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EMPOWERMENT

in a Time of Pandemic



Laurie Kelley

When you first hear devastating news—the death of a family member, loss of a job, a personal illness, the diagnosis of a bleeding disorder like hemophilia,¹ or now, the prevalence of COVID-19—the powerful feelings you have in response can hijack logical, rational thinking that might help you take good next steps. Instead of taking time to learn more, staying open-minded, and seeking solutions, we often become anxious and fearful when our emotional response goes into overdrive.

If the devastating news is on a grand scale, as with COVID-19, millions of us face strong emotions. We sit daily, glued to the TV or computer screen, reading about the escalating death rate. The nation is already polarized politically; not since the Vietnam War, perhaps, has the nation faced such divisiveness. And we have elections coming in November. All of this adds to stress, anxiety, isolation, and tension. Much of what we experience, like the initial diagnosis of hemophilia, is fear of the unknown.

We already have realized, after a number of recent suicides in the bleeding disorder community, that anxiety and depression run

deep in people with chronic disorders. Now we have the added stress of managing a new way of life with COVID-19. With children at home constantly, job loss, restriction of freedom to socialize or travel, and even cancellations of our annual or local community meetings, we are increasingly isolated. If we turn to social media for support, we may face even more tension and stress.

To manage your emotions and learn to cope with a new world order, apply some techniques you may have learned when your child was diagnosed with a bleeding disorder. You can learn to think more clearly, feel calmer, and protect yourself mentally, just as you are protecting yourself physically from COVID-19.

Feeling Powerless?

When something unwanted, unexpected, or harmful happens to you and it's not your fault, you may feel powerless. Hemophilia is something that you didn't want and possibly didn't expect.

1. The information presented here applies to any bleeding disorder, and indeed, any chronic disorder. But for simplicity in writing, we will use hemophilia as an example.



When I learned in 1987 that my son had hemophilia, my world was suddenly and completely upended. Usually, I was a logical thinker and a great planner. But hemophilia was not in my plan. My husband and I had just finished

graduate school, gotten married, and landed good jobs, but we were broke and in debt. The stress was off the charts. How would I handle this? Could I return to work? Who would care for my son? What would a bleed be like? I was terrified. But I got to work—on myself. Through the next few months and years, I collected many experiences and wisdom from family, friends, co-workers, my spouse, the medical community...everyone had something to offer. Much of it was great advice; some was unwanted and unwarranted. This highly emotional time often left me feeling burned-out and exhausted. But I found my way through it, and channeled all the emotions into something positive: a new company, a mission, and educational books and newsletters for the bleeding disorder community.

Recently, I realized that learning to cope with the hemophilia diagnosis and a completely altered life is not unlike learning to cope emotionally with the pandemic. Early this year, life went on as planned. Then suddenly, our world changed. When I began learning to live with a chronic disorder, I realized two key things: (1) My attitude and mindset about hemophilia contributed a lot to my anxiety. (2) Communication with others depended on how I responded to their comments and behavior. So I learned how to be more self-aware, set boundaries, take responsibility, get educated about hemophilia, and communicate my needs and wishes to others in a positive way.

Our world now seems filled with anxiety, uncertainty, and the threat of a devastating diagnosis. To cope with this new COVID-19 world, you can apply techniques you've learned from coping with a bleeding disorder diagnosis. A bleeding disorder, and a global pandemic, can become less stressful when you have greater self-awareness and a positive mindset; when you set goals, create strong boundaries, and improve communication with your family and community. This issue of PEN is dedicated to helping you improve your mental health during the pandemic. We may be facing a new world, but we will get through it, together.

Laurie Kelley

In the past, it was incorrectly believed that only men could have hemophilia, and women with the gene were labeled asymptomatic "carriers." It's now recognized that women are not just carriers of hemophilia, but can also have hemophilia and experience symptoms if less than 50% of their factor is active. Most diagnosed patients are male. For editorial simplicity in PEN articles, when we refer to a person with hemophilia, we may alternately use "he," "she," or "they."

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EDITOR-IN-CHIEF Laureen A. Kelley

SENIOR EDITOR Sara P. Evangelos • SCIENCE EDITOR Paul Clement

CONTRIBUTING WRITERS

Richard J. Atwood • Cazandra Campos-MacDonald

Debbie de la Riva • Derek Markley

LAYOUT DESIGNER Tracy Brody • PUBLICATIONS MANAGER Jessica O'Donnell

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37-39 West Main Street #8
Georgetown MA 01833 USA
978-352-7657

info@kelleycom.com • www.kelleycom.com

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as i see it

Derek Markley

Being Better at Life

I often wonder how our son Bubba can remain unperturbed in situations that would drive me to the brink of insanity. After navigating life for nine years with severe hemophilia B, Bubba's greatest source of recurring stress is losing games of Fortnite or FIFA on his PS4. His ability to stay calm in virtually every part of his life is a mystery to me. It's an understatement to say that I envy this part of his personality.

How does he do it? A recently released study from Italy¹ suggests that people with hemophilia may be more effective at dealing with life's challenges and hardships than people who do not have a chronic condition. The study included 84 participants with severe hemophilia and 164 who had no history of a chronic illness. The research team found that the percentage of individuals "flourishing" with respect to mental health, as measured by the survey instrument, was higher for the participants with hemophilia than the participants with no chronic illness.

Bubba is the only member of our family who knows what it's like to have severe hemophilia, and he has developed his own strategies for handling the challenges that come with it. While our son was not a part of the Italian study, elements of his personality lead me to believe that he will flourish, too. I think he will teach us a few things along the way. In fact, I'd like to think that we as parents have also become more understanding, and more open to increasing our knowledge of the challenges people face daily.

I was perfectly happy believing this to be true, and felt no need to have my belief tested.

But life doesn't work that way...

Last summer my wife Ashley spent 11 days in the hospital, with five of those days in ICU. She suffered from severe acute necrotizing pancreatitis. Her condition was incredibly serious, and I had an uncomfortable discussion with a physician about



Derek Markley and family

mortality rates. We were very fortunate that Ashley made it through despite significant damage to her pancreas. Her physician warned us that her recovery could take up to a year, and that we shouldn't expect things to be "normal" in the near future.

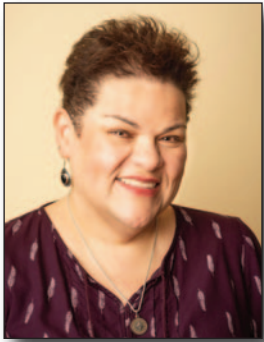
In December 2019, Ashley began experiencing significant weight loss, nausea, loss of balance, and a number of other issues. Labs during a trip to our nurse practitioner indicated that Ashley's glucose level was over 400. A person's glucose level should be under 140 two hours after eating. We were immediately sent to the local ER, where we learned that Ashley was suffering from diabetic ketoacidosis. At age 39, my wife was diagnosed with type 1 diabetes. After three days in ICU and another three days in recovery, Ashley returned home as a diabetic for the first time. She will be insulin dependent for the rest of her life.

