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Parent Empowerment Newsletter

photo: Smiths Medical MD, Inc.

Seven Reasons to Remove a Port

by Denise D. Vermeulen

Ports are beneficial for many patients with hemophilia—but there are sometimes good reasons to remove them.

For Kristin and David Prior, the decision to remove their son Lynden's port came suddenly, but under no duress. Nine-year-old Lynden, who has severe hemophilia, is a third-grader in Shelburne, Vermont. Lynden enjoys snow skiing, camping, boating, fishing and horseback riding. Prophylactic factor treatment through a port accounts for Lynden's active lifestyle. His port was surgically implanted when he was three, and until last fall, the Priors had no reason to consider removing it.

Lynden's mom Kristin is active in the hemophilia community, serving on several committees and boards, and attending annual NHF meetings. The NHF meeting that Kristin and David attended in November 2003 proved pivotal in their decision to remove Lynden's port.

At a seminar during the NHF meeting, the Priors heard about a research study recently conducted at a treatment center in Indiana. The study's short-term data pointed to an increased incidence of thrombosis (blood clots) at the tip of the port line after five years. Combined with the fact that there is little research about long-term use of ports, this study gave the Priors second thoughts about Lynden's port, and begged the question: *When is it time to remove a port?*

Understanding Ports

A port is a device surgically implanted under the patient's skin, typically in the chest wall. Dr. Kerry Bergman, a pediatric surgeon at Overlook Hospital in Summit,

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inside PEN

LA Kelley Communications, Inc.

welcome

Freedom to Choose

LAST MONTH I SPENT A lovely morning on Ft. Lauderdale beach, Florida with my ten-year-old daughter Mary and my friend Kim, the mother of an adorable six-year-old boy with hemophilia. As I bent to lift Kim's son into the surf, I felt the telltale sign of a port and immediately put him down, not wanting to cause him discomfort. His T-shirt protected him from the sun, but also hid his port. Kim mentioned, "I've been thinking of when we should have it removed." But she admitted that she was unsure about when that should happen. She wanted to protect her son's veins, but also wanted him to feel free to choose to take off his shirt at the beach. We discussed the possibilities, and I told her that this month's *PEN* might have some answers.

Port removal–when, how, why. It's one of the many choices that we must make as parents of children with hemophilia. And while medical decisions like this may seem hardest, for some, decisions about family planning are even tougher. With more medical knowledge and scientific technologies come more choices and more decisions. If we know that hemophilia runs in our family, how do we approach the decision to have another child? We discuss this question, and much more, in this issue of *PEN*.

Decision-making can seem overwhelming, particularly when it involves young children. Of course, we always want to make the right decision, at the right time, with the right information. We fear making wrong decisions. Sometimes, we're hard on ourselves for not knowing enough, or not acting quickly enough. I was often like this while raising my son. But through the years, I've learned that we can lessen our fear of making wrong choices by simply being grateful for *having* choices–having the freedom to choose. Not everyone in the hemophilia world has these choices. In our Project SHARE story, a young man with hemophilia in the Philippines, with limited financial and medical means, struggled with a tumor that threatened his dreams. Thanks to our many generous readers, including HTCs, chapters and home care companies, we were able to donate enough factor and funds for Galo to have a lifesaving operation. We were able to give him a small measure of the many choices we are free to make daily in the US: freedom to move and sit, freedom to work, freedom from pain, even freedom from impending death. Freedom to choose–at once daunting and exciting; equally a privilege and a responsibility. And, I think, always a gift.

PARENT EMPOWERMENT NEWSLETTER AUGUST 2004

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Are you interested in submitting articles to PEN?

PEN is looking for medical professionals, advocates and consumers with good writing skills to submit articles. *PEN* pays $800 for original feature articles and $50 for *As I See It*. For submission guidelines, contact us at info@kelleycom.com. *PEN* will work with authors on editing and content but cannot guarantee that submissions will be printed. Overseas authors welcome!

letters

PAUL CLEMENT'S ARTICLE explaining CJD ["Mad Cows and CJD in the USA," *PEN*, May 2004] is very well written. He has made a complex topic very understandable.

DOREEN EATON
Baxter BioScience

continued on page 19

by Lori Kunkel

"Your Child's Port Needs to be Removed."

When you hear these words, you may feel frightened–just as frightened as you felt when someone first suggested placing a port. Facing decisions about port placement, replacement or removal isn't easy for any parent. If you're considering placing a port for the first time, you may fear the unknown: *How much pain will my child feel? Do the benefits outweigh the risks?* Yet while you may be reluctant to have a port placed, in time you learn to enjoy its security and convenience. If you're considering replacing or removing a port, you may feel guilty because you know that your child must undergo another painful procedure. Whatever your situation, it's essential to include your child in the decision process. Age-appropriate communication will help you make the correct decision for your individual child.

I'll never forget the day my son Brady's port stopped working. I was getting ready to give him his prophylactic dose. After accessing ports for more than four years, this was routine: mix the factor, sterilize the area, access the port, pull back the plunger of the syringe, check for a blood return. Then my heart fell to my stomach. There was no blood return. X-rays confirmed my fears: the catheter was not in the proper place. Brady's port needed to be removed.

I suddenly found myself back at square one, but with a new twist: I knew what to expect this time. Feeling all the emotions of a stressful situation, I found myself again deciding whether to place a port. But because I had already lived through the experience with Brady's first port, I felt a little more educated the second time around. Still, sometimes ignorance is bliss: I found it hard to think about putting Brady through the pain I had witnessed the first time. Then, good thoughts began to surface: infusions had become so much easier for Brady with a port.

Your child may be mature enough to participate in the decision to place a second port–or he may not. My husband and I felt that it was important to speak with our preschool son on a level that he could understand. We explained to Brady that his port was "broken," and the doctors were going to help him by taking it out. Then we told Brady that the doctors could put in a new port if he would like to have another. If he didn't want one, we explained, we would use the veins in his arm to give him his factor infusions. Brady replied that he wanted a new port, but he wanted to pick the color! What a difference between our son's thoughts and our own. Sometimes we forget the simplified thinking of a young child. Brady was concerned about color, while we were concerned about the port placement procedure and the short-term pain that we knew would follow.

Our decision wasn't easy. We had to contend with feelings of guilt: because of our original decision, Brady was now facing another surgery. It's hard for loving parents to watch any child go through something that causes pain. However, we knew that we had to think past the immediate future and the pain involved. Having a port had made life easier in so many ways, not only for Brady, but for us. A port had made infusions less stressful, since we didn't have to try to find a vein on a chubby toddler's arm. Brady had come to accept infusions as something that helped him, not that caused him pain. He told us why he wanted a second port: "It doesn't hurt to have a needle put in your port, but it does hurt when someone puts a needle in your arm."

It's been about three years since Brady's second port was placed, and he is now eight years old. We haven't once regretted our decision. Our two other sons have gone through a similar process: nine-year-old Cody had a second port placed at age six; and Jeff, age 20, is using peripheral venous access after having his first port removed at age 17.

As parents, we should remember that each child is as unique as each family. In our family, we try to base our decisions on the needs and desires of each child, so every decision is the right one–for our sons as individuals, and for our family. While they work well for us, our personal decisions may not be right for someone else's family. It's so important to communicate with your child and find out what he wishes–you'll be much happier with the results. After all, the decision will affect *him* the most.

