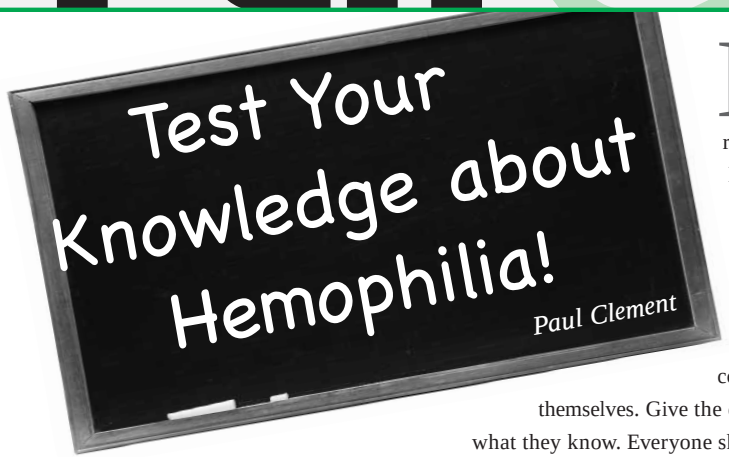
PEN

inside



It's almost back-to-school time for our kids! Soon enough, they'll be tested on what they've learned in the classroom. We've all learned the hard way in the real-life world of bleeding disorders. But when was the last time we were tested on hemophilia?

In this issue of PEN, we offer a great checkup on your knowledge of all things hemophilia. And it's multiple choice, so it's not too tough. You'll find the answers on pages 17–19, so you can quiz yourself and even learn something new. Download extra copies from our website and have family members test themselves. Give the quiz to audiences at your local chapter events, to see what they know. Everyone should know the answers to these basic hemophilia questions. Circle your answers, and then check your responses at the end. Good luck!

All about the Science

1 Hemophilia A is caused by a deficiency of which clotting factor?

a) factor VII
b) factor VIII
c) factor IX
d) factor III

2 Factor VIII is produced by the endothelium, the thin layer of cells that line the interior surface of blood vessels. Factor VIII is also produced in which organ?

a) kidney
b) thyroid
c) liver
d) bladder

3 About how many people in the US have hemophilia?

a) 5,000
b) 17,000
c) 50,000
d) 1 million

4 What's the average number of babies born with hemophilia in the US each year?

a) it depends
b) 400
c) no one really knows
d) 1,000

5 The half-life of factor VIII concentrate is closest to

a) 2 hours
b) 12 hours
c) 18 hours
d) what's the half-life?

6 Which of the following does *not* describe the complication of hemophilia known as inhibitors?

a) shyness in social situations
b) shortened factor half-life
c) affects about 20% of people with hemophilia A
d) more common in people of African American heritage

7 In 1964 Dr. Judith Graham Pool discovered the first effective treatment for bleeding episodes in people with hemophilia. What was it?

a) factor concentrate
b) plasma transfusion
c) cryoprecipitate
d) hypnosis

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welcome

Laurie Kelley and her daughter Mary.

My 16-year-old daughter Mary is studying for her driver's permit. When we reviewed some of the test questions that might be asked, I realized that I didn't even know the answers!

My coworker Zoraida often helps Latino families in our area when they want to study for their citizenship test. When I hear the kinds of questions they review, I'm surprised at how much I've forgotten about American history.

And when our newest employee, Kathryn Ondek, asked about factor half-lives, I hemmed and hawed. I'm always confusing the factor VIII and factor IX half-lives. Which one is 12 and which one is 24? Do *you* know?

It's easy to forget things that we should know about our history, our laws, and even about hemophilia. As we get ready for another school year (two in college, one to go!) I thought it would be fun to take a hemophilia quiz: What have we learned? What have we retained from those early days of the first diagnosis? There's something for everyone in our quiz: ports, prophy, and products; inhibitors and infusions; joints, genes, and growth mediums. You won't know all the answers, but I'll bet you know a lot. And I bet you'll learn a lot, too!

Share this issue of PEN with other moms, dads, and patients at your next family or chapter meeting. Share it with your teen or young adult with hemophilia. Challenge yourself: refer to this quiz in a few months to see if you remember what you've learned. Even for parents like me, who don't have a child with hemophilia living at home, hemophilia has a way of coming home when you least expect it. Be prepared!

In other news, we say goodbye to Rachel Ruggles, director of Project SHARE. Rachel is off to western Massachusetts to begin a new chapter of her life with her new husband Jon. We wish them best of luck! For any inquiries on Project SHARE, please contact Kathryn Ondek at share@kelleycom.com.

inbox

I enjoyed your February PEN article, "New Treatments in the New Decade." As always, you are at the forefront of sharing exciting and timely news with the community. As your article mentioned, we at Biogen Idec are researching and developing long-acting, fully recombinant therapies for factor VIII and factor IX. We noticed that the chart listing clinical progress showed our rFIX program in a preclinical phase; however, we wanted to share that in January we entered a phase II/III pivotal study titled B-LONG, and dosed our first patients. We shared the results of our phase I/IIa clinical study at the World Federation of Hemophilia meeting in Buenos Aires in July. Anyone interested in learning more about our clinical programs can visit us at www.biogenidechemophilia.com or search www.clinicaltrials.gov for B-LONG or rFVIIIFc.

Jess Swann
Associate Director, Product & Market Development
Biogen Idec Hemophilia

I am the mom of a seven-year-old with hemophilia B. He has a port and we do home infusions twice weekly. Your blog posts on Africa are incredible; what you are doing is truly inspirational. I posted the link to the group I've created, PEI Chapter of the Canadian Hemophilia Society. I have recently connected with a few hemophilia moms on Facebook and found many groups. We often feel like we're the only ones dealing with this, so making the connections sure can make a difference.

Amy Milligan Robinson
Prince Edward Island

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PARENT EMPOWERMENT NEWSLETTER • AUGUST 2010

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Laurie Kelley

Facebook and Factor: Donor Beware

By anyone's standards, Americans are big givers. In 2009, Americans donated an estimated $303.75 billion to charitable causes, according to the Giving USA Foundation. About half of this went overseas, to help with natural disasters, refugees, and social reforms. What's truly amazing is that about 75% was donated by individuals, not corporations or foundations. Whatever the world thinks of Americans, you can't deny that we have big hearts.

And when the US hemophilia community hears of a suffering child in an impoverished country, it's not hard to find someone to help. Most often, help comes in the form of donated factor. The 4 million IU donated to Project SHARE each year is a testament to the US hemophilia community's compassion.

But donating factor just got trickier. Social networking, through forums like Facebook, allow anyone from a developing country to ask hemophilia parents and patients directly for a factor donation. Not knowing international laws, Americans who want to donate factor can unwittingly put themselves in a dangerous, even illegal position, while putting the people they wish to help at risk.

Logistics

Vickie learned this the hard way. In March she sent factor to the Philippines to help a family with hemophilia. "They still haven't received the package," she says. "It was the first time I ever filled out a customs form. That little package cost a lot to mail." Customs is the official process of screening every package that comes into a country: items must be declared in a specific way, or risk being delayed or seized. Biological products like factor are generally not allowed into a country without careful screening, and must be registered with the government. If you're not connected with a nonprofit and don't have paperwork filled out correctly, chances are your package won't get through, and valuable factor will be lost.

Are you even allowed to ship it? Factor is normally shipped only by companies licensed to sell pharmaceuticals. They know the correct assay size for the patient, and can make sure the "cold chain" is intact–correct temperature is preserved from the time the product leaves the manufacturer. That's important because most requests come from countries in the tropics. Without a license, nonprofit status or designated program, you could find yourself in a "gray market" of factor distribution, outside the law–not a great place to be!

Who's Doing the Asking?