During Ashley's time in recovery, Bubba watched her check her sugar levels and inject insulin. His response was understated, as usual: "At least I'm not the only one who gets stuck with needles now."

Everyone had a quick laugh, including the nurse. It was

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1. Luca Negri, Andrea Buzzi, Anna Brigida Aru, et al., "Perceived Well-being and Mental Health in Haemophilia," *Psychology, Health & Medicine*, Jan. 26, 2020.



Cazandra Campos-MacDonald

Depression, PTSD, and Inhibitors: My Family's Experience

Everyone processes stressful events differently. Moving, starting new jobs, changes in relationship status, and financial hardships are among the most stressful events that people experience. Living with a bleeding disorder and an inhibitor is also a reality that can be extremely stressful. We wait to reach a certain Bethesda Unit (BU) to begin immune tolerance, access a port daily, and deal with bleeds that take a long time to heal. How we handle these problems can affect our outlook on life and can raise other concerns.

Diagnosed with an inhibitor at age 11 months, my youngest son Caeleb is now 14 years old. Many complications, including target joints and an allergy to factor VIII, resulted in a year when Caeleb spent more days in the hospital than at home. It was an incredibly stressful time for our family. As a second

grader, Caeleb didn't easily understand why hemophilia was so difficult and painful. As his mother, I needed to remain calm to advocate for my son.

As a woman with clinical depression¹ and anxiety, I work hard to manage my mental health issues. During Caeleb's journey with an inhibitor, I found that writing was the best way to deal with my feelings. I blogged, posted on social media, and wrote articles that expressed my fears and anxieties. It was my process. My husband Joe is quite the opposite. When a problem arises, such as a terrible bleed that Caeleb endured, Joe gets incredibly quiet. He says he is a master "builder of walls," not allowing emotions to get the best of him. Joe and I work together exceptionally well as a team, but it wasn't until a few years ago that I discovered something remarkably interesting about my husband.

While I wrote my book about my experiences raising two sons with hemophilia and inhibitors, in the early stages of writing, Joe was my first editor. It often took some time for him to complete a chapter because reading and reliving the experiences we had endured brought up emotions that Joe hadn't processed. He realized then that the situational depression² he felt—which he had attributed to work issues and life in general—was partly caused by Caeleb's active struggles. Many years later, the lingering effects of these traumatic experiences continue to take a toll on our family.

What came next was the reality that post-traumatic stress disorder (PTSD) was now a part of our family. PTSD is a mental health condition triggered by a terrifying event. Our journey into the frightening part of living with an inhibitor began with Caeleb's repeated hospitalizations; with a team of nurses holding him down to access his port while he fought. Not only did the trauma of these hospitalizations and being

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1. Clinical depression is a term often used to refer to one or more types of serious depressive disorders that may occur with or without the presence of a specific stressor (nami.org). 2. Situational stress can generate emotional or behavioral symptoms that look and feel very much like clinical depression (nami.org).





Richard J. Atwood

Publishing for Hemophilia: Dr. Elihu Hubbard Smith

Are you a writer with a creative urge to put your thoughts in print? Or are you a reader, always eager to soak up information?

Reading and writing are separate forms of communication, bonded by the power of words. The publication of those words, either in print or electronically, is an important way for the bleeding disorder community to share and spread vital information.

A significant contributor to early American publishing in hemophilia was Dr. Elihu Hubbard Smith (1771–1798), better known for his literary pieces than for his medical articles. Unfortunately, Smith's publications about hemophilia, from 200 years ago, are virtually unknown.

Smith grew up in a medical family in Litchfield, Connecticut. His father, Dr. Reuben Smith, and his cousin, Dr. Josiah Root Jr., practiced medicine there. Smith passed the entrance exam for Yale College at the young age of 11, then earned his bachelor's degree in 1786. At only 15, Smith was too young to enter a profession. Instead, he spent two years at Greenfield Hill Academy for a literary and classical education. He wrote poetry, including five published sonnets. Smith next studied medicine by apprenticing with his father for two years, while also submitting anonymous poems and essays to the Litchfield newspaper.

Smith moved to Philadelphia in 1790, to attend medical lectures led by Dr. Benjamin Rush from November to February. One of Rush's lectures was titled "Nature of Blood." Completing these courses, and with his apprenticeship, Smith could call himself a doctor, but he could not use the MD initials after his name. Plus, he was too young to earn his medical degree.¹ Smith befriended Charles Brockden Brown, considered the

father of the American novel. With his literary colleagues, and using pseudonyms, Smith wrote a series of newspaper essays in the fall of 1791.

Smith then moved to Wethersfield, Connecticut, to practice medicine. He maintained a reciprocal correspondence with Rush.

Smith joined other literary men, mostly Yale graduates, in the Friendly Club, whose

members were known as the Hartford Wits. Smith wrote satire pieces that were published in the local newspaper, then later collected in a series to be published in *The Echo* in 1805. He also published the first anthology of American poetry, *American Poems* (1793). Seeking a better medical practice, Smith moved to New York City in 1793.

After passing a qualifying oral exam to practice medicine in New York when he was only 22, Smith took over another physician's private practice. Besides treating private patients, Smith was elected to the City Dispensary in 1794, and then to a staff position at New York Hospital in 1796, where he treated charity patients and taught medical students. With other literary men, Smith participated in weekly meetings of the Evening Club, which later changed its name to the Friendly Club. He wrote the opera *Edwin and Angelina*, performed in 1798. Smith joined the New



Dr. Elihu Hubbard Smith

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1. Smith only attended the lectures. Yet with his apprenticeship, he could call himself a doctor and practice medicine. But this put him in a lower class of physician compared to someone earning a doctorate of medicine (MD), who attended more lectures, did clinical work, and published a dissertation. This tiered system of medical practice lasted for over 100 years. By 1800, only three US medical schools had graduating students, so opportunities to earn an MD were limited. The minimum age to earn an MD was 24. The top tier of physicians usually attended medical school in Europe.



Mental Health During Coronavirus

and YOU

Debbie de la Riva

The collective pursuit to control the spread of coronavirus has resulted in an enormous challenge for the bleeding disorder community. The economic fallout of sheltering in place has affected our need for a steady income, health insurance, access to medical treatment, and—equally important—our access to each other. The degree of impact on our families is hard to determine, but it's safe to say this pandemic has been very stressful.