Lori Kunkel, a trained phlebotomist, is hemophilia resource coordinator for a home care company. She lives in Rochester, Minnesota with her husband Joe and their three children with hemophilia: Jeff, age 20, Cody, age nine, and Brady, age eight.

by Paul Clement

Having Another Child *with Hemophilia*

The decision to have children, and how many children, is basic to most marriages. In fact, it's such an important decision that most men and women sound out potential marriage partners: How do they feel about having children? How many would they like? Often, potential partners with diametrically opposed viewpoints on children cross each other off the "list" of eligible partners. For women who are carriers of hemophilia, the decision to have children takes on another dimension—and requires taking into consideration many factors regarding the risks of having a child with hemophilia.

After marriage, many couples engage in some form of family planning, factoring in such considerations as finances, housing, health insurance, and spacing of children. Many young couples postpone having children until they have stable finances and secure jobs with health insurance. Socioeconomic status and culture play a major role in the perception of what defines the ideal family size. Family planning decisions are deeply personal and require serious thought.

Adults with a family history of a bleeding disorder—and parents who have already given birth to a child with a bleeding disorder—must deal with the same decisions as other families. But they must also consider the possibility that their children may have a bleeding disorder, and carefully weigh the pros and cons of giving birth. This may involve compromising or abandoning a life-long dream of having "the perfect family" and the "right number of children."

Carrier Status and Family Planning

A woman with a history of a bleeding disorder who contemplates having children must grapple with the initial question: *Am I a carrier?* Surprisingly, many women with a family history of a bleeding disorder don't know whether they are carriers. In the case of hemophilia, they often don't even know their factor levels. For some women, the question of carrier status is easily answered. Women whose fathers had hemophilia are *always* carriers, or "obligate carriers." A woman who has a son with hemophilia and a male relative with hemophilia is *assumed* to be a carrier, as are women with two or more sons with hemophilia.

In other cases, a woman's carrier status is not so easily determined and requires further testing in addition to determining her factor levels. A woman with a relative with hemophilia but no sons with hemophilia is a *possible* carrier, as are women with one son with hemophilia and no other family members with hemophilia. Measuring the factor level of a possible carrier is the first step in determining carrier status. For hemophilia A (factor VIII deficiency), the test for factor level should be repeated at different times because factor level can vary with hormones and stress. Women who are carriers often have low factor levels. Knowing only a woman's factor level and family history, a genetic counselor can determine, with 85% accuracy, whether the woman is a carrier.[1] For women with one son with hemophilia, no family history and a normal factor level, genetic testing may be required to determine carrier status. Genetic testing can determine carrier status with a 95% confidence level.[2] For a small percentage of women, carrier status cannot be determined.

I'm a Carrier. Will My Child Have Hemophilia?

Once a woman's carrier status is known, she and her partner might ask the next question: *What's the probability that our baby will have hemophilia?* For hemophilia A and B, this is easy: hemophilia A and B are both sex-linked recessive disorders, with the defective gene located on the X sex chromosome. Sex-linked disorders affect primarily males. This inheritance pattern is easy to predict: women who are carriers have a 50% chance of passing the gene for hemophilia to their sons, and a 50% chance of passing the gene to their daughters.

For the most common form of von Willebrand Disease, Type I, all children of affected parents will have the disorder. For other forms of VWD, a geneticist may be needed to help determine the inheritance pattern.

Insurance Matters[3]

The US does not have universal health care: individual citizens must purchase coverage from an insurance company,

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[1]World Federation of Haemophilia, Facts for Families, 1996. Published jointly by the World Health Organization and the World Federation of Hemophilia. www.wfh.org/ShowDoc.asp?Rubrique=31&Document=180&Contentid=194, [2] Canadian Hemophilia Society, Heredity of Hemophilia. www.hemophilia.ca/en/2.1.4.php,
[3] The insurance discussion in this article has been simplified to save space. For more information, and before making any decisions about insurance, contact your employer, home healthcare company, the pharmaceutical company of the product you use, and other organizations such as Patient Services Incorporated (www.uneedpsi.org/about.cfm).

Galo Villamil: A Teacher's Courage

by Annie Schwechheimer

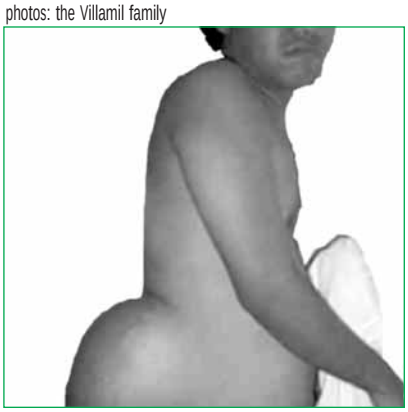
photo: the Villamil family

Galo Villamil lives in Zamboanga, Philippines with his elderly mother. Her pension is just enough to cover the cost of everyday living–but not much more. If Galo were an ordinary 26-year-old man living in a developing country, the situation would be challenging enough; but unfortunately, Galo has hemophilia and must deal with its added financial and emotional stress.

Galo was diagnosed in 1994 after falling on wet pavement and experiencing severe pain in his right leg. He couldn't walk for two months. Realizing that the injury would not heal, Galo went to Cebu Doctors Hospital in Cebu City. He informed the attending physician of his family's medical history–his brother had hemophilia and died in 1988 due to complications from a bleed. Galo was immediately tested, and received the ominous news: he also had hemophilia.

Since then, with hard work and determination, Galo has managed to excel. He earned a bachelor's degree in elementary education and passed his teacher certification examination. During his time in school, Galo experienced bleeds that were serious enough to require hospitalization; this interfered with his studies, but he never lost the focus he needed to fulfill his dream. In order to pay some of Galo's medical costs, his mother was forced to sell a small parcel of land that she had inherited.

photos: the Villamil family



Humiliation, suffering, poverty: Galo's disfiguring tumor.

Nearly three years ago, a fist-sized tumor appeared on Galo's buttocks. Since the tumor was relatively small, Galo's doctor advised him to go home and watch for any changes. By June 2002, the tumor had increased significantly and Galo began to have problems walking and standing. His life was now in danger, and, because of the pain and discomfort, he had to make the difficult decision to stop teaching. When Galo's doctors explained that he would need 120,000 IUs of factor VIII for his surgery, he became depressed. *Where would he get this medicine?* Only temporarily disheartened, Galo was soon bolstered by friends and family, who encouraged him to ask organizations outside the Philippines for help.

One of these organizations was Project SHARE. In January 2004, Project SHARE was contacted by Dr. Mary A. Chua, chief hematologist for the Santos Tomas University Hospital in Manila, and also a founder of the national hemophilia organization of the Philippines, HAPLOS (Hemophilia Association for Love and Service, Inc.). Fortunately, Project SHARE was able to donate 60,000 IUs of factor VIII and broker the donation of the remaining 60,000 IUs through a private donor.

This factor donation enabled Galo to have lifesaving surgery. Today, still recovering in the hospital, Galo remains positive about his ordeal. He is truly a testament to the merits of hard work and determination, and a teacher to us all.

To learn more about Project SHARE and how you can help patients like Galo, please visit www.kelleycom.com/iha/projshare.html or contact Director Annie Schwechheimer at (978) 352-7657 or annie@kelleycom.com.

Project SHARE[SM] is an international humanitarian program administered by LA Kelley Communications, Inc., in partnership with ZLB Behring, Baxter BioScience, Bayer HealthCare, Hemophilia Health Services and Novo Nordisk Pharmaceuticals, Inc. Factor donations are primarily from private sources.