In an effort to "do good," Americans can be naïve. A friendly person from a developing country on Facebook befriends you, chats with you, shares photos…and then asks for factor. It's exciting to offer help. But what do you really know about this person?

Does he have hemophilia? Which type? Has he been properly tested? Many countries don't have proper screening to accurately determine factor deficiency and levels. Can he administer factor by himself? Who is supervising his care? If he lives far from a hospital, he may not be under a doctor's regular supervision. In many developing countries, few hospitals specialize in hemophilia care, and most patients do not self-infuse. Do you want to risk sending an injectable drug to someone you know almost nothing about?

What will happen to your factor if it *does* arrive? Will it be used by the patient, or sold to other patients? Sounds cynical, but once the factor leaves your hands, it's out of your control and enters the gray market, where anything can happen. Factor is like gold in developing countries: sometimes it brings a hefty price that can pay for food or rent. Deep need can bring deep desperation.

Whose Factor Is This, Anyway?

"Gary" recently ordered factor from his home care company, but was dismayed when the shipment contained a lot of product that was ready to expire: the company had offloaded on him, and factor can't be returned once it's sold. Gary knew he couldn't use all the factor before expiry, and he didn't want to throw it away. So he called Project SHARE, and we accepted it. There are some valid reasons to donate your factor: when you switch products on your doctor's recommendation, develop an allergic reaction to a brand, migrate to the latest product generation, or even receive the wrong assay size.

But what if you simply want to donate a few vials of your child's in-date factor to help impoverished children? *Don't do it.* Donating factor meant for you or your child, without a valid medical reason, is insurance fraud–a felony in most states, punishable by a fine and prison time.

Before you take any action to donate factor, contact Project SHARE. We're the US experts in screening international requests for factor. We do extensive background checks on each patient, his physician, the national hemophilia organization, and the country's customs requirements. We're also the experts on receiving donations. If you want to donate factor, talk to us first. We'll ensure that it's done properly and legally.

We're also the experts on factor shipping. From one vial to a million IU at one time, we know how to package and process factor, prepare the paperwork, ship it and track it. And we follow up to make sure the factor is used properly.

Vicki was disappointed after her factor went missing. "Next time," she wrote us on Facebook, "I'm going to get advice from you first." Americans are generous, but they need to know that donating factor isn't simple. When you donate through Project SHARE, you can make your compassion work for you and for those you're trying to help–efficiently, effectively and expertly.

Laurie Kelley

Inhibitor Insights is a PEN column sponsored by Novo Nordisk Inc.

Attending School When You Have Inhibitors

Attending school is at once a triumph and a challenge for families with inhibitors. Trying to maintain attendance and good grades while battling inhibitors, and sometimes chronic pain, is difficult and draining. Planning ahead and working closely as a team with the school and your HTC is the key to success.

Working with School Staff

An initial meeting with school staff should take place before school starts and should include your child's head teacher, assistant teacher, teacher's aides, principal, administrative assistant, physical education (PE) teacher, playground aides, and school nurse. Ask your HTC nurse coordinator to attend. He or she may also want to meet with school staff in private to answer any questions they are reluctant to ask in front of you.

"We gave the school written information, and our outreach worker visited them to talk about hemophilia," says Lynley Scott of New Zealand. "We try to build a good relationship with teachers and office staff so they feel comfortable approaching us, asking questions, and calling. We gave them many contact numbers, including the hospital, HTC, and extended family and friends."

Keep your messages to staff simple: Believe your child when he says he needs to call home or needs treatment. Take normal precautions. Observe standard first-aid procedures, if needed, while they are trying to notify you.

Dosing

An inhibitor often requires a demanding dosing schedule when you're treating a bleed or undertaking immune tolerance induction (ITI). If you use FEIBA VH, you'll be treating approximately every twelve hours, so the doses can be given at home before and after school. If you use NovoSeven®RT, you could be dosing every two hours. You may need to infuse at school. Ask these questions: Can you store factor at school? Infuse on school property? Can your child infuse himself at school?

"We treat every six hours if it's a bad bleed," notes Shari Luckey of Michigan. "We used to infuse every two to three hours with small doses (90 mcg/kg) but that was difficult when our son started school. When he had a bleed, we had to go to school every two hours to infuse. Now we give higher doses.* On school days, we infuse before he leaves and right when he gets home."

Missed School

Almost every child with inhibitors is challenged with school attendance, due to active bleeds, constant pain, or even surgeries.

"Leland was in pain every day for eighteen months from sixth to seventh grade," recalls Jane Smith of Massachusetts, "with lots of bleeds and a major surgery. He missed an incredible amount of school. He only misses school now due to pain."

Keep your child up to date when he is absent. Have his classmates bring his homework or have it emailed. Give the teacher weekly updates, and ask for weekly updates on the returned work. Ask for classroom news about special events or new students so your child doesn't feel left out.

Accommodation Plans

All US public schools are required by federal law to provide services for children with special needs. The eligibility criteria and implementation of services vary by state and school district: check your state's special education department website about its accommodation plans and state laws.

A school district may be required to provide tutoring, adaptive equipment or assistive technology devices, an aide, school transportation, extra time to get to class, and more.

Accommodation plans formalize special assistance. They include an IHP (Individual Health Plan), IEP (Individualized Education Plan), and 504 plan.

IHP

This plan ensures access to education for students with special healthcare needs, regardless of whether the student is classified as eligible for special education. It's a formal written agreement involving the school administration and licensed registered school nurse, the student's family, the student, and the student's healthcare providers. The plan coordinates health services at school, to and from school, and at home. For example, your child's IHP might allow for infusions to be done on school property.

IEP

An IEP is designed to meet a child's unique academic and functional needs. It

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* Megadosing is the doubling or tripling of an initial dose, rather than using a lower dose spread out over several hours. Megadosing has not been approved by the FDA and is considered an off-label use. It may increase the risk of thrombotic complications (excessive clotting at other sites). Do not try any dosing changes without first consulting your hematologist.

Kevin Correa

Transitions is a PEN column sponsored by Baxter BioScience

Your Online Persona: Employers Are Reviewing It. Why You Should, Too.

Social networking sites like Facebook* have revolutionized the way people forge new relationships and rekindle old ones. For the hemophilia community, these sites provide an ideal platform to connect with others across the globe to share experiences, information, and inspiration.

Everyone, it seems, is opening a Facebook account. Even corporations have entered the social networking realm, but not just for marketing purposes. Increasingly, companies are using social networking sites to screen potential hires. If you have hemophilia and you're looking for a job, could you be screened out of the running if you post personal health information online?

Red Flags

In a 2009 survey of more than 2,600 hiring managers, the job listing website CareerBuilder found that almost half of employers use social networking sites to screen potential employees.

This means that when you apply for a job, there's a good chance that someone in human resources will snoop around the Internet looking for information about you that wasn't necessarily on your resume–like hemophilia?

First, the good news: according to the CareerBuilder survey, several companies report hiring someone based on information they found in the candidate's Facebook profile. Here are some of their reasons:

- The profile provided a good feel for the candidate's personality and fit within the organization.
- The candidate was creative.
- The candidate showed solid communication skills.

And the bad news: a greater percentage of companies report *not* hiring someone because of information they found in the candidate's profile. Some of the reasons:

- The candidate posted provocative or inappropriate photos or information.
- The candidate posted content about alcohol or drug use.
- The candidate showed poor communication skills.

Cleaning up bad language and removing sketchy photos is easy enough. But what about references to hemophilia? What–if anything–should you do about those?