But stress is not a new concept for the bleeding disorder community. In fact, our community has been dealing for years with the emotional angst of fighting for what is needed to manage our medical conditions. Remember our fight for safer products, or our fight for laws to protect us from job discrimination? Today's battle, for our community, is to deal with the stress resulting from the pandemic. So let's follow the same steps we have taken so many times before.

Let's get informed, find our resources, and stick together.

Get Informed

To learn to manage stress, we need to become familiar with how our central nervous system works. Our brain comes pre-wired with an intricate system that functions to keep the rest of our body alive. This is the *limbic system*, which provides the “fight-or-flight” response. If the brain determines that the body is in danger, it initiates a chemical chain reaction that gets the body ready to either fight the challenge or run from it. This response begins when sensory information is picked up by a part of the brain called the *amygdala*. If the amygdala determines there is a threat, it signals other parts of the brain and body to release hormones such as adrenaline and cortisol. These hormones instruct the heart and lungs to increase their output in order to create the energy needed to meet the challenge. This fight-or-flight response is extremely effective when a person needs physical energy to avoid a danger such as jumping out of the way of a car. But most of today's challenges are emotional, and they don't require the extra energy provided by the stress response. The result is a steady supply of stress hormones circulating in the body at all times. The image that comes to mind is a person standing next to an IV pole and steadily receiving drips of adrenaline and cortisol. In other words, our body remains in a constant state of high alert.

The good news: We do have the ability to slow down the stress response. Since we now know that the brain is constantly scanning our body and our environment to determine if it should go into stress or relaxed mode, we can intentionally offer cues to indicate that we're not in danger. In fact, this is how meditation works. The first goal of meditation is to slow down your breathing rate. This is important, because once your brain receives the signal that your breathing rate is lowered, it will interpret this to mean that you're not in danger, and will turn off the stress response. The second goal of meditation involves your focus. You want to be focusing on the present—instead of musing about the past or anticipating the future—



COVID-19 is something no one wants and no one predicted. But *feeling* powerless shouldn't leave you without power. The difference between powerlessness and empowerment lies in your ability to accept a diagnosis—or a situation like COVID-19—and to move forward proactively, with new awareness and new choices.

One major difference in stress level between these two stressors is time: the diagnosis of hemophilia may have given you some time to learn to accept it. Bleeds usually don't happen right away. But COVID-19 hit us like a sledgehammer, and within weeks, we had to accept its presence and its consequences. Just as it was up to you to integrate hemophilia into your life and grow stronger with your new normal, we all need to integrate COVID-19 into our lives until a vaccine is found.

Becoming Empowered

Being “empowered” means that you believe you can influence your own destiny through the decisions you make. It means that you realize you are accountable for your responses to situations and challenges. These responses include your thoughts, feelings, beliefs, and actions.

Although being accountable is the key to becoming empowered, being so responsible can make you feel anxious. For a new parent of a child with hemophilia, responsibility means making decisions and taking action. You must read about medical procedures, choose a factor concentrate brand, stick your baby with a needle, watch for symptoms of a bleed, and choose a treatment option. As a US citizen, you are responsible for taking proper precautions to avoid contracting COVID-19 or passing it on to others. You are responsible for knowing about and following your state's health guidelines and social distancing rules.

One way to conquer fear, lower anxiety, and accept responsibility is to observe how you problem-solve. Do you spend a lot of time worrying or obsessing about the challenge you face? Does COVID-19 keep you up at night worrying? If so, you're probably focusing on what *is* happening—which is truly frightening—or what *will* happen, instead of focusing on what you can *make* happen.

Instead of dwelling on a problem, try to spend more time on a solution. Try to spend 90% of your time on a solution, and only 10% on the problem. This means you need to respond with action. Your child has hemophilia: How can you learn more about it? Have you scheduled an appointment with your hemophilia treatment center (HTC)? Have you contacted National Hemophilia Foundation (NHF) or Hemophilia Federation of America (HFA)?² Have you subscribed to hemophilia magazines or ordered books? Have you checked out pertinent



websites or searched for hemophilia groups on Facebook?

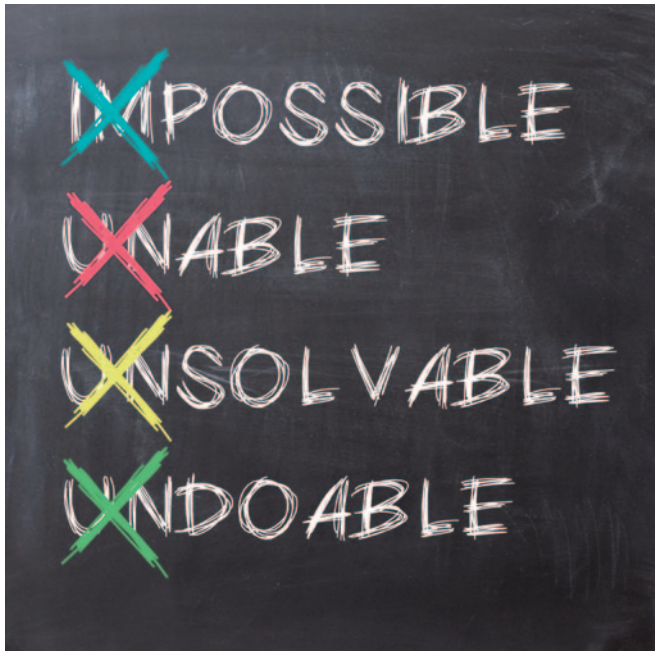
With COVID-19, you can read about coronaviruses. Many books are available, written in clear, simple terms. Google COVID-19. But don't just listen to the nightly news and the increasingly polarized debates. Dig into some science, learn the facts, and stay open-minded. Wear a mask according to your state's mandates, or as often as you feel you need to. Reevaluate your lifestyle and health: Do you need to lose weight? Being obese is associated with a higher risk of developing severe symptoms and complications of COVID-19, and more than 42% of the US population is obese. Smoking is also a risk factor. Quit smoking—*now*. Start walking, running, cycling. Set goals for better health and healthier living; goal-setting is a fabulous way to be empowered. And goals for exercise and eating better will lower your stress amazingly, throughout the day. Goals give you the feeling of empowerment, and indeed, goals empower you.



GOAL-SETTING MADE SIMPLE

- Set manageable, measurable goals to create small victories.
- Add a deadline—this will motivate you to act.
- Share your goals with family or co-workers—this will keep you accountable.
- Tape your goals to the refrigerator.

2. NHF: hemophilia.org. HFA: hemophiliafed.org.



Identify and Channel Your Feelings

You may experience a range of emotions and reactions when you receive the diagnosis of a bleeding disorder like hemophilia. Just as our nation did, when we went from shock to disbelief to caution, and then to full-scale alert when COVID-19 was confirmed.