Eric Dostie Memorial College Scholarship *Winners 2004*

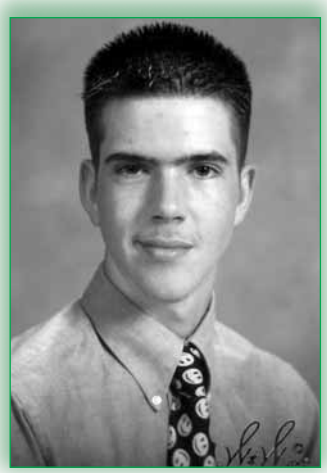
LA Kelley Communications is pleased to announce the winners of the Eric Dostie Memorial College Scholarship for 2004. The scholarship honors the memory of Eric Dostie, a five-year-old boy with hemophilia who was murdered in 1994. Funds provide financial assistance to students and family members in the US bleeding disorders community. The Eric Dostie Memorial College Scholarship is generously funded by NuFACTOR of Temecula, California. We extend our deepest thanks to NuFACTOR, its president Patrick M. Schmidt, and the 2004 review committee for their hard work, compassion and dedication.

Megan Procario

Megan Procario plans to pursue a pre-medical degree, and hopes to become a pediatric oncologist. Her interest in this field began when she was 12. Since then, Megan has volunteered more than 320 hours for the pediatric and oncology wards at a local hospital. She was also a member of her high school's "mock trial" team, and Latin and science clubs. The children she has seen with cancer, says Megan, are some of the "bravest people in the world." She is passionate about her career choice: "Every child saved from a premature death is a miracle."

Michael Reutershan

Michael Reutershan attends Bowdoin College, where he plans to major in biology. He hopes to attend medical school and pursue a career in pediatric hematology. Michael enjoys swimming, basketball and spending time with friends and family. "Although hemophilia can be burdensome," he admits, "I must pay homage to it as it has helped me set concrete goals."



Joshua WIlliams

Joshua Williams plans to study nursing in college and ultimately become a doctor. He explains that having hemophilia increased his desire to enter the medical field because he can relate to patients' experiences. Joshua enjoys making patients feel comfortable, safe and informed about their care, and hopes to "help them through a tough, sometimes frightening time in their lives." In addition to following his passion for medicine, Joshua volunteers at his local high school.

Erin Baschal

Erin Baschal attends the University of Colorado, where she is chemistry major. After graduation, Erin plans to pursue a Ph.D. in pharmacology with the goal of finding cures for genetic disorders. Erin enjoys working with children and volunteering for the Girl Scouts and her local library. Through her love of science and her career, Erin would like to "help people live longer, healthier and less stressful lives."

Jaime Leonard

Jaime Leonard attends Longwood University, where she is pursuing a pre-medical degree. Jaime is an active volunteer for a local hospital and bleeding disorders organizations. She says that spending a lot of time at the hematologist's office piqued her interest in medicine. While she knows she has much work ahead, Jaime affirms, "Every day I know I am one day closer to becoming a doctor and helping people understand what is going on with their health and how to get better."

Brandi Jones

Brandi Jones attends Salem College, where she studies psychology and religion. She plans to obtain a master's degree in child development. In the future, Brandi hopes to become a child life specialist and work with terminally and chronically ill children. Brandi is a long-time employee at Camp Carefree, a camp for children with chronic illnesses. She says, "Children are this world's future, and if I am able to help them have a better life, then I feel that this will help humankind."

Christopher Rasch

Christopher Rasch plans to pursue a biology degree and ultimately attend medical school. He hopes to become an orthopedic surgeon. Christopher explains that the time he spent in hospitals due to hemophilia has helped him become passionate about science, medicine and helping others. He enjoys swimming and weightlifting. By becoming a doctor, says Christopher, he can enable people to "live their lives normally and without permanent handicaps."

Rebecca Guy

Rebecca Guy attends Boise State University, where she is pursuing a degree in nursing. For Rebecca, involvement in the medical field has always been a goal. She feels that it would be meaningful to help cardiac and hemophilia patients professionally on a daily basis. A proud mother of five children, one with hemophilia, Rebecca enjoys stamping, hiking and photography.

Melody Selby

Melody Selby plans to pursue a degree in elementary education and become a first grade teacher. Through various local and civic organizations, Melody has devoted considerable time to teaching and guiding children. Recently, she participated in a mission trip to Mexico. Melody, the daughter of a hemophilia patient, explains, "I want to do whatever I possibly can to help children learn and grow from their hardships, be successful in life and make healthy decisions."

Applications for the Eric Dostie Memorial College Scholarship 2005 will be available through NuFACTOR after November 1, 2004. For more information, please visit the LA Kelley Communications, Inc. website at www.kelleycom.com/finaid/finaid.html or contact NuFACTOR at (800) 323-6832.

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[Q] Gender Selection:

If possible, would you choose to have a girl just to avoid having a boy with hemophilia?

A student doing research for an English paper posted this question to the Bleeding Disorders mailing list. Here's how readers responded:

[A] I have a 14-month-old son with hemophilia A. When I became pregnant, I knew that hemophilia ran in my family: my grandfather and his brothers had hemophilia, and all of his daughters were carriers. My aunts also have sons with hemophilia.

I would never try gender selection for a girl just because males are more affected by this genetic disorder. Since I am a carrier, each of my two daughters has a 50% chance of being a carrier, and may have sons with hemophilia. By choosing to have only daughters, you're just skipping out on having a son with hemophilia–only to deal with grandsons who have hemophilia. What's the point? You get what God gives you, no matter what problems the child may have. All children are precious, and we should be thankful for them.

ANONYMOUS

[A] I think that science is doing great things! Yes, you have a 50% chance of passing the hemophilia gene to a girl. But in 20 years, what if the eggs can be sorted, allowing your daughter to choose *not* to pass on the gene? My main problem with my son Colton–and his future–is the cost of factor. His factor bill now runs about $10,000 per month. That is a huge burden to put on a child. And as an adult without insurance, how would he make it? It would be difficult for me to have another child knowing that he'll live his whole life with the decision I make. I believe that God gives you only what you can handle, but I also think that God shows you when you're in over your head. I respect any family that chooses to have a child when the mother is a carrier, but we all know how much we can personally handle.

RANDI CLITES
Ohio

[A] There is no right or wrong answer. Our second son doesn't have hemophilia, and we just learned that a third is on the way. I don't know whether we're doing the right thing, but it's too late now!

As I write this, I watch my four-year-old with severe hemophilia A and my unaffected two-year-old play together. With our second child, we obviously made the right choice, but you don't know if you're right or wrong until after the child is born. My 12-year-old cousin has six months to live due to terminal brain cancer; my neighbor accidentally ran over and killed his two-year-old son last summer. None of this–including hemophilia–makes any sense to me.

DOUG STEWART
Michigan

[A] I see no real difference between having a girl and having a boy. If you're a carrier, you risk passing the gene to your children–and either your son will have hemophilia, or your daughter's children may have hemophilia.

