GROWING OLDER WITH BLEEDING DISORDERS
AGING WELL WITH HEMOPHILIA

Today, people with hemophilia and other bleeding disorders are living longer, fuller lives because of advances in factor treatments. However, as people with bleeding disorders get older, they face the same health issues that other people do. We spoke with Nurse Practitioner Angela Lambing to learn more about what people with bleeding disorders can do to help them age well.

STAYING HEALTHY INTO YOUR GOLDEN YEARS

Follow these tips to help you stay healthy as you get older.

THE MANY CHAPTERS IN THE LIFE OF BERNIE FINN

Bernie Finn’s life has been a roller coaster. He is 63, with hemophilia and human immunodeficiency virus (HIV), and he is confined to a wheelchair. But you won’t find Bernie complaining. In fact, he feels more fortunate than many people he has known. Read about Bernie’s life with hemophilia and the happy turn it has recently taken.

JUDY & SUSI: A MOTHER/DAUGHTER TEAM TAKES ON VON WILLEBRAND DISEASE

Judy Madsen and her daughter, Susi Prola, have been facing von Willebrand disease (VWD) together for nearly a decade. They both have rare type 2M VWD. Read about how Susi helps her mom manage her condition and get the care she needs.
To learn more about aging with bleeding disorders, we spoke with Angela Lambing, MSN, RNCS, NP-C. Lambing is the nurse practitioner and coordinator of the Henry Ford Hospital Adult Hemophilia and Thrombosis Treatment Center in Detroit. She is certified in both adult primary care and geriatric care.

Bleeding Disorders and Healthy Aging

Lambing says that the most common aging issues for people with bleeding disorders are similar to the issues other people face. These include diabetes, high blood pressure, heart disease, and cancer. And as for other people, there are many ways that bleeding disorder consumers can help maintain their health as they age.

“People need to look at healthy lifestyle choices. These choices can help reduce the chances of having diabetes and high blood pressure. They can also reduce the pain of joints affected by arthritis,” Lambing says.

Lambing counsels her older hemophilia patients to take the following steps to stay healthy:

- Maintain a healthy lifestyle. This includes avoiding alcohol and smoking.
- Follow a healthy diet to keep your weight down.
- Have a primary care provider in addition to providers from your hemophilia treatment center [HTC].* Also have other specialists as needed.
- Get screened for cancer and other conditions related to aging.
- Keep up with your vaccinations.
- Take factor as prescribed by your HTC.
- Maintain a regular exercise program within your limitations.

As different aging issues arise, people with bleeding disorders may find that they need care from several providers. These may include specialists in diabetes, heart, liver, or kidney care. Communication between these specialists, the patients’ primary care provider [PCP], and care providers from their HTC is key.

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This same level of communication is needed when people with bleeding disorders have other testing. This includes a colonoscopy,* as well as screening for prostate cancer, which is the number one type of cancer in men as they age. In addition, Lambing suggests that patients talk with their healthcare team about checks for thyroid function, bone density, vision, hearing, and kidney function. Each of these areas is affected by age. Some of these checks are especially important for patients with HIV and hepatitis C (see page 6).

“People may also want to consider supplements such as calcium. This is because bone density can decrease as we age. There is also increased focus on vitamin D because it helps the bones absorb calcium for bone health. People should discuss these options with their healthcare providers.”

Controlling Pain

Managing pain has always been a concern for people with bleeding disorders because of the pain caused by joint bleeds. Lambing says that when those same joints become affected by arthritis, pain can be an even greater issue. Lambing and her colleague,
between addiction and dependence. For think that taking the medicine every day don't understand the term addiction. They says Lambing. “This is largely because people concern about addiction.

The fear of addiction can be a barrier when opioids should be considered without misusing it, which is addiction.”

Lambing cites a national study that examined the risk of addiction and misuse of opioids. The study stated that the risk is about 3% in the general population. Lambing has found the same to be true in the hemophilia population.

“We forget that 97% of hemophilia patients don’t misuse pain medications and manage very well.”

**HIV and Hepatitis C**

For older people with bleeding disorders, health issues may include infection with hepatitis C or human immunodeficiency virus (HIV). This is because of transmission of these viruses through factor used in the 1970s and ’80s. Fortunately, things have changed since those days. According to the National Hemophilia Foundation, no transmissions of HIV through factor products have occurred since those days. According to the National Hemophilia Foundation, no transmissions of HIV through factor products have occurred since 1986. And there have been no reports of hepatitis C transmission through factor that has been treated with new screening processes. And there’s more good news – patients who do have these conditions are enjoying much better health due to improved treatments.