What may surprise you is how differently people respond when stress hits. Family members may react in various ways to a hemophilia diagnosis. For example, one parent may close off and withdraw, trying to appear stoic. Another parent may be more emotionally expressive, talking or crying. A grandparent may deny the baby's diagnosis, while an uncle may become the "expert" on hemophilia. Similarly, with COVID-19, your Facebook friends may shock you with their opinions about CDC recommendations, state safety mandates, or what the White House should do to overcome this pandemic.

Though it's natural for anyone to experience intense emotions under stress, you can be disempowered by feelings when (1) they affect others negatively, or (2) you allow others' emotions to affect *you* negatively. Intense feelings can interfere with your personal relationships, your personal growth, and your parenting.

Feel more empowered by choosing how to respond to your feelings. Sometimes you can't control what you feel, but you can train yourself in how you act and what you say.

First, identify your feelings. Look inward, and try to find the true motive or core emotion behind your actions. It may seem simple: A person who yells is angry, right? But after looking deep within, the angry person may find that the core feeling is really fear. When you identify your core feelings, you'll find it easier to choose your responses.

Second, pay attention to your physical reactions. They can also offer clues to what you're truly feeling. For example, you're preparing to go to the HTC. Outwardly, you may appear calm, but your heart is racing and your palms are sweating. If you realize, "I must be feeling anxious," then you can decide how to take action to reduce your anxiety. Believe it or not, just becoming aware of your feelings can help you take deeper breaths and relax.

Third, look for a way to channel intense feelings into something positive. Intense negative emotions are often a cue to turn off the TV, read a favorite book, take the dog for a walk, or take a drive. Try a hot bath, gardening, or practicing yoga. You may feel angry because you've been furloughed from work or because hemophilia has caused mounting medical bills; but anger without action can make you feel depressed. If you acknowledge your anger, you can try channeling it into healthy activity, like calling a trusted friend to vent, exercising, playing with your dog, or engaging in a favorite hobby.

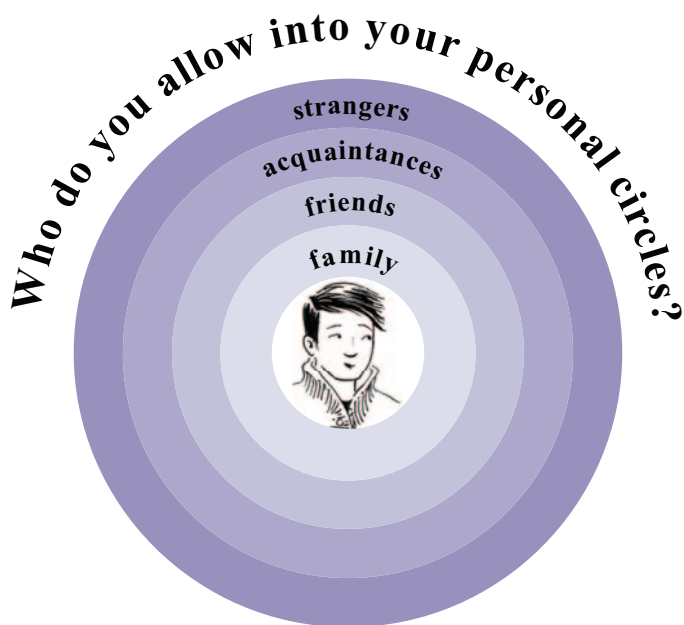


KEYS TO IDENTIFYING AND HANDLING INTENSE FEELINGS

1. Look behind the immediate feeling to the core emotion.
2. Check your physical response for clues.
3. Name your feelings.
4. Channel your feelings into healthy responses.

Maintain Boundaries

One of the most important things you can do to feel empowered is to strengthen your personal boundaries. Boundaries are the emotional, mental, and physical barriers we set for ourselves, barriers that define how comfortable we feel in relationships. It's like our skin: it's there to protect us, allowing certain things in (sun, cream, medicine, moisture), and keeping out harmful things (bacteria, viruses, poison). Emotional boundaries define



which behaviors and words from others will affect us—what we will and will not allow. With healthy boundaries, we ask to be treated respectfully, and we offer respect to others.

Think of boundaries as concentric circles drawn around you, like a series of fences guarding the perimeter of your emotional, mental, and physical being. As you begin to trust people, you let them into your circles. When someone you don't know—or don't trust yet—gets too close, you feel your boundaries being violated. Have you ever had a stranger stand much too close to you? You go on alert, the hair stands up on your neck, and you instinctively pull your arms tighter around you. Someone has penetrated your inner circle, and you feel stunned, surprised, defensive—even angry.

Being aware of your boundaries is important in hemophilia, because you may need to reach beyond them to trust new people, like your HTC staff. And you need to learn how to handle people who may push through your personal boundaries because they're curious about your child's hemophilia. The correlation to COVID-19 is obvious: physical boundaries are essential, down to how many feet we may approach people, and what seating in restaurants will eventually look like. Boundaries keep us all safe.

We experience conflict with people when our ideas about boundaries clash. Some people have rigid boundaries, and some have none at all. Do you know people who never open up and share what's happening? They may have rigid boundaries. Do you know people who share their entire life stories, with every intimate detail, at your first meeting? They seem to be boundary-less. *You* are responsible for clearly defining your boundaries, for

respecting others' boundaries, and for reestablishing your boundaries when others have violated them.

Our country has been clashing about boundaries since COVID-19 appeared. We ask, "Whose right is it to determine physical boundaries due to COVID-19?" "How do we respect others' boundaries?" Social distancing and stay-at-home policies to contain the spread of the virus have led to entire states shutting down, even entire countries. People who are angry over COVID-19 boundaries—keeping them, or relaxing them—vent on social media, at the White House, and at each other. Some people in the hemophilia community are no longer on speaking terms on social media because of clashes over what constitutes safe pandemic boundaries.


To lower anxiety related to clashes over boundaries, you can decide which comments, actions, and even attitudes you will and won't accept. For example, you don't have to share information about your child's hemophilia with anyone. If someone asks, "What's that bump on his chest?" and points to your child's port, you decide: Do you want to smile and pass it off? "Oh, it's just a bump!" Or do you want to invest the time to educate someone? It's your choice.

You can also protect your boundaries by shifting focus. Instead of directly answering someone's question, you can ignore it, or redirect it: "I prefer to concentrate on all the things that are right with my baby." You can use this change of focus as an opportunity to educate someone: "Because you're curious about his bump, let me tell you about hemophilia."



TO MAINTAIN HEALTHY BOUNDARIES

1. Identify your feelings immediately after an exchange with someone.
2. Know who is most likely to violate your boundaries.
3. Understand which actions and questions trigger your deep feelings.
4. State your boundaries clearly to the boundary breaker, or shift focus.