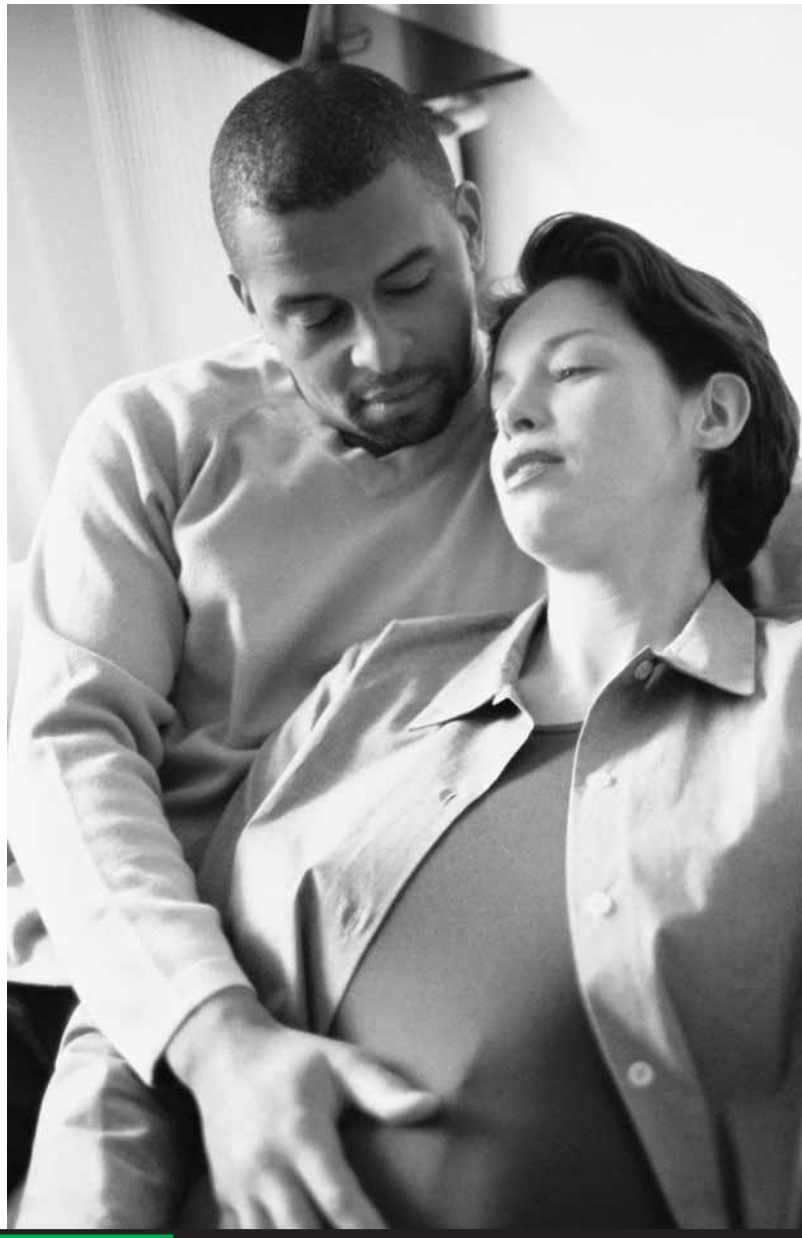
With so much new technology at our disposal, we're faced with making tough decisions about what we will pass on to our kids. God gives us what he chooses, and we are called on to love and care for our children. I don't pretend to say whether it's responsible to knowingly bring children into the world with a bleeding disorder, but it deserves great consideration. When we have children, we must commit to caring for them, regardless of whether they have bleeding disorders, Downs Syndrome or other chronic disorders.

My husband and I have decided not to have more children because we feel that we have an awesome responsibility: to use the knowledge we possess both in planning for the well-being of a child to come, and in meeting our commitment to the children we have.

We were blessed to have a daughter first, and planned our second birth without the conflict of knowing that our second child could have hemophilia or be a carrier. Now that we do know, it has changed everything. Knowing our own limits and capabilities, we assessed our goals for our family–specifically our son–and decided not to have more children.

Since our son has done well, we manage his disorder mainly by watching him closely. Although we haven't done prophylaxis, that takes one-on-one supervision, which we couldn't do for two children. We don't know yet whether our daughter is a carrier, but we'd be concerned to put that responsibility on a daughter, knowing that as a parent, she may face the same challenges we have faced.

It wasn't an easy decision, but it was best for our family. Would I trade my son for a child without hemophilia? *No!* Would I have terminated my



pregnancy had I known that he had hemophilia? *No!* Would I terminate a future pregnancy knowing that the child had hemophilia? *No!* But making the decision to intentionally take the chance of having child with a lifelong disorder–that's another story.

I encourage anyone trying to make this decision to weigh the short- and long-term realities, benefits and fears. We may know how we feel about passing hemophilia to future generations; but we have no way of knowing the feelings of those who will live with it. If I had another son, he might be happy to be alive, taking his disorder in stride. A different child might be angry about having to suffer the complications that hemophilia may bring.

But we won't hear from the son we might have had. Maybe we aren't brave enough. Maybe we're wise. We'll never know. We have to live with the choices we make–for ourselves as individuals, for our children, and for generations to come.

Gina Cindrich
California

[A] My husband and I both wanted more children, but decided not to have another child. We still wonder if our decision is the right one. I don't feel brave and I don't feel wise. We're just doing the right thing for us. I don't wish to tell anyone else what to do.

My son is one of the healthiest, sturdiest boys with hemophilia that I know. But after everything that he has experienced–first bleed, tests and assays, first infusion, port implantation, hospital stays, agonizing port accessing, port infections, second port implantation, constant attentiveness to everything he does, school, sports and explanations, peripheral sticks–neither my husband nor I wish to put another child through any of it. When my son says, "I'm really tired of the stickings," and Dad responds, "Well kiddo, it just has to happen," our collective hearts break. Would I have aborted my son if I knew that he had hemophilia? *Absolutely not!* Would I love to have another child? *Absolutely yes!* Will I purposely attempt to get pregnant knowing that I'm a carrier and my child will have a 50% chance of dealing with hemophilia in some form? *No*. I don't think that's fair to a child. Would I abort a second child knowing that he has hemophilia? *Absolutely not.* It's a conundrum.

Kathy Mackay
Georgia

[A] After our son Stephen was diagnosed with severe hemophilia A, my husband and I decided to have no more children. We wanted more children, but after four difficult pregnancies, constant morning sickness, high blood pressure, bed rest for two months, and two miscarriages in the second trimester, I just couldn't go through it again. And we didn't want to risk having another child with hemophilia or a daughter who is a carrier. Our decision still hurts sometimes, but it's the best decision for us.

Leslie Houvenagle
Kentucky

[A] When our first child, Sam, was diagnosed with hemophilia, we assumed that we would have no more children because hemophilia seemed overwhelming. Then, 14 months later, Sam's port was placed and we began prophylaxis... and our lives changed. Hemophilia no longer controlled us. Four months later, we began to consider having a second child. We certainly didn't *hope* to have one with hemophilia.

As we weighed the option of adding to our family, we wondered whether Sam would think that something was "wrong" with him if he learned that hemophilia was the reason he didn't have a sibling. Our limited financial resources made adoption virtually impossible. Then we considered trying to conceive.

When I became pregnant with Nat, we chose not to have genetic testing, but did learn that we were having another boy. I went through the grieving process again—we just assumed that he'd have hemophilia.

Six years later, we have no regrets. Nat, age six, and Sam, age eight, are best friends. They don't feel isolated or "strange" because they have hemophilia. Each boy has a companion who intimately understands what it's like to live with a bleeding disorder.

Fortunately, our experience with hemophilia has been smooth. We've had three ports, no infections, no joint bleeds, no breakthrough bleeds. Both boys participate fully in activities that interest them. Both boys are candid with friends and classmates about their medical bracelets and weekly Band-Aids® on their elbows. We're lucky to have good insurance.

