P T U M N N
A N B A T H I N G S U I T X G
N G B R F B S A J L Y F U Z L
S V I B K T W V S K A I P K A
W X C E F I S H I N G E R I S
I M E C B Z K B L S S W M V S
M X C U P S A N D C A S T L E
M G R E T F U N P H P F I V S
I C E T Q Q H U L A H O O P W
N B A S O C C E R H S J C V M
G H M N J L E V O S M C Z N A



Word Bank

BARBECUE
BASEBALL
BATHING SUIT
BEACH

BIKINI
CAMPING
FAMILY
FISHING

FUN
HULA HOOP
ICECREAM
OCEAN

SANDCASTLE
SOCCER
SUNGLASSES
SWIMMING

Summer Word Scramble

AUTSGU

UNSHSINE

CATIONVA

LLABVOELLY



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For the latest **B Scene** video sharing real life stories of members in our community, please visit our social media sites:

Website: www.hemob.org

Facebook: www.facebook.com/HemophiliaB/

Twitter: <https://twitter.com/coalitionhemob>

For more information, please contact Kim Phelan:

kimp@hemob.org or call 917-582-9077



Wishing You a Wonderful Autumn!