When dealing with COVID-19 boundaries, try to be aware and understanding. Emotions are running high. Many people say things on social media that they wouldn't dare to say face-to-face. From "I'm going to the beach and not wearing a mask! I'll do what I like!" to "We all must stay inside and away from each other," people all have opinions, and most will not change regardless of how much you argue. If you do respond on social media to rants or even just opinions that don't agree with yours, ask: Am I responding thoughtfully, with compassion and understanding, or am I overreacting? Is the comment I just read really violating my boundaries? Am I acting like a bull with a red flag in front of me? Remember, most people on social media are looking for a reaction. You can choose to do what they want—react—or you can leave the conversation, and then practice lowering your anxiety, using the examples provided earlier.

If you rage silently when others break your boundaries, then the situation becomes your problem. If you respond to others respectfully and clearly—clarifying what you will tolerate and what you won't—they may be surprised or offended, but that's their problem. Boundaries protect us, but they don't necessarily separate us from others. Boundaries keep us in healthy relationships, physically and emotionally, with

the people we love, and with our neighbors, friends, and bleeding disorder community members. Know your personal boundaries, and protect them. And know your state's boundaries regarding COVID-19.

The Internet: Double-Edged Sword

The internet is a common resource for information on hemophilia, and on COVID-19. Anyone can google "hemophilia" and instantly find professional websites with information on symptoms, coping with the diagnosis, treatment options, HTC listings, financial aid, and more. Within minutes of the diagnosis, new parents can connect with hundreds of other parents of children with hemophilia to find comfort, advice, ideas, resources, and new friends. YouTube has videos of infusions, summer camp, and parents and patients sharing their stories. Instagram has thousands of photos of children and families with hemophilia.

Information on COVID-19 is available and still emerging, and reports can be conflicting. Add to this the politicizing of the pandemic, and the internet becomes a double-edged sword: a source of both information and misinformation—and anxiety. Who to believe? Who to trust?





There are three ways that the internet can actually rob you of empowerment, whether about hemophilia or COVID-19. If you're aware of these risks, you can use the internet to your advantage.

First, you may get overwhelmed. If you search hemophilia, you'll find online discussions of dosing schedules, statistics, bleed symptoms, new drugs. And you may come across grisly photos of bleeds; some are superficial, but to a new parent, they will look shocking. With COVID-19, discussion about how to safely distance, when to open up states—and even your constitutional rights—can turn ugly and political.

Second, you may get misinformation. With hemophilia, other parents want to share what they know, but unless they are registered healthcare professionals, you won't know which information is accurate. Always question what someone says about treatment, and question their credentials. The same goes for COVID-19, maybe even more so, because there is still so much we don't know. As states grapple with when and how to open and whether we should wear masks, you may find yourself swayed by opinions from people who don't have all the answers, or who post incorrect information. Ask questions. Identify your emotions so you can recognize any personal bias first. Then *think*, and respond carefully and logically. Don't believe everything that's posted; always check sources. Does a posted article, statistic, or quotation get a rise out of you? Find out if the information came from a reputable website or organization.

Third, you may spend so much time on the internet that you neglect your family—and yourself. Your family needs you now. And you need to care for yourself. Unfortunately, being online is psychologically addictive, especially in this polarized political

climate. Friends, colleagues, and even family get into shouting matches on Facebook about government policies and science. It's not helpful, and it only raises anxiety. Limit the amount of information you seek at one time, and limit the number of hours you spend online. Balance your online time with “me” time and family time.

Turn to your HTC first for answers to your medical and childcare questions. Turn to multiple and reputable sources for information about COVID-19. If you find your strong emotions—fear, anxiety, frustration, anger—are affected by people on Facebook, the news, and social media, then limit your time with these. Follow the advice presented earlier: identify your feelings, channel them in a healthy way, set goals, and strengthen yourself internally. Maintaining emotional, thoughtful balance is vital to staying empowered, and key to limiting anxiety.

Cultivate Inner Strength

Remember that becoming empowered means knowing you are accountable for your responses to situations and challenges. To respond, you need to know what you believe, what you stand for, what your values are. This means you have a belief system for interpreting the world. When you receive a hemophilia diagnosis, or live in a world with emerging viruses, stress can challenge your beliefs. You may turn to your beliefs for comfort, or you may abandon them altogether!

Try using the challenge of stress to understand yourself better. What *do* you believe? Major crises in life are often interpreted as a path to deeper spiritual growth. Your spiritual beliefs and practices can offer strength to help you cope with problems in life.

Spirituality encompasses many practices, inside and outside organized religion. Meditation allows you to connect with yourself quietly and peacefully, producing great health benefits. Nature gives solace and support. Music and poetry provide comfort and inspiration. Mother Teresa once said that humanitarian service to people in need is the greatest form of worship we can offer. Spirituality often recognizes a higher power, and

BELIEVE
IN YOURSELF

seeks to expand our consciousness beyond our routine lives.

Our belief systems can also help place our crises and difficulties in a wider context, bringing a broader perspective and with it, perhaps, relief. In his book *The Problem of Pain*, the Christian author C. S. Lewis wrote that pain is God's "megaphone to rouse a deaf world."³ Yet the first of the Noble Truths in Buddhism is that suffering exists inherently in life. Whether or not you follow a particular religious belief, you may start asking new questions about life when you suddenly face a crisis—especially one that involves your child's health, and our nation's health.

Your Empowerment Legacy

The diagnosis of hemophilia, like the emergence of COVID-19, may leave you feeling overwhelmed by an array of emotions. You may feel stripped of your ability to care responsibly for your child with hemophilia. You may fear this new world where viruses can appear seemingly out of nowhere. But when you begin to accept what you cannot change; when you gain awareness of how you solve problems and how you respond to life's challenges; when you understand how you can change within...then you will become empowered.

The nation, the political parties, the news anchors, the people around you—even your own family—may have negative attitudes and incorrect information. Your situation may be tough right now, while you balance homeschooling, working from home, inability to visit family, and even job loss. But you've begun to see that you can focus on positive solutions, starting with your ability to manage your emotions and responses. Your inner strength, resourcefulness, and efforts to create a positive environment for yourself and your child will guide others to do the same. Be an example of empowerment for your child, who

will one day accept ownership and responsibility for his own life and his own disorder. Be an example of empowerment for our country, which desperately needs to heal.

It's your life, your child, and your destiny. Don't stay stuck in fear and anxiety. Start living your best life today, no matter if it's a diagnosis of hemophilia, or a new world with COVID-19. ☺

Adapted from *Empower Yourself About Hemophilia* by Laureen A. Kelley.
Available at www.kelleycom.com or amazon.com in print or as an ebook.



3. C. S. Lewis, *The Problem of Pain* (San Francisco: HarperSanFrancisco, 2001 [1940]), 91.



PROJECT
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PROJECT SHARE
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Do you have factor to donate?