For our family, it came down to "the devil you know" versus "the devil you don't." Our experience living with hemophilia has strengthened our family. Our marriage is solid, and the early adversity we faced has deepened our commitment to each other and to our children. Still, if I had watched my family deal with the devastating effects of AIDS, and life before recombinant factor, I'm fairly certain that my thoughts would have been different.

JILL LATHROP
Wisconsin

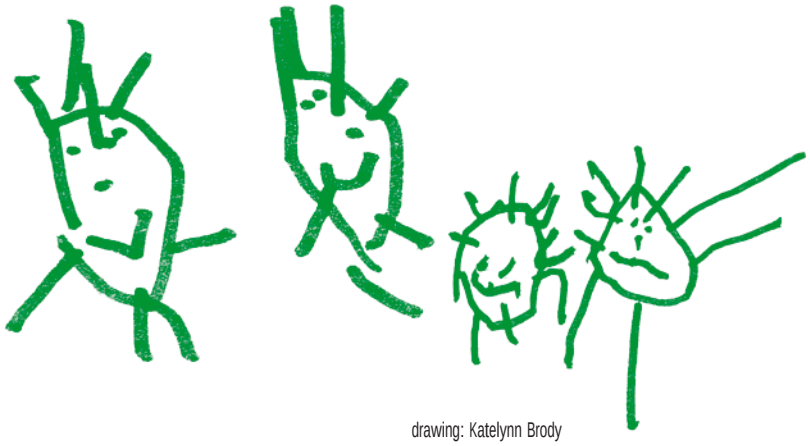
[A] I have four children: Alex, who is eight and has hemophilia; Julia, who is five; and Nathan and Owen, identical three-year-old twins with hemophilia. Our twins were a surprise, although we did plan for our second child, Julia.

I grew up with an uncle and brother with hemophilia. While I was pregnant with Alex, I learned that he had hemophilia. I was devastated. But after Alex was born and we started prophylaxis at 18 months, my attitude changed. It just wasn't a big burden. It was manageable. So we decided that we would try for girl, but a boy would be no problem. With this pregnancy, we tried gender selection twice, but it became too expensive. Although we knew that we had a good chance of having another boy with hemophilia, we eventually got lucky on our own and Julia was born.

Hemophilia has affected Alex so little—he doesn't seem to think there's anything different about him. He doesn't mind the sticks, and even enjoys going to the HTC. He plays T-ball, soccer and basketball, and loves life and his friends. Even with bumps in the road—we've had insurance problems, and one of the twins suffered a cerebral hemorrhage at age ten months—we have been lucky.

If I could order a custom-made child, I would not choose one with hemophilia. Yet I wouldn't give up the joy of having my boys in my life. We can't control everything in our lives, and I believe that hemophilia has made us stronger.

MICHELLE BLOODWORTH
Illinois



drawing: Katelynn Brody

The information provided in Parent-to-Parent should **not** be construed as medical advice. It is advice from one parent to another. Please consult your HTC for information on any medically related questions.

Seven Reasons... continued from cover

New Jersey, describes the port as "a small cup made out of metal (called a *portal*), covered with silicone and attached to a tube." The thin and flexible tube, called a *catheter*, is attached to a large vein in the patient's chest. The port is about the size of "four stacked nickels," says Dr. Bergman.

The surgical procedure is virtually risk-free, according to Dr. Bergman. "The biggest risks," he notes, "are long-term." These risks include the possibility of infection and clotting. Dr. Bergman, who has performed about 300 of these procedures in his career, explains that performing the surgery in small children requires general anesthesia and is usually an outpatient procedure. Patients often return home on the same day as the surgery, although some are kept overnight.

Dr. Steven Halpern, a pediatric hematologist/oncologist at Tomorrow's Children's Institute at Hackensack University Medical Center in Hackensack, New Jersey, treats about 40 patients with hemophilia annually. "Ports have been around for over twenty years," he says, adding that ports were "originally for patients with cancer." Dr. Halpern explains that about a decade ago, physicians got the idea to use ports in patients with hemophilia.

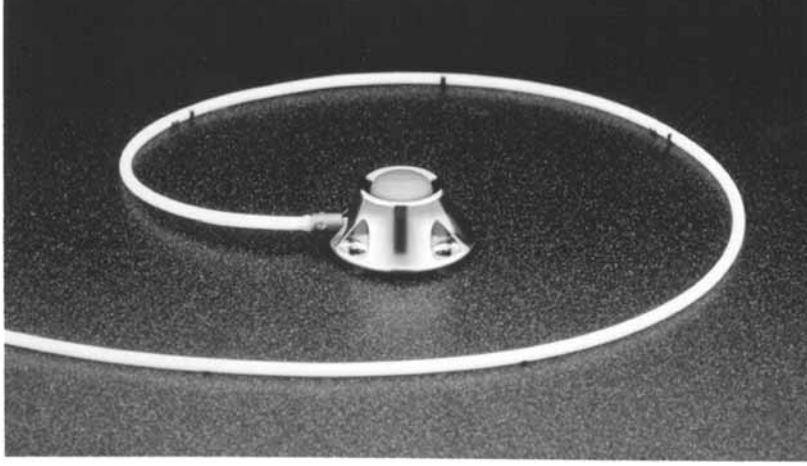
Today, cancer patients still represent the majority of patients using ports. Nevertheless, according to Dr. Halpern, ports have proved to be a "terrific advance in medicine and have made prophylaxis possible for children with hemophilia." He adds, "Ports allow them to live normal lives." In other words, ports allow patients to receive regularly scheduled infusions of clotting factor to *prevent bleeds* without having to access a vein. Accessing a vein is often troublesome, particularly in very young children.

All Good Things Must End?

The many advantages of ports include regular treatments, fewer joint bleeds and ease of infusion. But for children with hemophilia, eventually the port must be removed. Parents often worry about making the transition from port to vein because they—or their child—will need to learn how to infuse a vein. For children who can't remember life without a port, this change can be unsettling since children often think of the port as part of their bodies, and must now get used to repeated needlesticks.

For some children, medical circumstances may dictate removal of the port. Among the most serious reasons for removal are dangerous infections and blood clots. Other reasons to remove a port include outgrowing the port, ending prophylactic treatment, insurance concerns, and moving past the crisis that precipitated implanting the port. Parents may also choose to have their child's port removed simply b

photo: Smiths Medical MD, Inc.



A port: About the size of "four stacked nickels."

because the child is physically and emotionally ready for the change. Here are seven common reasons to remove a port:

1 SERIOUS OR FREQUENT INFECTIONS

A port allows direct access to a child's bloodstream—for factor or bacteria. Parents of children with hemophilia are taught a specific procedure to administer factor. Correctly following this procedure greatly reduces the risk of infection. Typically, a home healthcare provider teaches the procedure and works with parents until they are comfortable doing it alone. Parents must "practice rigorous precautions when accessing the port," explains Neala S. Schwartzberg, Ph.D., author of the article "Venous Access Devices—A Better Way?"[1] Dr. Schwartzberg writes, "Preliminary hand-washing with antibacterial soap is a must, as is cleaning the child's skin with an antiseptic and then again with an alcohol wipe according to instructions."

To help prevent port infections, parents must learn the proper procedure and follow it precisely. "Bacteria like to run to artificial devices," explains Dr. Bergman. He emphasizes that aggressive sterile technique is required at the port site, and with the factor bottle. However, remarks Dr. Halpern, "a very meticulous parent is not likely to have a problem." He adds, "Sterile technique is not usually the issue… Infections can occur despite meticulous attention to sterile technique." Of course, serious bloodstream infections, called *sepsis* or *bacteremia*, require immediate medical attention.