Friend Requests

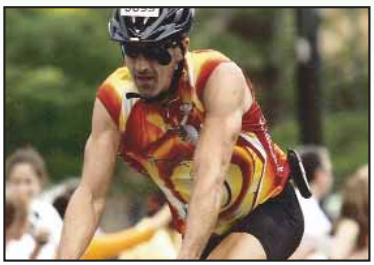
Barry Haarde, 44, is a member of what he calls the 3-H Club. He has hemophilia, hepatitis C, and HIV. Until recently, Barry did not share that information outside of his family and close friends.

But last year, he did something he couldn't have imagined a couple of years earlier: he joined Facebook, and let the world in on his secret.

The catalyst for the change was the 2007 death of his brother, who also had hemophilia, hepatitis, and HIV.

"I felt some personal regret that I wasn't contributing more to the community," recalls Barry. "Now, I'm more engaged and see myself as a self-styled educator."

The Internet, and specifically Facebook, has helped Barry connect with others in the bleeding disorder community. "A year ago, I hadn't anticipated that I would be doing as much as I am," he says, referring partly to the fact that



Barry Haarde

he has gone from relative anonymity to having videos of his story posted on the Web for public consumption.

Though secure in his current employment (his employer is aware of his medical conditions), anxiety creeps into Barry's mind at the thought of having to search for another job. "I'm aware that [half] of employers search the Internet for information about job seekers," he says. "If I lost my job, I would panic. When you google my name, all that information about hemophilia, hepatitis, and HIV is out there."

For Barry, the concern in a job hunt is no longer fear over the stigma of HIV. His real worry is that an employer would focus only on the high healthcare costs associated with his medical conditions, rather than on his qualifications as a candidate.

In spite of this risk, Barry has not censored the information he shares on the Web.

Self-Censorship

Hemophilic Facebook users face a conundrum. On the one hand, the community benefits greatly when people like Barry openly share their experiences. On the other hand, the information they

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* Facebook is only one of several popular social networking sites, but for simplicity we use it here to refer to all such sites.

PROJECT SHARE

It's time to give back

Laurie Kelley

Simon's Struggle

In the US, we honor and reward people with bleeding disorders who excel in golf, karate, and even scaling mountains! These are all activities enjoyed by members of our community. In Africa, we need to honor people like Simon, who simply survive each day.

I met Simon, 26, in April during a humanitarian visit to Kenya. My partners on this journey were Maureen Miruka, founder of Jose Memorial Haemophilia Society–Kenya (JMHSK), and Paul Kamau, 21, a young man with hemophilia, who is administrative assistant for JMHSK.

To reach Simon's home, we drove nearly three hours north of Nairobi, deep into the Kenyan countryside, dodging terrible potholes. The shoulders of the road drop swiftly, so we had to be careful not to veer off. Unmarked speed bumps, or "sleeping policemen," blend in with the road and can cause severe damage.

We arrived in Nyahururu, a bustling town. The Kenyans are always walking, so I was surprised to see a long line of young men with motorcycles idling–the local transport. We spotted Simon sitting on the ground under a gas station sign, waiting for us. I gave up my front seat when I saw how painfully and slowly he was walking.

Simon directed us to his home via a road made of the red, rich soil of Kenya. As the road ascended into the hills, we dodged bicycles with massive loads of grass or wood, their drivers pushing them resolutely upward. Women and children walked, carrying huge loads of potatoes in sacks supported by bands around their heads. Cattle stumbled past us down the road into town.

Our tires spun in the muddy soil, but the car clambered up a dirt path strewn with rocks, and we finally arrived at Simon's farm, perched high on a hilltop with a spectacular view of the Kenyan countryside.

The farm is a rambling collection of split-log cabins and pens for animals. Rural living, even primitive, if judged by American standards. Simon's mother greeted us, but did not smile. The entire family was grim, tight-lipped. "They are stressed," Maureen told me quietly.

We all sat inside the small home, more like a shed, where Simon's mother lives. I glanced at the corrugated tin roof, typical in the developing world, the "wallpaper" of cardboard sheets stapled together, and the light peeping in through the gaps.

Simon hobbles on crippled legs. When he gets a bleed, this is what happens:

photos: LA Kelley Communications, Inc.

Simon of Kenya: Rural living means hours to the nearest hospital

In pain, Simon must walk down the dirt path to the dirt road, and then walk three miles on this road down the hill and into town. There he waits for the local bus to drive him to Nairobi–to the only hospitals able to care for people with hemophilia. Our drive had taken more than two hours; the bus takes two to three times longer, with stops.

And all the time, Simon is in great pain.

It was clear to us all that Simon needs to keep factor at home. He knows how to self-infuse. But the Kenyan government buys no factor, so there is rarely any available.

We presented Simon with a gift of factor VIII, and we pledged that somehow, we'd keep him stocked with factor. By the time we shuffled back to the car, everyone was beaming–like a ray of light brightening his future–but no one more than his mother. Fortunately, factor from Project SHARE can help ensure that Simon survives.

Read more about Simon, and about Laurie's trip to Kenya at http://blog.kelleycom.com

In rural Kenya, isolated hemophilia families suffer without factor

richard's review

Richard J. Atwood

Ann Woodruff

Hemophilia in Pop Culture

Authors use their imaginative powers to create fictional characters, or they base them on real people. Who would have guessed that these extraordinary fictional characters had bleeding disorders?

» A nameless, tattooed circus clown with hemophilia who becomes a murder victim

Feeling Very Strange: The Slipstream Anthology
James Patrick Kelly and John Kessel, eds.
Tachyon, 2006

Originally published in the *New Yorker*, Michael Chabon's story "The God of Dark Laughter" appears in this collection of short stories. In their introduction, the editors explain the slipstream genre of fiction, a style that indeed makes the reader feel very strange.

Recounting the murder investigation of a circus clown, a district attorney notes that the nameless victim was identified as a clown by his purple trousers and jacket, orange waistcoat, oversized shoes, and makeup kit. The clown's forearm was tattooed with a grinning baboon. He had been shot in the head, and all the skin on his cranium had been removed.

Performing the clown's autopsy, the coroner diagnoses hemophilia—another case of delayed blood coagulation throwing off a time-of-death estimate. But the coroner wrongly associates hemophilia with inbreeding in European royal families.

» Aaron, a Jewish vampire with hemophilia who hates the sight of blood and cannot bite his victims

The Saga of a Jewish Hemophiliac Who Can't Bite So He Opens a Vampire Blood Bank
Harry Zelenko
Zai, 1999

A nurse at Samaritan Hospital in New York relates his involvement with Aaron E. Newman, a Jewish man with hemophilia who was bitten by the first-ever Jewish vampire while walking through Central Park. Aaron turns into a vampire, but because he hates the sight of blood, he can't bite any victims. Aaron and the nurse open the profitable Eleventh Hour Blood Bank to meet their needs, but soon find that other vampires want a piece of the action.

Aaron's treatment, for both his hemophilia and his thirst for blood, comes from the blood bank's stockpile: four bags of whole blood daily, enough to cover his prostate surgery. Aaron finds that being a vampire improves his blood clotting and reduces his hemophilia symptoms.

Zelenko's farcical novel, stuffed with laughs for a mature audience, has no redeeming value other than laughter, which is sometimes the best medicine.

» Mayfly, a drug-addicted, high-velocity hit woman with a rare form of hemophilia, who almost assassinates Wonder Woman

Wonder Woman: The Ultimate Guide to the Amazon Princess
Scott Beatty
DK Children, 2003

William Moulton Marston introduced the superhero Wonder Woman in 1941. An Amazon princess, Wonder Woman (aka Diana) comes to America as an ambassador of peace to teach ideals of love, respect, and coexistence. Fighting evil in a wide variety of fiendish foes, she meets Mayfly, who suffers from a rare (unidentified) form of hemophilia and is addicted to the drug Velocity-9. Wonder Woman captures Mayfly, who kills herself in prison. Mayfly appears in the September and November 1993 issues of *Wonder Woman* comics, and her character is summarized in Beatty's book.