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— NONPROFIT —

COVID-19 Relief Is Here

Hemophilia Federation of America (HFA) is providing financial relief for members of the bleeding disorder community facing a significant loss of income due to COVID-19. The fund will provide emergency financial assistance for payment toward one essential household bill, such as mortgage, rent, water, electricity, phone, or car payment. **Why this matters:** If your application is approved, funds will be paid directly toward a bill (payment to landlord, for example) to give quick financial relief.

To apply: hemophiliafed.org



Find an HTC Near You

The US Centers for Disease Control (CDC) has a searchable database of hemophilia treatment centers (HTCs) throughout the country. Use the HTC directory to search for names and contact info for HTCs and staff that are part of the federally funded HTC network; create and save lists; print mailing labels; send emails. **Why this matters:** When patients travel or move, the listing will help them quickly find a needed HTC.

For info: CDC.gov and search: HTC

New Hemophilia Numbers

De-identified data collected 2012–2018 from US HTCs was used to estimate incidence and prevalence of hemophilia in the US (last done in 1998). Based on this latest data, the average hemophilia incidence in the US is 1 case per 4,334 live male births (higher than the previous estimate, 1 in 5,032). Researchers estimate that there are between 29,761 and 32,985 males with hemophilia in the US today (higher than the previous 20,000). Unfortunately, women with hemophilia weren't counted in this data. **Why this matters:** New numbers indicate that males with hemophilia are one-third more common than we thought, which may affect our healthcare system and pharmaceutical companies.

For info: onlinelibrary.wiley.com and search: Occurrence rates of haemophilia among males in the United States



The Problem of Pain

Pain Podcast is a series of audio discussions about living with chronic pain. It includes info from pain specialists on the frontline. Topics include treatment using opioids, cannabis, chiropractic, surgery, psychodynamic therapy. New episodes are released on Wednesdays. Produced by BloodStream Media.

Why this matters: Pain is often overlooked as a serious issue in patients with bleeding disorders; it can lead to increased suffering or drug addiction.

For info: bloodstreammedia.com



— SCIENCE —

Rabbits That Make Factor

The US Federal Drug Administration (FDA) approved Sevenfact® (coagulation factor VIIa [recombinant]-jncw) by HEMA Biologics. This new bypassing agent treats and controls bleeding episodes in adults and adolescents 12 or older with hemophilia A or B with inhibitors. Manufactured by LFB, Sevenfact is an innovative recombinant form of human factor VIIa. Its active ingredient is expressed in the milk of genetically engineered “transgenic” rabbits that contain the gene for human factor VIIa.

Why this matters: Transgenic animals can produce large amounts of factor in their milk; production can be scaled up by milking more animals. This will increase availability of factor VIIa and perhaps decrease cost.

For info: groupe-lfb.com



— INDUSTRY —

Takeda Increases Assistance During Pandemic

As the COVID-19 crisis continues, Takeda has enhanced its assistance programs for US patients who qualify and need extra help. Highlights: (1) Job Loss Provision helps eligible patients who have lost jobs and have financial hardship. (2) Eligibility period will be extended for certain patients currently enrolled in Takeda's patient assistance programs. **Why this matters:** Extended eligibility periods can help reduce the time patients spend on securing medication approvals, so they can focus on health and safety during the pandemic.

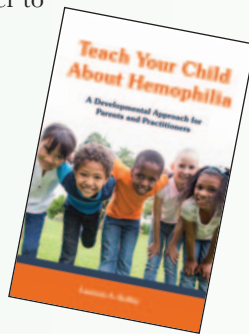
For info: 800-830-9159, 8:00 am – 8:30 pm ET, Monday – Friday

— LA KELLEY COMMUNICATIONS —

Newly Revised!

Teach Your Child About Hemophilia by Laureen A. Kelley is an in-depth exploration of the way children think and understand hemophilia as they mature from preschooler to adolescent. The book examines children's understanding of hemophilia concepts at different ages: cuts, healing, blood, severity levels, blood clotting, infusions, genetic transmission. *Teach Your Child* prepares parents to appropriately answer children's questions and encourage independence.

To order: kelleycom.com



— ADVOCACY —

No More Copay Assistance

The US Department of Health and Human Services (HHS) finalized the “2021 Notice of Benefits and Payment Parameters” rule, allowing health insurers to continue using accumulator adjusters. This means that health insurers can exclude manufacturer copay assistance amounts when calculating a patient's overall deductible or out-of-pocket maximum. **Why this matters:** Excluding factor manufacturer copay assistance amounts can mean higher out-of-pocket costs and a serious barrier to access to care for some.

For info: hemophiliafed.org and search: HHS Finalizes Rule Allowing Insurers to Exclude Copay Assistance



— SOUNDBITES —

- **COVID-19** will likely be with us for the next few years, until a vaccine is developed and widely used. **NHF** has developed an info resource for people with hemophilia: hemophilia.org/Newsroom/COVID-19-Information
- BioMarin Pharmaceutical announced that despite the COVID-19 pandemic, its application to the US FDA remains on track, seeking approval of valoctocogene roxaparvec, an experimental **gene therapy** for adults with severe hemophilia A.
- **Novo Nordisk** announced that three clinical studies evaluating concizumab (investigational anti-tissue factor pathway inhibitor for treating hemophilia A and B) have been paused, following nonfatal thrombotic events in three patients enrolled in ongoing phase III trials.
- **Bayer** and **Ultragenyx Pharmaceuticals** reported results of a gene therapy trial that increased levels of factor VIII and prevented or reduced bleeding in the first two subjects with severe hemophilia A treated in a phase I/II clinical trial. The trial used a modified adeno-associated virus (AAV, called AAVhu37) to deliver a functional B-domain-deleted copy of the factor VIII gene to liver cells.
- US government and blood industry leaders are urgently appealing for **voluntary blood donations** to avoid a severe blood shortage. As of March 19, tens of thousands of blood drives have been canceled, resulting in the loss of several hundred thousand blood donations.
- The worldwide therapeutic plasma proteins market reached **\$24 billion** in 2018, up 13.3% from 2016, or 6.6% annually, according to a new report by the Marketing Research Bureau, “The Worldwide Plasma Proteins Market–2018.”
- Genentech has reminded healthcare professionals that if hemophilia A patients on **Hemlibra**® develop severe COVID-19 infections and clotting problems similar to disseminated intravascular coagulation (DIC), Hemlibra can interfere with some blood tests that diagnose DIC. Alternative tests should be used.
- Pfizer Inc. and BioNTech SE announced that the first US participants have been dosed in the phase I/II clinical trial for the BNT162 vaccine program to prevent **COVID-19**.

good timing on Bubba's part.