Parents of children with ports must report to their healthcare professional any fever of over 100 degrees. Then, says Dr. Bergman, "A blood culture would be done to check for bacteria." If an infection is confirmed, a seven- to ten-day course of antibiotics would be used. The antibiotics would be given at the port site. Dr. Bergman says that antibiotics are often the key to getting the infection under control. If the drugs don't work, the port must be removed. A minimum of a few days should pass after the port is removed to be sure the infection is cleared up before implanting a new port.

[1] *Hemalog*, July 1997.

According to Dr. Bergman, "Port removal for infection is tailored to the individual. Repeated, separate infections are simply bad luck, but if the patient relapses shortly after [antibiotic] therapy is completed, then the port was never sterilized." He adds, "A more aggressive antibiotic regimen might be tried, but failure will require removal [of the port]." Whether a patient will eventually have a new port implanted depends entirely on the individual's case history and medical needs. It's also interesting, notes Dr. Bergman, that "Just having one or two infections doesn't mean the patient is susceptible [to infections], but might mean someone accessing the port is breaking sterile technique."

While these port infections are typically curable, parents should know that a bloodstream infection is serious and could even prove fatal. Swift and aggressive action is required by parents and healthcare professionals in the event of an infection.

2 ENDING PROPHYLAXIS

"An increasing number of families turning to prophylaxis for their children have opted to infuse through a port system," writes Dr. Schwartzberg. "Ports and prophylaxis are a powerful combination. With prophylaxis, children who have hemophilia can act and feel like their friends, pursuing the same activities without worry about a bleed from a minor injury, or the embarrassment of an external line pointing out their medical condition."

Using a port allows factor continuous access to a child's veins, and protects veins from scarring. Children can lead active lives–running, playing, even swimming. At some point, however, parents may consider ending prophylaxis. Perhaps their child is old enough to cooperate with needlesticks. An older child's veins are usually developed enough that infusions can be rotated among various veins without damage. A child no longer using prophylaxis may have little reason to keep a port. If a patient is switching to on-demand treatment, the number of needlesticks will be greatly reduced.

3 THROMBOSIS

The port, with its catheter inserted into a vein, is a foreign object. Whenever a foreign object is inserted into the human body, thrombosis is more likely to occur. Dr. Halpern explains: "Catheters as foreign objects damage blood vessels and disrupt blood flow. The body forms a small blood clot to repair the damage, and this may eventually lead to a larger blood clot. Part of this larger blood clot can break off and travel to the rest of the body." The results can be deadly. A blood clot could travel to the lung, making the heart beat harder and impairing breathing. Of course, since a hemophilia patient's blood doesn't clot properly, thrombosis is less likely than in other patients with ports. But ironically, even in a hemophilia patient, it's still possible for clots to form around the port and its tubing. To help prevent clotting, ports are flushed with each use. A blood thinner called *heparin* is pushed through the port and tubing to help prevent a clot from forming.

According to Dr. Bergman, a minor clot around the port area does not pose a threat to the patient. Doctors keep an eye on minor clots, but generally expect them to be absorbed into the body naturally. "Small clots near the catheter," says Dr. Bergman, "are of no significance, and could probably be seen in every patient if one were to look hard enough." However, he adds, "Major clotting would completely or nearly obstruct the vein that the catheter lies within, and may extend toward the heart." If a major blood clot is detected, a blood thinner might be administered to dissolve it. If the clot persists, the port would need to be removed.

4 OUTGROWING THE PORT

As a child grows, his veins grow. Because larger veins are easier to access, the need for a port diminishes. An older child is able to strengthen and care for his veins by doing hand exercises. He may also have grown responsible enough to self-infuse. And he may get a psychological boost by gaining some control over his health. Doctors agree that these are all valid reasons for removing a port.

Although Diane and Gary Horbacz of New Jersey have never had serious problems with their sons' ports, they don't see the need to use ports indefinitely. Matthew and Justin Horbacz both have severe hemophilia, and have had ports from infancy. The Horbacz family is aware of both the benefits and potential disadvantages of ports. Now that their older son Matthew is nine, Diane and Gary think it's time to begin making the transition from the port to venous access treatment.

Diane emphasizes that a "gentle transition" is key. She and Gary are working carefully with Matthew to help him make a gradual, uneventful transition with little stress and physical pain. Matthew still has his port, but Diane and Gary use it only if they aren't successful in accessing the vein. "The vein is harder," admits Diane. "You don't know if you'll get the stick." Diane, who fainted the first time she practiced needlesticks with an HTC nurse, isn't sure when Matthew's port will be removed. She reports that while Matthew wants to learn to infuse a vein, he isn't ready yet to make the switch.

There are other developmental reasons to consider removing a port. As a child grows, says Dr. Bergman, the "catheter tip may not be in the right place [to infuse it successfully]." He explains how this change occurs: "The tip of the catheter needs to be in a large

photo: Diane Horbacz

"Gentle transition" is key: Diane Horbacz recommends a gradual transfer to peripheral venous access.

vein near the heart. As a child grows, the distance between the insertion site and the heart increases, but the catheter remains a fixed length. So, with growth, the catheter 'pulls back' into a smaller vein, further from the heart. If [the catheter] is too peripheral, it will need to be replaced." By the time that happens, it may no longer be such a big deal to infuse a vein. After all, an older child usually doesn't mind the needlesticks, while a small child may. An older child's veins can be located with greater ease, and can withstand repeated needlesticks without scarring or a lot of stress.

Although most children are not self-conscious about the small bump on the chest caused by a port, as they get older this may become a larger issue. Looking "different" from other kids–at the beach for instance–can be a real concern. Kristin notes that Lynden is very thin and his port was quite visible. Although this didn't bother Lynden, Kristin worried that in the future, it may have made him self-conscious. Looking just like peers, and avoiding differences due to health challenges, are paramount concerns for adolescents.

5 A CRISIS PASSED

The port was a lifesaver during a crisis for the Prior family. Lynden was diagnosed with hemophilia at age eleven months. During the first two years after diagnosis, the Priors treated Lynden's bleeds on demand. But his right knee proved to be a target joint with frequent bleeds. The Priors' local HTC, says Kristin, taught them how to "hit a vein and to do it ourselves." For the next three months, prophylactic treatments went well and Lynden received factor every third day.

"Once he was on [prophylaxis], he ate and slept better," explains Kristin. "His energy increased and our family was less stressed… We were not willing to go back on demand." But this easy transition was short-lived. "Suddenly," recalls Kristin, "we were having a horrible time–no one could find a vein." Inserting a port was the answer to the Priors' problem. However, after the November 2003 NHF meeting, the Priors were ready to move on. They decided to have the port removed after Lynden's crisis had passed.

6 INSURANCE CONCERNS

The majority of hemophilia patients in the US have a lifetime maximum on insurance reimbursement for prophylactic treatment. The NHF estimates that the cost of healthcare for a child with hemophilia is approximately $100,000 annually. A lifetime maximum may cover prophylactic treatment while the child is young, but may run out as the child gets older. Many individuals with hemophilia will exceed their lifetime caps within five to ten years of making claims, according to research done by the NHF.