These unusual fictional characters with bleeding disorders may not seem real to you when you read their stories. But can you really tell who has a bleeding disorder when you enter a crowded room? Regardless of who they are or what they look like, everyone with a bleeding disorder, whether real or fictional, is extraordinary.

8 Monoclonal refers to which of the following?

a) a purification method
b) a viral inactivation method
c) a cloned recombinant product
d) the source of the plasma used to make factor

9 Recombinant factor is made from factor secreted by cells that contain the human gene for clotting factor. The cells secrete factor into a culture or growth medium, and then the factor is later removed. Not just any cell is able to correctly produce a large protein like factor VIII. The two most common "cell lines" used to produce factor are made from what?

a) Chinese hamster ovary cells
b) baby hamster kidney cells
c) bovine (cow) kidney cells
d) swine (pig) liver cells

10 Which of the following is *not* a viral inactivation method?

a) pasteurization
b) solvent-detergent
c) dry heat
d) nanofiltration

11 Inhibitors are a complication of hemophilia, but they don't always last a lifetime. What's the name for inhibitors that go away on their own after a few weeks to months?

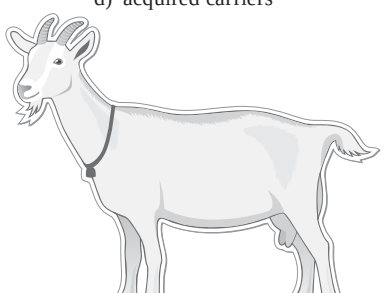
a) intermittent
b) transient
c) evanescent
d) ephemeral

12 What's the chance that your baby boy will have hemophilia if you are a carrier of the hemophilia gene?

a) 25%
b) 50%
c) 75%
d) 100%

13 Most women who are carriers of the hemophilia gene carry the gene in their body's cells as well as in their eggs. But some women do not carry the gene in their body's cells; the hemophilia gene may only be in some of their eggs. What are these women called?

a) obligate carriers
b) genetic mosaics
c) indeterminate carriers
d) acquired carriers



14 When did the first factor concentrate become available in the US?

a) 1930
b) 1968
c) 1980
d) 1992

BONUS QUESTION!
What was its name?

15 Before viral inactivation was implemented in the factor-manufacturing process, thousands of people with severe hemophilia were infected with several types of hepatitis, and later on, with HIV. When was the first virally inactivated factor product available in the US?

a) 1960
b) 1983
c) 1992
d) 1978

16 What is the only known cure for hemophilia?

a) gene therapy
b) kidney transplant
c) liver transplant
d) bone marrow transplant

17 What is a transgenic animal?

a) an animal with human genes in some of its cells
b) an animal with human body parts
c) an animal that has been cloned from another animal
d) an animal with no gender

Medical Musings

18 What are the two most common serious complications of ports?

a) infection and line blockage
b) infection and deep vein thrombosis (blood clots)
c) deep vein thrombosis and port failure
d) skin erosion and cost

19 What does prophylaxis mean?

a) treating a bleed after it occurs
b) infusing factor regularly to help prevent bleeds
c) a treatment for inhibitors
d) a birth control device

20 In severe hemophilia, about 85% of bleeds occur in joints. Of these joint bleeds, up to 90% occur in only three joints. From most frequent to least frequent, which three joints are most susceptible to bleeding?

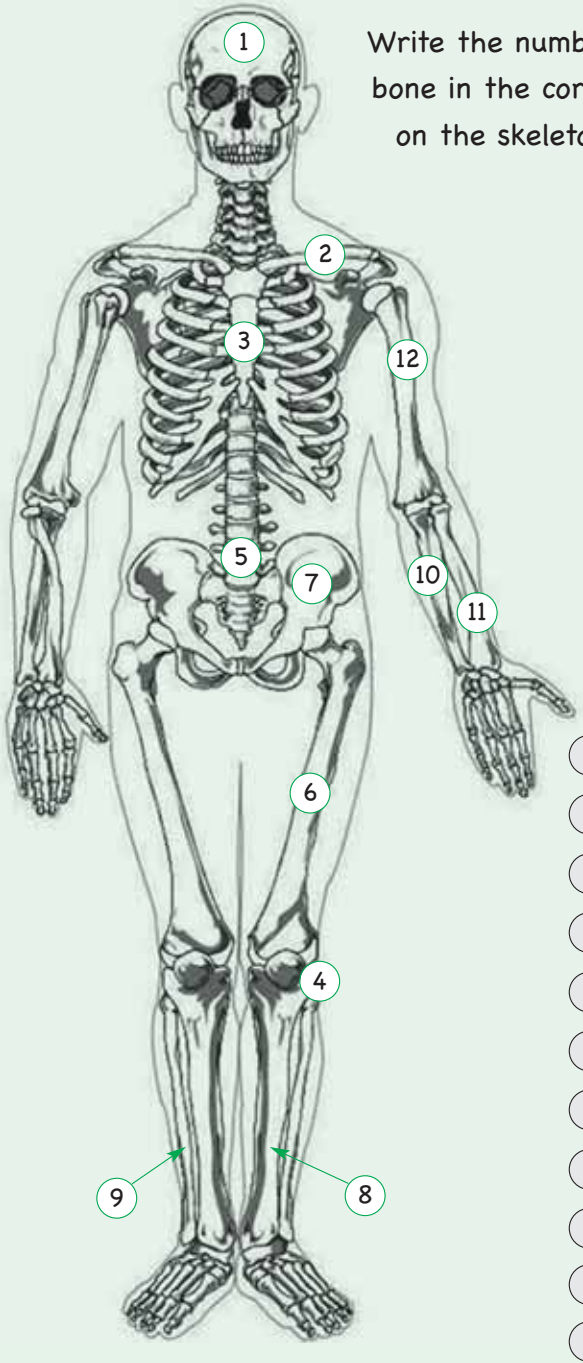
a) ankle, knee, elbow
b) knee, ankle, elbow
c) knee, elbow, ankle
d) elbow, ankle, knee

21 What's the most common major complication of severe hemophilia?

a) intracranial bleeds
b) joint damage
c) anemia
d) excessive bleeding

Bone Up on Your Anatomy!

Write the number of each bone in the correct circle on the skeleton's body.



- ◯ Radius
- ◯ Vertebrae
- ◯ Sternum
- ◯ Pelvis
- ◯ Patella
- ◯ Tibia
- ◯ Femur
- ◯ Fibula
- ◯ Humerus
- ◯ Cranium
- ◯ Ulna
- ◯ Clavicle

» *answers on page 19*

22 **Tranexamic acid and aminocaproic acid are two drugs that help retain blood clots by slowing fibrinolysis–the break-down of the clot. What are their brand names?**

a) DDAVP® Injection and Cyklokapron®
b) FEIBA VH and Amicar®
c) Amicar and Cyklokapron
d) fibrin glue and Amicar

23 **Which of the following drugs should never be taken by a person with a bleeding disorder?**

a) Motrin®
b) aspirin
c) Tylenol®
d) ibuprofen

24 **People with mild or moderate hemophilia A can sometimes control a bleed by taking a drug that releases stores of factor VIII inside their bodies. What is this drug called?**

a) DDAVP
b) FEIBA VH
c) RICE
d) Amicar

25 **What does on-demand treatment mean?**

a) treating a bleed after it occurs
b) infusing factor regularly to help prevent bleeds
c) a treatment for inhibitors
d) a protocol that allows you to get factor immediately when you ask for it at the ER

26 **When infusing through a port, why should you always use a syringe that is 10 mL or larger?**

a) a small syringe creates higher pressure, which can rupture the catheter
b) you can infuse more factor with a larger syringe
c) you can infuse faster with a large syringe
d) a small syringe causes you to waste some factor