A week later, Ashley and I met with a diabetes educator. At one point, the educator asked me if I would be able to give Ashley an emergency injection of glucagon if necessary.

My wife and I both smiled and laughed. Our diabetes educator was clearly confused. I explained that we had a son with severe hemophilia and that we did weekly infusions at home. The idea of mixing medicine and a subcutaneous injection was not very intimidating.

That was the moment when I realized how much we'd grown as a family. In the midst of learning about Ashley's lifetime of treatment, we could both find the humor in a question about our ability to give an injection. Neither of us had spent the past nine years as a person with hemophilia, but living with Bubba had taught us a lot. Part of our learning curve included ports, infusions, sterility, and the physical treatment of issues related to his bleeding disorder. That was only part of our growth and development.

Being Bubba's parents taught us how to deal with our emotions when health-related issues occurred. Bleeds can be scary. Surgeries cause significant stress. Raising a child with severe hemophilia meant that we experienced high levels of emotion and stress on multiple occasions. We learned how to focus on

the well-being of our son, not on all the other things surrounding his condition. We grew in our ability to more fully understand the impact of a medical condition on individuals and their immediate family members. Hospital trips, needles, and monitoring a loved one's condition were simply parts of our normal life. Most people would not characterize any of this as normal, but it's what we do daily as a hemophilia family. We have learned how to persist and work through challenges.

I do not have hemophilia, nor do I have diabetes. I can say, with great assurance, that hemophilia taught our family how to flourish when faced with a significant challenge. Without Bubba's presence in our life, I'm certain that a diabetes diagnosis would have been far more difficult. We're a little tougher because of what we've learned as Bubba's parents. I know that Ashley is more confident in her ability to move forward managing her diabetes because of our experiences. I don't have a study to prove it, but I'm pretty sure we get a little better each day. ☺

Derek lives in Saltillo, Mississippi, with his wife Ashley and their children Abbey and Bubba. He is the executive director of two University of Mississippi regional campuses and an assistant professor in the School of Education. Ashley is a fourth-grade math teacher in the Tupelo Public School District. Derek is the author of The Bubba Factor, available on Amazon in Kindle format and in paperback.

Inhibitor Insights... from page 4

held down for infusions affect Caeleb, but PTSD is also evident in my life, and in Joe's.

Once the frequent hospitalizations ended, our family experienced fantastic freedom. However, the times we went to the hospital for regular clinic visits and blood draws triggered our PTSD. For us, the smells and sounds of the hospital are unlike those of any other places we visit, so walking into the building immediately raises our defenses. When Caeleb goes to the clinic for a blood draw, it takes him extra time to prepare himself mentally for the stick. He handles needles well, but the immediate physical response he can't help is *fear*. PTSD is an issue that affects our family, and memories of the traumatic experiences Caeleb endured come back in a flash.

Joe and I treat our depression with the help of medications prescribed under the careful supervision of a psychiatrist. Not all primary care physicians have a specialized understanding of the medications needed to treat depressive disorders; that's why Joe and I have a psychiatrist who can closely monitor our individual needs. The other component that helps us maintain good mental health is regular therapy. A therapist is a necessity on our path to good mental health. Often, people think that treatment is a sign of weakness. In my experience, I see therapy as a sign

of strength. Struggling with mental illness can be frightening, but when working with a therapist, you develop the tools you need to get through the times when depression and anxiety can take over your life.

When left untreated, mental health issues can be overwhelming and prevent you from living your best life. Hiding your problems, hoping they'll resolve on their own, is not in your best interest. It takes a great deal of strength to admit that you need help. We all struggle, but often we just learn over time how to hide the issues we feel make us "weak."

COVID-19 has brought even more stress and anxiety to our lives, and this can lead to depressive episodes. If you deal with mental health issues, please know that you are not alone! Many people have the same challenges; yet many fear that revealing their issues will stigmatize them. True freedom comes when you lower your defenses and share your struggles with loved ones. Maybe you know someone in the bleeding disorder community who has mental health issues—especially someone living with an inhibitor. Reach out. Talk to them. Ask questions. If you're struggling yourself, they may be able to help you find the resources you need to get on track and work toward making your mental health a priority. ☺

York Manumission Society, whose goal was to free slaves. He became a trustee of the society's school, then called the New York School for Colored People.

Earlier in 1794, Smith had written a letter to Rush summarizing the bleeding episodes of his cousin, James Hawley. An extract of this letter was published in an 1805 medical journal, after Smith died. Though Hawley had no known family history of bleeding, his hemorrhaging is considered a case of hemophilia, even if questionable.² After the 1794 yellow fever outbreak, Smith wrote ten letters to Dr. William Buel of Sheffield, Massachusetts, describing that disease. These letters, collected and published in 1796 by Noah Webster, demonstrated Smith's medical knowledge. In an 1817 medical journal, Buel and his brother Samuel, a physician as well, would publish a summary of the Collins family, also of Litchfield, that included multiple cases of hemophilia.

Smith continued to learn about blood by reading medical texts. He read William Hewson's *Inquiry into the Properties of the Blood* (1774) and John Hunter's two-volume work *Treatise on the Blood, Inflammation, and Gun-Shot Wounds* (1796).

Smith even proposed doing a chemical analysis of blood.

In 1797, Smith invited Rush to teach at Columbia College, only to have the offer vetoed by Alexander Hamilton for divisive political reasons.

Along with Samuel Mitchill, MD, and Edward Miller, MD, Smith is remembered as one of the founders and editors of *The Medical Repository*, the first American medical journal. Both Smith and Rush contributed articles, beginning with the first volume in 1797. This medical journal published the 1803 article by Dr. John Conrad Otto that is considered the first clear description of hemophilia ever published.

During the 1798 yellow fever outbreak, Smith remained in New York to practice medicine. He contracted the disease and died that September, at age 27. We can only imagine what he might have accomplished during a full lifespan.

Smith surrounded himself with other literary men, always with the intent to publish, whether it was for the arts or for medicine. Smith is better remembered by literary scholars who acknowledge him as an early American poet, playwright, satirist, editor, and publisher.³ Regrettably, medical scholars are less likely to recognize Smith's contributions to medical publishing, especially his connection to hemophilia. But Smith certainly wielded the power of the printed word. 🍷

2. James Cronin, who edited *The Diary of Elihu Hubbard Smith 1771-1798* (1973), claimed that in 1794, Smith successfully treated a patient with hemophilia in New York using common salt as a remedy. This claim can't be substantiated because Smith treated a case of hemoptysis, or bleeding from the mouth, with salt. Salivation was Smith's favorite cathartic treatment. He often applied mercury to the gums, a treatment that produced saliva and blackened teeth. Cronin may have misdiagnosed Smith's case by labeling it hemophilia; Smith himself diagnosed it as hemoptysis. But because, at that time, all bleeding was treated the same way—with purgatives—it wouldn't matter whether the patient had hemophilia or hemoptysis. 3. Smith didn't own a publishing company, but he provided the money to publish—for himself and his friends—because printers wanted payment up front. One way to print was to obtain subscribers before the printing, but someone had to solicit the subscribers. Smith was never wealthy as a physician because he used his income for publishing.