The need to reduce insurance costs and prolong lifetime maximums, or the absence of insurance are both valid non-medical reasons for considering port removal. The patient can switch from prophylaxis to infusing the vein with factor as needed. The result is likely to be less costly and will still meet the patient's needs, provided there are no complications with the patient's treatment or medical condition.

7 SKIN BREAKDOWN

Occasionally, a patient with a port experiences a breakdown of the skin covering the device. According to Dr. Bergman, a minor breakdown of the skin is similar to an abrasion, and is often due to multiple needlesticks at the port site. "Minor breakdown is treated topically with antibiotic ointments," he explains, "and may necessitate avoiding port use for several days."

If the entire incision covering the port opens as a result of infection, the skin breakdown is considered more serious. This would typically happen within several days after port implantation. "An exposed port," says Dr. Bergman, "due to incisional breakdown must be removed." However, this kind of port complication rarely occurs.

Removing the Port

According to Dr. Bergman, the procedure to remove the port is straightforward. First, the surgeon reopens the old scar. The port will have scar tissue holding it in place, which the surgeon cuts away. The surgeon then pulls out the port and closes up the wound.

The Priors scheduled Lynden for port removal surgery within weeks of making their decision. The family practiced needlesticks on each other: if David or Kristin missed Lynden's vein on the first try, they

Tips for Parents Considering Port Removal

- Take time to understand the pros and cons of continuing to use the port.
- Work as a team to make a decision with your healthcare providers.
- Determine whether your child is emotionally and physically ready for port removal.
- Be well prepared for the transition from using the port to infusing a vein.
- Keep your child well informed about the transition from port to vein.
- Have a back-up plan in place when you need help infusing a vein.

went right to the port. Kristin considers EMLA® numbing cream on the needlestick site "a miracle." She's also learned that Lynden must be well hydrated for his veins to "plump up."

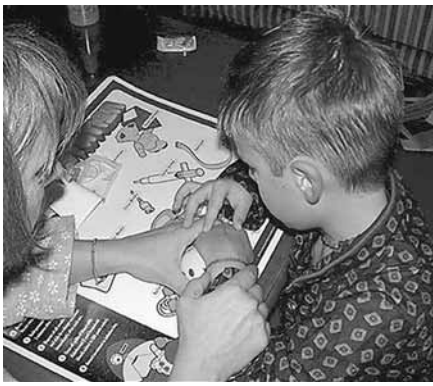
Lynden's port was removed in December 2003 in a 20-minute procedure. He stayed overnight in the hospital so the healthcare team could monitor his factor levels. To help close the wound initially, the sutures were located on the inside of his skin, with adhesive strips on the outside. Lynden has a small scar from the procedure, which Kristin calls "pretty uneventful." Six months later, Lynden is slowly learning to infuse himself, but his parents are not pressuring him.

Dr. Bergman stresses that port removal is a simple procedure. In fact, he often removes the ports of older children in his office using only a local anesthetic. Only a simple analgesic like Tylenol® is required afterward. The task of preparing the child for port removal, and the subsequent venous infusions, generally falls to the parents and home healthcare team or HTC.

Planning, Transition, Teamwork

The transition from using a port to infusing a vein can be stressful, but parents should remember that help is available at their HTC. Diane Horbacz believes that all children need to be assisted through this period of change. Parents should communicate with their children about the transition, and stress the importance of learning to access a vein. As with any kind of change, involving the concerned parties at every step helps ensure success.

Deciding to remove the port requires planning. Parents should carefully weigh the pros and cons of using a port. Kristin Prior encourages parents to be fully prepared: not only must they learn to infuse through the vein, but they should have a back-up plan in case they are unsuccessful one day. Parents may need to go to the emergency room or HTC for venous infusion if they fail to get a vein. Coordinating this back-up plan with the local HTC, says Kristin, is especially important.



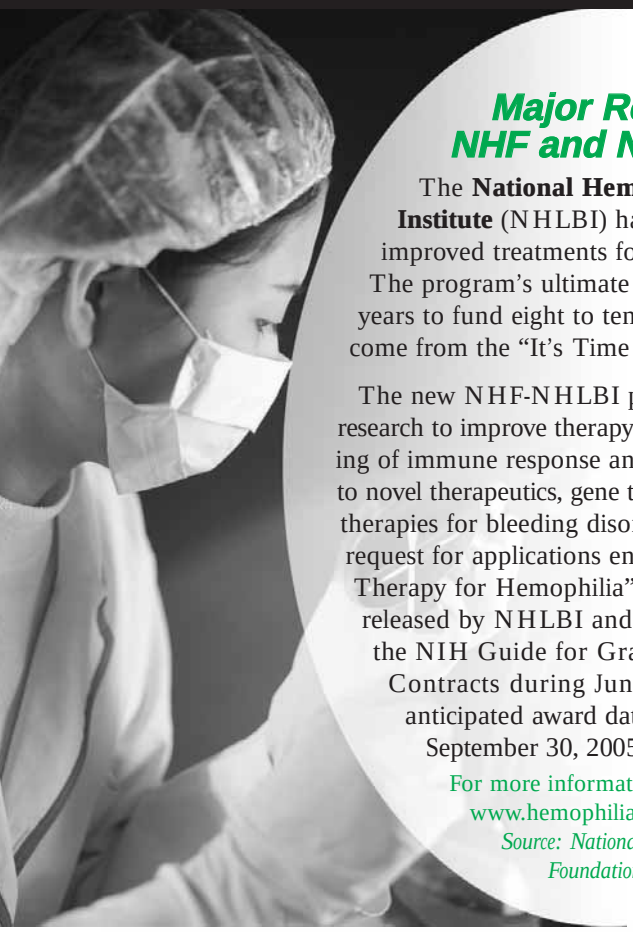
Life after a port: Lynden learns about self-treatment.

Ports have proved a wonderful addition to the treatment plan for many children. Most physicians and parents find more reasons to use the port than not use it. But for most families, eventually it makes sense to remove the device. Each individual case requires planning and a transition phase. Patience—and help from your HTC—will combine for good teamwork.

photos: the Prior family

When the time was right: Lynden Prior (and friend) ready for port removal surgery.

Denise D. Vermeulen is a freelance writer and former public relations director at two pediatric hospitals. She lives in Massachusetts with her husband and three children.



Major Research Venture for NHF and NHLBI

The **National Hemophilia Foundation** (NHF) and the **National Heart, Lung, and Blood Institute** (NHLBI) have agreed to jointly sponsor a collaborative research program that will lead to improved treatments for hemophilia, von Willebrand Disease and other hereditary bleeding disorders. The program's ultimate goal is to find a cure. NHF and NHLBI will commit $6 million each over four years to fund eight to ten research grants, from fiscal year 2006 to fiscal year 2009. NHF funds will come from the "It's Time for a Cure" campaign.

The new NHF-NHLBI program will stimulate basic research to improve therapy and enhance understanding of immune response and safety issues related to novel therapeutics, gene transfer or cell-based therapies for bleeding disorders. A one-time request for applications entitled "Improved Therapy for Hemophilia" will be released by NHLBI and published in the NIH Guide for Grants and Contracts during June 2004. The anticipated award date is September 30, 2005.

For more information visit www.hemophilia.org

Source: National Hemophilia Foundation

Hemophilia B Clinical Trial Discontinued

Biotechnology company **Avigen** is discontinuing its phase I trial of Coagulin-B, an investigational gene therapy technology to treat hemophilia B. According to Avigen, the halt is due to "certain scientific, regulatory and clinical hurdles" specific to hemophilia. Avigen plans to redirect its resources into product candidates addressing neurological disorders, including Parkinson's disease and chronic neuropathic pain. **Bayer HealthCare**, which partnered with Avigen to develop Coagulin-B, says that while it is disappointed, it will remain focused on research and development in hemophilia. Bayer will keep options open to continue supporting the Coagulin-B development program, and hopes that Avigen will resolve the current scientific issues.