27 **Which of the following procedures might your hematologist recommend if your child is a "hard stick" but needs frequent venous access?**

a) angioplasty
b) plasmapheresis
c) kidney dialysis
d) port implantation

28 **While trying to stand at the coffee table, your toddler with severe hemophilia falls backwards and hits his head on the hardwood floor. What should you do first?**

a) wait and see what happens
b) call the hemophilia treatment center (HTC)
c) infuse, then call the HTC
d) rush to the ER

29 **Your toddler fell backwards from a standing position and hit his head. You have infused him and are now at the HTC's ER. The attending physician wants to order a CT (CAT) scan of your son's head. What do you do?**

a) always agree to a CT scan after a head bump
b) agree to a CT scan only if your son has symptoms of a head bleed or has had a previous head bleed
c) agree to a CT scan only if your son has inhibitors
d) never agree to a CT scan

30 **Your toddler has hard-to-find veins, making infusions difficult. Which of the following would *not* make it easier to find a vein?**

a) making sure your child is well hydrated
b) making sure your child is relaxed
c) giving your child a warm bath or putting his hand in warm water
d) tightening the tourniquet

Consumer Challenge

31 Your toddler has a mouth bleed. Which of the following would *not* help control the bleed?

a) applying pressure to the bleed
b) using ice or offering an ice pop to reduce bleeding and relieve pain
c) giving factor if pressure and ice don't stop the bleeding after 20 minutes
d) letting your child use only straws to drink during the bleed

32 When infusing through a port, why must you use only a special infusion needle called a Huber needle?

a) it's sharper and easier to insert into the port
b) it's longer so it can reach the back of the port
c) it won't bore holes in the reservoir of the port
d) it can be left in place for several days

33 Which of the following factor concentrates is *not* a recombinant product?

a) Alphanate®
b) NovoSeven®RT
c) Kogenate® FS
d) Advate

34 What's a 504 plan?

a) a type of insurance plan
b) a payment plan for factor
c) an agreement between you and your child's school
d) a state disability plan for people with bleeding disorders

35 Who should wear a medical alert bracelet or medical ID tag?

a) people with severe hemophilia
b) people who participate in risky activities
c) anyone with a bleeding disorder
d) all of the above

36 At what age does a minor have the rights to consent to treatment and to control protected health information?

a) age of consent
b) age of majority
c) age of minority
d) age of freedom

37 Which of the following is *not* a usual source for ordering factor?

a) your local pharmacy
b) your HTC
c) your home care company
d) you product's manufacturer

38 The Public Health Service (PHS) Drug Pricing Program allows HTCs to buy factor at a low cost and to sell it at a profit. This program is often known simply by its section number. What is the number?

a) 504
b) 402
c) 340B
d) 401(k)

39 You're standing in line at the grocery store with your toddler on your hip. He has big bruises on his shins and knees. The person in line behind you looks either shocked or suspicious, and she asks what happened. How should you react?

a) say "MYOB."
b) ignore her
c) explain matter-of-factly that your son has hemophilia
d) tell her you've enrolled your son in a toddler wrestling class

How did you do? Turn to pages 17–19 to check your responses and read the correct answers to the questions. We guarantee you'll learn something new today!

headlines

manufacturer

First Mobile App for Treatment Management

Bayer announces FactorTrack™, the first customizable mobile application for people with hemophilia A that makes it easier to track and record infusions.

- Captures dosing history
- Records frequency and location of bleeds
- Alerts when it's time for the next infusion
- Allows emailing of infusion information to the patient or healthcare team

Not specific to any brand; available on iPhone, iPod touch, or iPad. Why this matters: This new app may facilitate recording of infusion history–so important in today's insurance climate.

For information: LivingWithHemophilia.com/FactorTrack

New Hemophilia Support Series

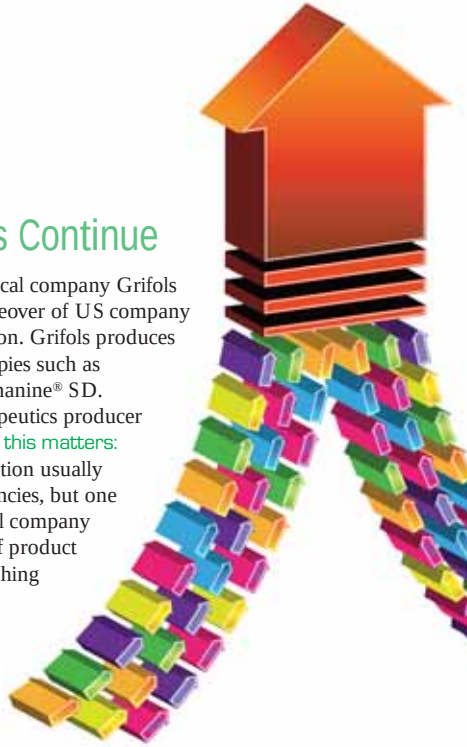
Baxter announces a new series of hemophilia kits to help families adjust to hemophilia.

- Hemophilia Starter Kit: helps newly diagnosed families take the first steps in understanding the basics of hemophilia
- Infusion Kit: provides overview of venous access options, and information on transitioning to home and self-infusion
- *In Full Motion* DVD: helps families and clinicians better understand joint care; includes animated segments and discussions with experts and people living with hemophilia
- Perspectives: experienced healthcare professionals discuss clinical evidence and topics that impact adults with hemophilia

Why this matters: New materials can generate interest in familiar topics, presenting concepts in new ways for greater retention.

For information: www.thereforyou.com

Acquisitions Continue



Spanish pharmaceutical company Grifols has announced a takeover of US company Talecris for $3.4 billion. Grifols produces plasma protein therapies such as Alphanate® and Alphanine® SD. Talecris is a biotherapeutics producer of Koate®-DVI. Why this matters: Continued consolidation usually means greater efficiencies, but one fewer pharmaceutical company may impact choice of product and price. Keep watching developments.

New Size for Advate

Baxter has added a 1,700 IU intermediate potency vial size to Advate, a recombinant factor VIII therapy. Advate now has six potencies, the broadest selection available, ranging from 250 IU to 3,000 IU. As with all therapies, please consult your hematologist before changing dosage size. Why this matters: More infusing and dosing options offer ease, convenience, and flexibility of infusing with a single vial of factor VIII concentrates.

Get Active! Get in Shape!

CSL Behring "Gettin' in the Game" is a one-day competition in golf and baseball designed exclusively for children with bleeding disorders who can't participate in high-contact sports due to their disorder. Professional athletes Perry Parker (golf) and Jessie Schrader (baseball), who both have hemophilia A, will be on hand to coach. Why this matters: Role models and an organized program can help motivate your child to stay fit. This year's dates and cities:

September 11: Pittsburgh, PA
September 25: Durham, NC
October 16: Los Angeles, CA

For information: www.cslbehring-us.com

manufacturer

Common Factors℠ Launched

CSL Behring has launched an innovative live program for people with bleeding disorders and their caregivers. Common Factors provides current insights and information on topics of interest to the bleeding disorder community. Why this matters: This program is available for your chapter or annual meeting.

For information: www.WeHaveCommonFactors.com
888-508-6978

DTC Ad Spending Up

Despite the economic recession, the pharmaceutical industry increased its direct-to-consumer (DTC) spending in 2009 to $4.51 billion, up by 1.9%. Overall DTC ad spending increased in 2009. Online advertising generated the strong growth in spending, increasing by 30.8% to $117.4 million. Why this matters: The costs of many of our nation's drugs are partly tied to huge expenditures on advertising. The US and New Zealand are the only countries that permit DTC advertising.