YOU... from page 6

and you want to intentionally focus on words or images that evoke feelings of peace or happiness.

You can bring up images of when you felt safe and happy, or you can think of words that reassure you. This tool is like anything else in life: it requires practice and commitment. But eventually, you'll find that you can truly create a sense of well-being, no matter what's going on in your life. Sound too good to be true? Do you need proof? Ask yourself how you feel when you're watching a scary movie, and compare that to how you feel when you're watching a romantic comedy. In other words, what we focus on creates how we feel inside. That same principle is at work when we intentionally think about what we are grateful for, as opposed to what we lack or what we don't like about our lives.

Learning to handle stress is more important than ever. Let's look at the many resources that have been created since the coronavirus pandemic started.

Find Resources

National Hemophilia Foundation (NHF) and Hemophilia Federation of America (HFA) have created content to help people in the bleeding disorder community deal with both physical and emotional impacts of the pandemic. The following links will take you to the current web pages with this information.

National Hemophilia Foundation
hemophilia.org/Newsroom/COVID-19-Information
Hemophilia Federation of America
hemophiliafed.org/news-stories/2020/04/coronavirus-covid-19-what-hfa-is-doing/

You can also purchase proven self-help workbooks on stress reduction at New Harbinger Publications: newharbinger.com

And yes, there is an app for learning to relax! Appropriately called the Calm app, it has hundreds of meditations and master classes on stress management: calm.com

You can find these resources and many others by visiting the Mental Health Matters Too website: mentalhealthmatterstoo.com

Seek Out Others

It didn't take long for our community to figure out how to be connected virtually. Though it isn't the same as being in a room together, it is nice to see familiar faces and get a chance to let someone know you are there for them.

Take a moment to check in with yourself, because it's very easy to feel lonely in isolation. If you find that you're exceptionally lonely, depressed, or anxious, it always helps to talk to someone trained to help you feel understood and supported. Online platforms like Talk Space and Better Help are reporting an exponential increase in the number of requests for counseling sessions right now.

Look for Purpose

One of the best ways to combat the feeling of helplessness that comes with a crisis is to look for a way you can help others. This sense of purpose gives people some control,

and helps them feel productive and useful. For me, contributing to Save One Life is one way I fulfill my need to have purpose in my own life. Each month, I have a small sense of satisfaction knowing that there are three young people with hemophilia who feel that someone else on this planet sees them and cares about them.

So, whether it's meditating, talking with someone, or just being there for another human, there are ways to combat stress. We will get through this pandemic as a community, the way we always have. We will get informed, find resources, and seek out each other. ☺

Debbie de la Riva, LPC, has been an active member of the bleeding disorder community since the birth of her son with severe hemophilia 25 years ago. She served as executive director of the Lone Star Chapter of NHF, was a co-chair of an NHF Annual Meeting, received a Ryan White Award for Advocacy Excellence, and has presented on mental health issues to chapter and national organizations. In 2018, Debbie founded Mental Health Matters Too as a way of combining her degree as a licensed professional counselor with her passion for helping community members who struggle with mental health challenges. To contact Debbie: www.mentalhealthmatterstoo.com or debbie@mhmttoo.com

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myPROBE App

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The myPROBE app allows respondents to complete the PROBE questionnaire on a smartphone or tablet.



What is PROBE?

PROBE stands for Patient Reported Outcomes Burdens and Experiences. PROBE is a long-term study aimed at measuring outcomes that people with hemophilia A and B think are important to their health status and quality of life. To gather data, people can complete this questionnaire via the myPROBE app.

Why use PROBE?

The PROBE study provides a way for people with hemophilia to report data on their health status and quality of life. Patient organizations can then use this data as evidence to advocate for better care and treatment.

Who can use PROBE?

The questionnaire can be completed by people with hemophilia A or B, including carriers. It can also be completed by people who don't have a bleeding disorder. They serve as the control group.



Get the App!

The app can be downloaded for FREE from the **Apple Store** for iOS and **Google Play** for Android and can be used on smartphones and tablets. Search for **myPROBE** in either the Apple Store or Google Play.



No device? No problem.

Complete the online survey at
myPROBE.org

Learn More

Periodic updates, scientific abstracts and publications from the study are available on the PROBE website: **www.probestudy.org**

inbox

“Meet Your Child’s Joint!” *PEN*, May 2020



I READ WITH INTEREST THE thorough review of joint bleeds in the latest edition of *PEN*. This article, as many of the *PEN* articles do, references current scientific research. Unfortunately, it mentions joint microbleeds, which is not supported by current scientific research. The concept of joint microbleeds in hemophilia patients was proposed as a hypothesis to explain unexpected imaging findings in the Joint Outcome Study. That study is over 15 years old, and since then, there has not been a single publication in the medical literature confirming the existence of joint microbleeds. Even though there is no evidence for joint microbleeds, this doesn’t mean they do not exist. However, I think you do a disservice to patients, leaving them with the impression that joint microbleeds are accepted and established by medical science when they are far from that.

JOHN PUETZ, MD

Adjunct Professor of Pediatrics
Saint Louis University School of Medicine
SSM Health Cardinal Glennon Children’s Hospital
St. Louis, Missouri

Response from Paul Clement, PEN Science Editor:

DR. PUETZ IS CORRECT THAT there is no direct evidence of joint microbleeds; but the *PEN* article does not say this or give the impression that microbleeds are established by medical science. The information in the article referring to microbleeds includes this: “Whether microbleeds are

responsible for joint damage over the long term is still unproven.”

Joint microbleeds, if they exist, are difficult to study—they are subclinical (no signs or symptoms) and involve microscopic amounts of blood. They are not detectible by standard x-rays. MRI, the best imaging method and gold standard for detecting joint disease, is time-consuming, expensive, often not accessible, and may require sedation in young children. The next best imaging method is musculoskeletal ultrasound. A clinical trial started in 2016, the Hemophilia Ultrasound Project (clinicaltrials.gov, NCT02807753), is scanning the joints of children with hemophilia every six months, monitoring the progression of joint disease. We may finally have an answer to the role of microbleeds in joint disease when the study is completed in July 2021.

I GOT MY COPY OF *PEN* with the joint article...an excellent piece it is! Very informative and extremely easy to understand. I highly recommend it to all with bleeding disorders and their families. You will learn a lot. By the way, you should all sign up for *PEN*. I’ve been getting it for several years. You can’t go wrong.

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