“Treatment for AIDS and hepatitis C is better than it’s ever been, so many of these patients are living into their senior years,” Lambing says. “This is often true even if the patients also have diabetes, high blood pressure, heart disease, hepatitis C, kidney disease, or cancer.”

**Other Aging Issues**

In addition to addressing health issues, growing older with hemophilia involves planning for the future. This doesn’t mean just planning finances. It also means paying attention to living arrangements and learning about Medicare.

“Planning for physical safety as patients age is important,” Lambing say. “This can mean changing to a home without stairs. It can also mean being sure to get regular exercise to maintain strength and balance. Cognitive* health is just as important. To stay mentally and socially active, people can continue working, volunteer, or take other steps.”

As for Medicare, some older people with a bleeding disorder are already on the program because of disability. Others will be reaching age 65 and starting on it. Medicare Part A pays for hospital stays, and Part B covers outpatient treatments. Part D pays for prescription drugs. Factor is covered by Part B (not Part D), but there are co-payments involved that can be quite high. Lambing suggests that people with bleeding disorders nearing the age of 65 should look at medical insurance supplemental plans to cover these costs.

Another issue for aging hemophilia patients can be the overall management of their health. It can be a challenge to keep all of a patient’s providers informed, as well as track all of the care information they provide.

“When people are trying to balance all of their care, they may need somebody, such as the patient’s provider, to take a lead role in coordinating care.”

*Please see Glossary on page 8.
A New Golden Age

Although people with bleeding disorders have learned to listen to their body about bleeds, aging can bring new, unknown symptoms into the mix. Communicating well with their providers can help people learn more about what happens to their bodies as they age. So, if you’re getting older with a bleeding disorder, be sure to ask questions. Request help when you need it.

And of course – don’t forget to enjoy your golden years.

*Glossary*

- **Cardiac catheterization**: A procedure used to test heart function. A long, narrow tube is inserted into a blood vessel and guided into the heart.
- **Cognitive**: Relating to mental activity.
- **Colonoscopy**: A test that is typically used to screen for colon cancer. A long, narrow tube with a camera on the end is inserted through the rectum into the colon. This test is recommended for all people over age 50.
- **Hematologist**: A doctor who treats liver disease.
- **Hemophilia treatment center (HTC)**: A healthcare center for people with bleeding disorders. Funded by the federal government, HTCs have specialists who are trained in the diagnosis and treatment of people with bleeding disorders. These specialists include hematologists, nurses, social workers, and physical therapists.
- **Human immunodeficiency virus (HIV)**: The virus that causes acquired immunodeficiency syndrome (AIDS).
- **Immune tolerance therapy (ITT)**: A treatment for people with an inhibitor (see below). With ITT, a person is exposed repeatedly to the deficient clotting factor. The goal is to “teach” the body to tolerate the factor. If the treatment succeeds, replacement therapy can be used to prevent or control bleeding.
- **Inhibitor**: A somewhat common complication in people with bleeding disorders. An inhibitor is an antibody (immune system cell) that the body produces in response to clotting factor treatment. The inhibitor treats the factor as a foreign substance and tries to destroy it. This prevents the factor from working.
- **Opioids**: Medications that can relieve pain. They may be used to treat pain that doesn’t respond well to other types of pain medications. Opioids are a type of narcotic.
- **Secondary prophylaxis of factor**: A treatment of factor that is used to manage pain. It is used in addition to primary prophylaxis. Primary prophylaxis consists of regularly scheduled treatments of factor on two or three days of the week.
- **Tainted plasma**: Plasma that is infected with a disease. In the 1970s and ’80s, many people with bleeding disorders became infected with HIV or hepatitis C through their factor. Fortunately, HIV transmission by any factor VIII or IX product in the U.S. has not occurred since 1986. This is due to improved blood-product treatment practices. [Source: The National Hemophilia Foundation, www.hemophilia.org]


*Please see Glossary on page 11.

BY VALERIE HANSEN | CONTRIBUTING WRITER

**THE MANY CHAPTERS IN THE LIFE OF BERNIE FINN**

Bernie Finn’s life has been a roller coaster. He is 63, with hemophilia and human immunodeficiency virus (HIV), and he is confined to a wheelchair.