Source: "Direct-to-Consumer Advertising: Review and Outlook 2010" www.pharmalive.com

global

Kuwait's New Hemophilia Society

On April 17, World Hemophilia Day, Kuwait Hemophilia Society celebrated its official opening. For the past three years, the society has helped 150 patients with hemophilia, and is now a registered nonprofit. Why this matters: A national hemophilia organization can help institute policies with the government and marshal resources to protect people with hemophilia.

Source: Abdullah Al-Qattan, staff writer
www.kuwaittimes.net

Hope for Haiti: Mitch and grandmother in his one-room schoolhouse

First Assessment of Haiti

Save One Life made the hemophilia community's first assessment visit to Haiti, beginning an outreach program to locate hemophilia patients after January's devastating earthquake. To date, only one patient with hemophilia is known in Haiti, but a team is in place to begin accepting referrals for other patients. Why this matters: Hemophilia resources for underdeveloped countries like Haiti exist, but in order to accept them, the country first needs a program or structure in place.

For information: www.saveonelife.net

Global Factor Sales Soar

The worldwide market for plasma products climbed to $11.8 billion in 2008, a 69% increase from 2005. North America accounted for 36.7% of worldwide plasma product sales. Recombinant factor VIII, factor IX, and factor VIIa products collectively totaled over $5.6 billion in worldwide sales in 2008, up 50% from 2005. Why this matters: Though factor sales are strong and profitable, only 25% of the world's hemophilia population can obtain factor.

Source: *The Worldwide Plasma Fractions Market – 2008*, a newly available research report from the Marketing Research Bureau marketingresearchbureau.com

nonprofit

Generous Grants

The Hemophilia Alliance donated over $300,000 to the hemophilia community, including $5,000 each for the 12 federal hemophilia regions to support their annual meetings. The Alliance is a nonprofit organization of 80 HTCs that either have or seek to have factor delivery programs under Section 340B of the Public Health Service Act. The Alliance's purpose is to promote the common interests of these HTCs. Why this matters: HTCs and hemophilia nonprofits face shrinking dollars from government and payers. By providing assistance to HTCs with 340B programs, the Alliance helps them deal with the shortfall of revenues so they can continue offering patients outstanding care.

For information: www.hemoalliance.org

NHF Annual Meeting!

November 11–13, 2010: New Orleans, LA

Attend NHF's 62nd annual meeting, "Marching Forward." Geared toward families with hemophilia, the meeting features lectures, programs, and plenty of fun. Why this matters: You can learn a tremendous amount and meet many families while having fun at the largest hemophilia event in the US.

For information: www.hemophilia.org

Connecticut Hemophilia Society, Inc.

New Hemophilia Organization

Connecticut Hemophilia Society, Inc. (CHS) is the newest US hemophilia nonprofit. CHS is dedicated to improving quality of life for people with bleeding disorders and their families. CHS provides education and financial support, and supports scientific research on a cure for hemophilia. CHS hosted its first event at Lake Compounce, CT, on June 19, with 200 attendees. Why this matters: Consumers formed CHS to address the unmet needs of state residents.

For information: www.cthemophilia.org

advocacy

US Launches Healthcare Website

HealthCare.gov is a new website from the Department of Health and Human Services designed to help people understand their health insurance options and the new law, and find a health plan. In October, the website will launch a tool that allows people to compare the premiums and prices of various insurance policy plans. Why this matters: You can use this tool to shop for the plan that best meets your needs and budget.

For information:
www.healthcare.gov

education

Alex Lieber Scholarship Winners

Awarded August 1, 2010

Seth Kelly
Alabama

"At a young age I would have said my greatest challenge was facing the embarrassment of going to school on crutches almost on a weekly basis. Now, it is creating an understanding among the same peers that I was so afraid of facing…I hope to be a motivating factor in the lives of those I touch."

Academics are a top priority for Seth. An exceptional student, admired and respected by his peers and teachers, Seth is also a leader, a unique young man with the drive to succeed in school and in his community.

Seth will be a sophomore at Jacksonville State University this fall.

Dylan Hillard
Texas

"My greatest passion in life is writing… Writing can found nations, create love, rally armies, build followings and point out evil in our world…I would like to help give shape to perception and bind humans together through my writing."

A remarkable young man and National Honor Society member, Dylan plans to study writing and communications. As a church volunteer and a role model at hemophilia camp, Dylan's involvement in community service shows his strong character, commitment, and compassion.

Dylan will be a freshman at Texas A & M University this fall.

Visit www.kelleycom.com after Jan. 1 for information about the 2011 Alex Lieber Memorial Scholarship award.

Inhibitor Insights... from p. 4

outlines the child's present levels of academic and functional achievement, and provides measurable yearly goals, listing any special and appropriate accommodations. The federal government reimburses the school for an IEP. This plan is a legally binding document and requires progress reports. For example, an IEP might provide a home tutor for a child who misses school regularly, or may allow for extended absences because of medical treatment.

504 plan

This plan prohibits discrimination based on disability. It requires schools to ensure that students with disabilities have full opportunities to participate in all aspects of regular school on an equal basis with students without disabilities. A 504 is offered to students who don't qualify for special education services, but who have needs that require some accommodation. This plan is funded directly out of the school's budget. A 504 plan might mean that your child receives extended time on tests or assignments.

Explore other ways to keep your child on track, such as cyber schooling, also called distance learning. With today's technologies, sometimes school is only a mouse click away, helping to keep our children with hemophilia and inhibitors in school and on the road to a better future.

Excerpted from *Managing Your Child's Inhibitor: A Practical Guide for Parents* by Laureen A. Kelley with Paul Clement, available in September 2010.

Transitions... from p. 5

share could potentially be used against them. Given that social networking sites are such a great resource for connecting people in the bleeding disorder community, it seems contradictory to advise anyone to censor the information they share on bleeding disorders. As Barry notes, "You hate to discourage anyone from expressing who they are."

Rather than discourage that self-expression on Facebook–even if it includes discussing hemophilia publicly–we should focus instead on helping people with bleeding disorders make informed decisions about what they post online. And we should stress that it's not just friends who are viewing the information.

The issue of employers screening candidates via Facebook has touched a nerve beyond this community, and several grassroots efforts aim to stop the practice. Also, legislation is now under review that may eventually make the act of self-censoring medical information on Facebook unnecessary.

GINA

Protection against employment discrimination under the Americans with Disabilities Act (ADA) was discussed in "Be Honest. Just Not Too Honest" (PEN *Transitions*, February 2009). Although ADA prevents employers from asking about a disability during the interview process, it does not directly address that employer's acquisition of medical information gleaned from social networking sites.

In 2008, the Genetic Information Nondiscrimination Act (GINA) was signed into law. Title II of GINA prohibits discrimination in employment based on genetic information. With a few exceptions, the law deems illegal the acquisition of genetic information–for example, that an applicant has hemophilia. But lawmakers are currently working to determine if genetic information acquired from Facebook will be prohibited or exempted under the law. In other words, it's unclear if a potential employer has broken the law simply by looking at Barry's Facebook page and learning that he has certain medical conditions.

Final Title II regulations were expected in May 2010, but have been delayed.

Private versus Public

Even if you don't talk about your hemophilia in your Facebook profile, it's a good idea to give your account the once-over. What does it say about you? What might a stranger reading your profile think? *This guy really has his head screwed on straight!* Or, *What a disaster!*

Does your profile need an overhaul?

Until you've thoroughly evaluated the possible consequences of sharing *any* private information on Facebook, especially about your bleeding disorder, it's a good idea to max out your privacy settings, giving the public as small a window into your personal life as possible. In the online world, once the genie is out of the bottle, it's hard to stuff it back in.

Barry doesn't envy the privacy decisions many younger guys will face. "Young men just entering the job market must really consider how they want to approach the issue. They have to think very carefully about what they want share."