But you won’t find Bernie complaining. In fact, he feels more fortunate than many people he has known.

“I think my attitude goes back to when I was in grammar school,” he says. “I wasn’t having many hemophilia issues, and I wasn’t in a wheelchair. My only issue was that one of my legs was shorter than the other because of a subdural hematoma* I had when I was born. However, this was before the EEOA [Equal Education Opportunities Act*] was passed. At that time, ‘handicapped’ kids were separated from the ‘normal’ kids, and hemophilia put me in the ‘handicapped’ category. As a result, I went to school with kids who were much more disabled than I was. They had muscular dystrophy and polio and such. I always felt normal because everybody else was so disabled.”

The first major change in Bernie’s life came when he entered his teen years. Suddenly he began having severe bleeds. Although factor had just become available, prophylaxis* to prevent bleeds was not.

“The 1960s were probably the worst time for me, physically, emotionally and everything else,” he says. “I practically lived in the hospital because of all the bleeds. My high school years were spent in my living room with tutors. I didn’t get out at all, but at the time I didn’t know any differently.”

**His College Years**

By the time Bernie graduated from high school in 1968, the EEOA had passed. With the chance for equal education provided by the new law, Bernie felt that he was expected to go to college. However, he was not emotionally or mentally prepared for college because he hadn’t physically attended high school.
More Battles

Bernie went to work for the city in 1976 and stayed there until 2007. But in 2006 he found himself facing another mountain: HIV. Through the years, Bernie had known that he could be HIV-positive.* But he had chosen not to be tested because there wasn’t a lot to be done about it in those days. Plus, he had watched a cousin’s difficult battle with AIDS.

“I refused to be tested. I didn’t want to know,” he says. “I felt fine. I had no symptoms.”

That changed in early 2006.

“On St. Patrick’s Day, my father took me to the emergency room,” he says. “I didn’t realize how sick I was. I had double pneumonia.”

As it turned out, at some point Bernie had indeed received factor made from HIV-tainted plasma. After his diagnosis, Bernie stayed in the hospital for a month, and was off work for nine months. The HIV put him in a really dark place.

“I wanted to crawl under a rock,” he says. “I didn’t want anybody to know. I’ve gotten much better at coping over the last four or five years, but that was a bad time.”

Returning to work made Bernie even more unhappy. During the nine months he was off, new management had been brought in. Bernie didn’t like the new team and no longer had the support of family that he did. With his father’s help, he found the perfect position.

“My parents gave me everything; they were always there for me. I wouldn’t be where I am today if not for them,” Bernie says. “At the time, my dad was good friends with the mayor of Bridgeport, Connecticut, where we lived, and took me to his office. As it turned out, I was in the right place at the right time. They were looking for someone right out of college. They wanted someone without previous experience so they could train them on their particular system.”

Welcome Fran

When his father passed away, Bernie was alone in the house for the first time in his life. Plus, he was in a wheelchair. [At times during his teen years, his bleeds had put him in a wheelchair. In recent years, however, the chair had become a necessity.] Other than the Coram nurses coming in to start his infusions, Bernie had little social interaction.

He felt isolated. A short time later, a good friend of his suggested that he go on an online dating site.

“She said, ‘Bernie, you are too special to spend your life alone. There’s got to be someone out there for you.’ I took her advice and met Fran two weeks later. We’ve been together for almost a year now. My life has changed so much since she has come into it.”

Bernie and Fran support each other in all ways. He was comfortable enough with her to tell her about his hemophilia and HIV shortly after they met. She didn’t bat an eye.

“Fran shared some of her life experiences with me, and again, in some respects I feel I was luckier than her,” Bernie says. “She didn’t have the support of family that I did. We have something really special between us.”

When Bernie retired, he still shared his parents’ house with his father [his mother had died in 1989]. In September 2012, Bernie’s father was admitted to the hospital. Three days later, Bernie had a severe knee bleed and ended up in the hospital himself. He spent a month in the physical therapy wing. Both he and his father returned home, but in April 2013, his father died.

Bernie Finn has been through a lot in his 63 years. He has not only survived, but is thriving. With his HIV and hemophilia well-controlled by medication and a loving partner on his arm, Bernie is welcoming the future with a big smile on his face.