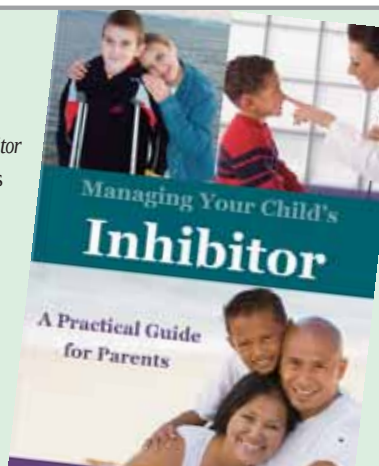
This is a complex question, and there isn't a *one size fits all* remedy. Talk it over with friends, family, and your HTC staff to help find a solution that works best for you.

World's First Book on Inhibitors

Pre-order your copy of *Managing Your Child's Inhibitor* by Laureen A. Kelley with Paul Clement. This groundbreaking book extensively covers pain management, surgery, family life, treatment, and more–from the parents' and patients' point of view.

To pre-order: 978-352-7657 or info@kelleycom.com

$18.95 plus shipping and handling



Inbox... from p. 2

MY SON IS FOUR YEARS OLD AND HE HAS severe hemophilia A. I am one of the ladies who started the Li'l Bruisers email group (it's graduated to a yahoo group now). I was given one of your books on the day my son was diagnosed, and it was the greatest help in those first few months.

Lyndsay Manley
Kentucky

THANK YOU VERY MUCH FOR YOUR inspirational book *Success as a Hemophilia Leader*, which I received from the Dutch Hemophilia Society. We had our first symposium in Suriname. As you can see [photo at right], I am holding your book up very proudly!

John Tjong Tjin Joe, President
Suriname Stitching voor Hemofilie Patiënten

I ENJOYED SPEAKING WITH YOU ON THE phone recently, Laurie. Your story was so helpful to me when my son was born in 1998 and I was trying to get my head around dealing with hemophilia. Thanks for the work that you do. It is so appreciated!

Megan Olson
Georgia

THANK YOU SO MUCH FOR THE resources that you sent to New Zealand. As always, they arrived in time for our New Families Camp. The parents all really appreciated *Teach Your Child about Hemophilia*. We do appreciate the support that you give us. Sometimes we can feel a little isolated way down here, and being such a small country, we do not have the financial resources or expertise to develop resources such as yours. Being able to access your resources is much appreciated, and they are brilliant.

Colleen McKay
Manager, Outreach Services
Haemophilia Foundation of New Zealand

THANKS SO MUCH FOR THE MAY PEN. As usual, I enjoyed reading it. I particularly liked the [*Transitions* column] on the medical identification bracelet. I recently got a MedicAlert® bracelet for my nine-year-old son Gavin (severe hemophilia A) and it is quite funky. The disc is on a velcro strap and nice to take off for a shower. It's also good for sports because the chain does not get in the way. I had

Members attend the first symposium of Suriname's hemophilia society

to explain to my son why he needs it: what happens if we are in an accident or if he is with friends and something happens? Since then, I have told other people to get them too.

Lindy Kerst
South Africa

Project SHARE

PROJECT SHARE'S ANNUAL REPORT is great. You should be very proud of the service you provide in making a difference in the lives of many. "A picture is worth a thousand words" and the pictures in the annual report say it all. Making a difference in one's quality of life and even saving lives is what I love about Project SHARE. Thanks for this service, and thanks to all who support it.

Dana Kuhn
President/Founder
Patient Services, Inc. (PSI)

MY SON ARYLL DAVID, 12, GOT an intracranial bleed in April 2009. He was confined in the hospital. I was hopeless until Andrea Trinidad gave us the contact number of Father Don Kill to tell him about my son's situation. He contacted Project SHARE. God is so great because Project SHARE sent us factor for my son. Now Aryll is in his first year of high school.

Rose David
Philippines

THANK YOU FOR THE HELP YOU OFFER us. At the hospital where I volunteer, there are many children with disabilities. Yesterday I saw a child with a very bad knee bleed, and the doctor told me that because of your help, my son Luis Carlos will not have such bad bleeds and such a severe disability, God willing. Thank you for your kind help and generosity, as well as the Project SHARE team. Also, thank you for the book you sent to me; it's helping me tremendously. I am learning a lot and trying to help other parents in Guatemala. Thank the Lord for your organization.

Gricelda González
Guatemala

I AM 21 AND HAVE SEVERE HEMOPHILIA A. I'm studying in the third-year university, and I have several problems at present: a lot of injuries, a psoas bleed, and bruises in the knees, hands, and shoulders. When I have a bleed and I go to a hospital in another city (120 km away) the doctor tells me every time, "We have little factor and we cannot waste it!" Now I have three swellings in both knees and my shoulder. The real problem is that this month I need to complete my studies. I can't do it because I can't walk or move. My family is very poor and cannot buy factor. Please help me.

Elassali Elmehdi
Morocco

Ed. note: Project SHARE is verifying the request and will send factor.

Test Your Knowledge... from cover

Check Your Answers!

1. Correct: B. Hemophilia A is caused by a deficiency of factor VIII. Hemophilia B is caused by a deficiency of factor IX. And there is no such thing as factor III!

2. Correct: C. Factors VIII and IX are produced mainly in the liver.

3. Correct: B. The US Centers for Disease Control and Prevention (CDC) estimates that about 17,000 people in the US have hemophilia. The CDC estimates the incidence of hemophilia at one case for every 5,000 male births. With a population of 300 million, the US probably should have about 30,000 with hemophilia, but many people with hemophilia died of HIV and HCV after being infected through their clotting factor concentrate in the late 1970s and early 1980s.

4. Correct: B or C. According to the CDC, based on the incidence of one case of hemophilia for every 5,000 male births, the closest estimate is that 400 babies are born with hemophilia each year in the US. But no one knows the exact number because hemophilia is not a reportable condition.

5. Correct: B. The half-life of factor VIII concentrate is closest to 12 hours. Half-life is the time it takes for half of the infused factor to become inactivated. For example, if you infused enough factor to provide 100% correction, then in 12 hours you would have half the factor left. In 24 hours you would have one quarter left, and so on.

6. Correct: A. Shyness in social situations is not a characteristic of inhibitors!

7. Correct: C. Dr. Pool found that when plasma is frozen and then slowly thawed at a low temperature, a precipitate (solid) forms that is rich in clotting factors. Soon after her discovery, cryoprecipitate ("cryo") became the first effective treatment for hemophilia. National Hemophilia Foundation (NHF) now gives an award for researchers in bleeding disorders, called the Judith Graham Pool Postdoctoral Research Fellowship—and rightly so!

8. Correct: A. Monoclonal refers to a purification method—a way to make your factor concentrate contain mostly factor and to exclude unwanted extra proteins. Plasma or growth medium containing factor VIII or IX is poured through a column coated with plastic-like beads. Attached to these beads are monoclonal antibodies. The factor sticks to the monoclonal antibodies, but everything else—including other proteins—passes through the column. The factor is then released from the column, resulting in almost pure factor.

9. Correct: A and B. The two most common cell lines used to produce factor are made from either Chinese hamster ovary cells or baby hamster kidney cells.

10. Correct: D. Nanofiltration is a purification process used to remove unwanted proteins in the production of some factor concentrates. Although it removes all but the smallest viruses, it isn't considered a viral inactivation process because it does not destroy or inactivate viruses. Several products use both nanofiltration and viral inactivation processes.

11. Correct: B. Inhibitors that go away on their own are called transient inhibitors.

12. Correct: B. It doesn't matter how many pregnancies you've had, or whether you already have a boy with hemophilia: if you're a carrier, the chance of each of your sons having hemophilia is 50%.