*Glossary

- **Equal Education Opportunities Act (EEOA):** A 1974 law that made it illegal to discriminate against students for reasons that include race, gender, or disability. It introduced “mainstreaming.” This meant placing students with physical disabilities into the main school population.
- **Hemorrhage:** An episode of bleeding that is fast and hard to control.
- **HIV-positive:** Having the human immunodeficiency virus, the virus that causes AIDS. In the 1970s and ’80s, many people with bleeding disorders became infected with HIV through their factor infusions. However, HIV transmission by any factor VIII or IX product in the U.S. has not occurred since 1986. This is due to improved blood product treatment practices. [Source: The National Hemophilia Foundation, www.hemophilia.org]
- **Prophylaxis:** Treatment to prevent bleeds in people with a bleeding disorder. It may consist of scheduled treatments of factor on two or three days of the week.
- **Subdural hematoma:** A collection of blood in the space around the brain. This type of brain injury can be life-threatening.
JUDY & SUSI
A Mother/Daughter Team Takes on von Willebrand Disease

BY VALERIE HANSEN | CONTRIBUTING WRITER

Judy Madsen and her daughter, Susi Prola, have been facing von Willebrand disease (VWD) together for nearly a decade. Susi was diagnosed first in 1996 at age 28. Her mother was finally diagnosed 10 years later, at age 61. They both have rare type 2M VWD.

It's possible that neither would be diagnosed today if Susi hadn't ended up in the emergency room one day nearly 20 years ago. On that day, Susi had a miscarriage and started to hemorrhage (bleed heavily).

"I was in the emergency room, and a young doctor just out of medical school was taking my doctor's place," Susi says. "When she started asking me questions, she seemed to know exactly what my experience had been. I looked around, like, 'Have you been following me?' She had read about von Willebrand's in medical school and suspected I had it. Unlike my other doctors, she asked all the right questions."

After talking to the doctor, Susi learned that getting a blood transfusion when she had her tonsils out at age five perhaps wasn't normal. She also learned that starting on birth control at age 13 because her periods were so heavy probably wasn't normal either. VWD would also explain the huge hematomas* that she had when her wisdom teeth were pulled at age 15. And it would explain the multiple blood transfusions she received after the birth of her children.

Even with all of these clues, Susi wasn't diagnosed until after her miscarriage. She was referred to a hematologist* at a local hemophilia treatment center (HTC).* After she was diagnosed, Susi began to search for more facts about VWD. These days she is teaching other people – parents, relatives, and often doctors – about the disease.

"Although it is not as rare as we were once told, von Willebrand's is often hard to diagnose," Susi says. "There are so many ways to get a false negative result, especially with type 2. Unlike types 1 and 3, we don't always have deficient factor. We have defective factor. So the doctors really have to know what to look for to get a correct diagnosis."

"Susi is very intelligent, and she isn't shy about telling people what she knows about von Willebrand's," says her mother, Judy. "Doctors admire her because she's right on top of it. They definitely listen to her."

After Susi’s Diagnosis

“When I was diagnosed, I knew right away that my kids had von Willebrand's because of the symptoms," Susi says. "They had baseball-sized hematomas from mild injuries. Their mosquito bites swelled to the size of silver dollars."

Once her son and daughter were diagnosed, Susi started working on her mother.

"I was a little skeptical," Judy says. "But when Susi went over some of the symptoms, it surprised me. When I was young, 10 days on a menstrual cycle was nothing to me. My mom was like that, too, so I think she also had it. One of my relatives was researching the genealogy* of the family. She found that my grandmother on my father's side died of a bleeding disorder."

Getting her mother diagnosed was a rocky road for Susi. The HTC nearest to her mother was a two-hour drive away. So they started at the local hospital. The tests they had done there came back negative. Then they went to a hospital in a bigger town nearby. Again, the results came back negative.

“It turned out that both hospitals had sent the blood sample to the same lab to be tested," Susi says. "I had learned enough by that time that I could tell just by the way the results read that they weren't doing it right." (Many labs don't have experience doing this highly specialized test.)

Finally, Judy agreed to go with Susi to her HTC. Susi knew that her mother’s anxiety about medical tests could impact the results. This is because of the surge in adrenaline* that anxiety can cause. To calm her down, Susi took her mom out to lunch before they went for the test. However, there was another hormonal issue that Susi didn't know about, because Judy didn't realize it was important. Judy had been taking estrogen for a number of years after a hysterectomy. Estrogen is another hormone that can raise factor levels and affect testing for VWD. When they

vom Willebrand disease (VWD) is a type of bleeding disorder. Von Willebrand factor (VWF) is a protein in the blood that helps clots form. People with VWD either don't have enough VWF in their blood (deficient), or their VWF doesn't work properly (defective).

- Type 1 VWD is the mildest form. With Type 1, the VWF levels are deficient.
- With Type 2, the VWF is defective. Type 2 is broken down into Type 2A, 2B, 2M, and 2N. Each of these causes a slightly different clotting issue. Type 2M is very rare.
- With Type 3, the most severe form of VWD, there is either no VWF produced in the blood, or very little.

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*Please see Glossary on page 15.
realized that the estrogen could be affecting the test results, they took care of that too.

“My mom stopped taking estrogen for six weeks to get it out of her bloodstream. Then she was retested and we finally got her diagnosed,” Susi says.

The results showed that Judy had the same type 2M VWD that Susi had.

**Susi Steps In**

Two weeks after Judy was diagnosed, she and her husband were in a serious car accident. Judy had a head injury. Before the doctors even took her in for a CT scan,* Susi took action.

“I called the hematologist and then took factor to the hospital to infuse her,” Susi says. “Our hematologist had called ahead to give orders that allowed me to infuse the proper dose of factor in the ER. If we hadn’t given her the factor, she would have had a serious head bleed.”

Susi doesn’t just ensure that Judy has factor when she needs it. She also makes sure that Judy has the proper access for infusion. This is especially important because Judy has small veins that can make infusion difficult.

“Whenever my mom has any minor surgery, I know I’m going to have to infuse her for at least a week afterwards,” Susi says. “Because of her veins, she is very hard to infuse. So, when she had open heart surgery, I asked them to put in a PICC line.* This helped because she was going to have to be infused daily for several months.”

The PICC line was placed in January, and Judy kept it in until mid-April. Around the twelfth week, her factor dose was tapered from a full dose daily to a half dose every other day. The reason she needed 16 weeks of infusions was that two weeks after her heart surgery, she was sent back to the hospital. She needed emergency gall bladder surgery.

“While I was in the hospital, Susi was at my bedside day and night,” Judy says. “She talked to the doctors about the von Willebrand’s and it was helpful to them. I got the best care. I couldn’t ask for a better daughter.”

**Living Life with VWD**

“In the past few years, Mom has gotten much better at realizing that using clotting factor shouldn’t be reserved only for surgeries or major injuries,” says Susi. “There were times when she had minor injuries when she should have infused and didn’t. That caused longer bleeding and healing time. She is learning that my constant reminder of ‘when in doubt, treat’ has merit, as she has seen the benefits of treating. Although it’s been a bumpy road at times, I feel it has helped us to connect in a very special way.”

Susi laughs and adds, “I’m sure many adult children caring for their parents know it can be challenging for both parties. Even though I know my mom trusts me, I sometimes think I will always be 12 years old in her eyes. But it’s very rewarding. I know that I’m able to help my mom and show my love by giving back a little of the care and love she’s always given to me.”

Today, Judy and Susi are relieved to finally have answers about their history of bleeding issues. In fact, they have found out that most of their family has VWD. With this powerful mother/daughter team on the case, VWD doesn’t have a chance.

*Glossary:

- **Adrenaline:** A hormone that may be released by the body in times of stress. Adrenaline can make factor levels in the blood increase.
- **CT (computed tomography) scan:** A type of test that uses x-rays to take pictures of bones and internal organs.
- **Genealogy:** The study of a family’s history and ancestors.
- **Hematologist:** A doctor who treats blood disorders.
- **Hematoma:** A collection of blood near a break in the wall of a blood vessel.
- **Hemophilia treatment center (HTC):** A healthcare center for people with bleeding disorders. Located throughout the country, HTCs are funded by the federal government. HTCs have specialists who are trained in the diagnosis and treatment of people with bleeding disorders. These specialists include hematologists, nurses, social workers, and physical therapists.
- **PICC (peripherally inserted central catheter) line:** A catheter that is inserted into the body through a vein (usually in the arm). It is threaded up to a large vein near the heart. A PICC line helps make infusion easier. PICC lines are most often used from two weeks up to six months or more.
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