13. Correct: B. Unlike most carriers of hemophilia, some women don't carry the

Pulse *on the Road*

LA Kelley Communications is taking *PEN's Insurance Pulse* on the road!

First stop...
Hemophilia of Indiana Annual Meeting on August 28, 2010

For more information about future events, please visit our website at www.kelleycom.com

LA KELLEY communications, inc.

Sponsored by
Baxter

gene for hemophilia in their body's cells. These women are called genetic mosaics. They have the gene for hemophilia in one or more of their eggs, which creates a unique problem: these women don't show up on genetic tests as carriers! The 50% chance of having a boy with hemophilia that applies to most carriers doesn't apply to genetic mosaics because the number of eggs that carry the hemophilia gene is unknown.

14. Correct: B. The first factor concentrate available in the US was introduced by Baxter in 1968.

Bonus answer: Hemofil

15. Correct: B. The first commercially prepared virally inactivated factor available in the US was introduced in 1983. Starting with the introduction of factor concentrate, people were being infected with hepatitis. Later, around 1978, people began contracting HIV from untreated factor concentrates.

16. Correct: C. Liver transplants have cured people with hemophilia A and B. Why? Because the liver is the primary site of factor production. Although research is still underway, experts believe that eventually gene therapy will successfully "cure" hemophilia—at least for a limited time, perhaps a few years, before the therapy would have to be repeated.

17. Correct: A. A transgenic animal has human genes (or genes from another animal) in some of its cells. Current research is exploring ways to incorporate the gene for factor IX or factor VIII into some of the cells of a female pig, goat, or rabbit. This would make these animals capable of producing human factor, expressed through the milk. It's hoped that this research will create low-cost factor through high production—great news to most of the developing world, which currently has little or no factor. And we've never heard of an animal with no gender!

18. Correct: B. Infection and deep vein thrombosis (blood clots) are the most common serious side effects of ports. Each affects about a third of people with ports over the lifetime of the port.

19. Correct: B. Prophylaxis means infusing factor regularly to help prevent bleeds. For hemophilia A, this usually means infusing every other day or three times a week. For hemophilia B, this usually means infusing every three days or twice a week. Why the difference? Factor IX has a longer half-life!

20. Correct: C. About 45% of bleeds occur in the knee, 30% in the elbow, and 15% in the ankle.

21. Correct: B. The most common major complication of severe hemophilia is joint damage. Excessive bleeding isn't a complication—that's what defines hemophilia!

22. Correct: C. Amicar and Cyklokapron are two antifibrinolytics available in the US. Antifibrinolytics help retain blood clots throughout the body, but are used mainly for clots in the nose or mouth. Mouth and nose bleeds are hard to treat because enzymes in the saliva and mucous membranes tend to quickly dissolve blood clots. This results in oozing of blood or rebleeding hours to days later. Amicar and Cyklokapron slow the breakdown of blood clots and are effective only after an infusion of factor concentrate.

23. Correct: B. Aspirin prevents blood platelets from becoming "sticky" and forming a platelet plug at the site of an injury. This stops a blood clot from forming. The effect lasts for the life of the platelets, about ten days. Ibuprofen (brand name Motrin) reduces pain and is an anti-inflammatory. It also affects platelet adhesion, or stickiness (for only about four hours) and it should not be taken during a bleed. Tylenol is the painkiller most often recommended for people with bleeding disorders.

24. Correct: A. DDAVP® Injection or a nasal spray called Stimate® both contain a synthetic hormone called desmopressin that, when inhaled, releases the body's own stores of factor VIII into the bloodstream. This product works only in people with mild or moderate hemophilia, who have some reserves of factor VIII that can be released by the drug. FEIBA is used to treat bleeds in inhibitor patients, and Amicar is used after an infusion to control mouth and nose bleeds. And RICE...well, if you answered C, then you need to read up on hemophilia!

25. Correct: A. On-demand means treating a bleed after it occurs. This treatment is usually less effective than prophylaxis at preventing long-term joint damage.

26. Correct: A. When compared to large syringes, and when using equal plunger pressure, small syringes generate a lot more pressure at the needle end of the syringe. If you use a small syringe with a port, you can rupture the catheter or separate the catheter from the port because of this excessive pressure!

27. Correct: D. A port is a device that is implanted in the body to make infusions easier. Ports offer a large target for the needle and eliminate the need to locate a vein.

28. Correct: C. For all hard head bumps, always infuse first (if possible), and then contact the HTC. Never wait and see!

29. Correct: B. Although some physicians and hospitals routinely order CT scans for any head bump, this is usually not needed. Head bleeds in people with hemophilia are relatively rare, and CT scans don't always detect early intracranial bleeds. X-rays can cause cancer, and the risk increases with the dose. Compared to adults, children are more susceptible to genetic damage from X-rays. The use of CT scans is currently being reviewed by the FDA.

30. Correct: D. Ensuring that your child is well hydrated, warm and relaxed will help you locate a vein. Making a tourniquet tighter will not make a vein easier to see. You can also have your child squeeze a rubber ball from time to time to build up the vein.

31. Correct: D. Children with mouth bleeds shouldn't use straws (or pacifiers) because the sucking force needed to use the straw can dislodge a blood clot.

32. Correct: C. Huber needles are specially designed so they don't bore a hole through the silicone septum of the port and destroy the septum.

33. Correct: A. Alphanate is a plasma-derived factor VIII concentrate manufactured by Grifols. NovoSeven RT, Kogenate FS, and Advate are all recombinant factor concentrates, meaning they are not derived from blood plasma.

34. Correct: C. A 504 plan is an agreement between you and your child's school to make sure that your child doesn't fall behind in school work if he misses many school days due to bleeds. Section 504 of the Rehabilitation Act and the Americans with Disabilities Act specifies that no one with a disability can be excluded from participating in any level of schooling if the school or district receives any federal funds. The word disability includes any physical impairment, such as a painful target joint. A 504 plan might include wheelchair ramps, access to elevators in multistory buildings, and tutoring or home instruction.

35. Correct: D. Anyone with a bleeding disorder should wear a medical alert bracelet or tag, or carry a medical ID at all times. An accident could incapacitate you or your child, preventing you from giving emergency responders the valuable health information that might save a life.

36. Correct: B. The Health Insurance Portability and Accountability Act of 1996 (HIPAA) gives minors of a certain age the right to consent to some types of treatment and protect some types of health information—for example, they can consent to having an HIV test and prevent their physician from releasing the results to their parent or guardian. The age at which parents are not allowed to see their child's medical records without the child's permission is called the age of majority, typically age 18. But for some kinds of health information, such as the results of HIV tests, the age of majority is 12.

37. Correct: D. Manufacturers are not legally allowed to sell factor directly to consumers.

38. Correct: C. The PHS Drug Pricing Program section used by HTCs is known as 340B, and factor sales operations by HTCs are often called 340B programs. This kind of program allows HTCs to purchase factor at a significantly reduced price. The HTC then sells the factor to consumers at a profit, and the profit helps to fund the HTC's operations.

39. Surprise! There is no correct answer, though we favor C. You can choose to ask inquisitive strangers to mind their own business, or just ignore them—though that might seem impolite. Everyone reacts differently to these situations, and we're sometimes exasperated with people who suspect child abuse. But it's often enlightening to explain simply, with a smile, that your child has a rare bleeding disorder. These inquiring minds are usually concerned with the best interests of your child—and all children. If you insult or irritate your questioner, you may risk an unpleasant visit from an investigator from Child Protective Services.

Bone Up on Your Anatomy answers from page 9

1. Cranium
2. Clavicle
3. Sternum
4. Patella
5. Vertebrae
6. Femur
7. Pelvis
8. Tibia
9. Fibula
10. Ulna
11. Radius
12. Humerus

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