For more information visit www.hemophilia.org or www.avigen.com

Sources: IBPN, *June 2004, National Hemophilia Foundation and Avigen*

Baxter Releases New Educational Program About Joints

Baxter Healthcare has developed a unique educational program called *Joint Health: A Guide for People with Hemophilia and Their Families*. The two-hour program explains how joints work, with illustrations of healthy and damaged joints and a detailed depiction of what happens during a joint bleed. The program stresses preventing joint damage through exercise, physical therapy, regular HTC visits, and adherence to factor regimens. *Joint Health* is available in CD-ROM and VHS formats. To order, visit www.hemophiliagalaxy.com and click on Joint Health Quiz, then complete the online ordering form. Or call Baxter's Customer Service line at 1-800-423-2090.

For more information visit www.baxter.com

Source: IBPN, *June 2004 and Baxter International*

Baxter Discounts Price for Advate

Baxter International has cut the price of Advate by about 10%. Advate is the first and only recombinant factor VIII concentrate with no added human or animal plasma proteins or albumin in the cell culture process. The new price is now only slightly higher than that of Baxter's older product, Recombinate.

For more information visit www.hemophiliagalaxy.com

Source: IBPN, *June 2004*

HHS to Acquire HRA

Tennessee-based home care company **Hemophilia Health Services** (HHS), a subsidiary of Accredo Health, Inc., is acquiring New Jersey-based home care company **Hemophilia Resources of America** (HRA) and merging operations. HRA plans to complete the acquisition by the end of this summer, and expects the merger to strengthen the service capability of both companies. Currently, HRA does not anticipate a name change, service interruptions or staff changes.

For more information visit www.hrahemo.com or www.hemophiliahealth.com

Source: Hemophilia Resources of America

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Insights... continued from page 4

or obtain it as part of an employee benefits package. For families with medical coverage supplied by a large company or government employer, insurance may not be an issue. But many families are not so fortunate. For some, the lack of adequate medical insurance coupled with enormous medical bills may cost them their homes and life savings.

Families without medical insurance through an employer often must rely on state aid, such as high-risk health insurance plans. Hemophilia treatment is very expensive, and most people can't afford the premiums for individual medical coverage. Small companies that offer their employees medical insurance are in the same boat: the premiums to cover medical insurance for one or two boys with hemophilia are exorbitantly high, and this cost can't easily be spread over a small number of employees. As a result, small companies in this situation have resorted to several imperfect solutions: 1) The company may drop all employee medical coverage. 2) It may cut employee salaries and/or withholding pay increases to cover the high cost of insurance. 3) Occasionally, a company may terminate the employee responsible for the higher medical premiums. In these extreme cases, such terminations are usually made under some pretense in order to avoid a lawsuit.

Even families enjoying adequate medical coverage from a large company for one son with hemophilia may run into problems if they have another baby with hemophilia. Many health plans have "caps"–a maximum amount the company will spend on a particular individual for a particular condition. One million dollars, the most common cap, is usually sufficient to last five to ten years. But children with complications such as inhibitors may spend through this cap at more than twice the normal rate. What do parents do when they reach the cap? Some employers offer more than one insurance plan, and allow employees to switch plans once a year–enabling parents to move to a new plan and start over with a new cap. Or a child can sometimes be moved to the other spouse's health insurance plan. Unfortunately, some families don't have these options, so they must search for a new job with new insurance, or go on state aid. In many states, to qualify for aid, income must be low; this forces parents to spend down their savings and take lower-paying jobs to qualify. In rare instances, parents have even divorced so that one parent could qualify for state assistance for a child.

Family Attitudes and Beliefs

How a family perceives hemophilia is a strong indicator of whether parents will risk having children with hemophilia. Families with a history of hemophilia–and a history of negative experiences–may decide not to have children. They have probably seen fathers, brothers and other male relatives suffer from joint destruction, chronic pain, HIV and hepatitis. They refuse to risk having children who might suffer. Parents of children who suffer from complications of hemophilia, such as inhibitors, also may be less inclined to risk having another child with hemophilia.

Conversely, parents of children with good insurance and positive experiences are likely to feel that hemophilia is something they can handle. They may choose to risk having another child with hemophilia. This attitude has grown over the past decade as prophylaxis has become accepted in the US, and boys with hemophilia suffer from fewer complications.

Family beliefs and traditions also influence the decision to have another child. Families with strong religious beliefs sometimes simplify the decision-making process by placing their faith solely in God. Some believe that if they have a child with hemophilia, it's God's will. Others believe that God gives them only the challenges they can handle.

Family Dynamics

Having a child with hemophilia is incredibly challenging. For most families, stress peaks during the toddler stage–when their baby son first learns to stand and walk, constantly falls, and can't say that he has a bleed. Stress usually decreases as the child gets older. If you ask parents of a toddler with hemophilia whether they would risk having another child with hemophilia, many will say *No!* If you ask the same question five years later, when stress has decreased and memories have faded, some of these parents will say *Yes!*

Having a child with hemophilia affects family dynamics. Families cope with the stress of hemophilia in different ways. Because they are so afraid that their son will be injured, some families become overprotective, severely restricting the child's activities and unfortunately ratcheting up the stress level of the entire family. Other families become overly permissive, sometimes allowing their children to do things a child with hemophilia shouldn't do. Some families avoid disciplining their sons with hemophilia. Some fathers, who had envisioned their sons playing contact sports, may show displaced anger or extreme disappointment. They may withdraw from interactions with their sons, increasing the stress level of the family and harming the son's self-esteem. And it's not unusual for a father or mother to be "in denial" about their son's chronic medical condition, placing more stress on the family.

What about feelings of guilt? Many women without a family history of hemophilia feel guilty about passing the

hemophilia gene to their son–especially if their son suffers from complications. This guilt is needless–but what about mothers who already have a boy with hemophilia? Having a second child with hemophilia is no longer a matter of chance, but a conscious decision. How will the mother feel if the second boy suffers from complications due to hemophilia? And how will other children in the family react to having another sibling with a medical disorder?

It takes a strong family to weather the stress of having a child with hemophilia. Stress affects each family differently: some families report that the stress of raising a boy with hemophilia brought them together and made them stronger. For other families, extreme stress widens tiny rifts in relationships, forming chasms that split the family apart. How a family deals with the stress of hemophilia plays a vital part in the decision to have more children. For some families worried about the challenges of having another boy with hemophilia, adoption has proved a viable option.

Gender Selection

What about families that want to have children, don't want to adopt, and don't want a child with hemophilia? Some families opt for gender selection to avoid having a boy with hemophilia. The term *gender selection* includes a broad array of technologies that can increase the chance of having a child of a particular sex. Some of these technologies are only moderately successful, while others are highly successful. Gender selection may employ natural methods, such as carefully timing when fertilization occurs, sperm sorting, and *in vitro fertilization* (*IVF*). IVF means fertilizing an egg outside the uterus, involves a small risk to the mother, and is often used in conjunction with *preimplantation genetic diagnosis* (*PGD*). PGD can be used to determine not only the sex of the child, but whether the child has a particular genetic defect. PGD can be used to select a boy who does not have hemophilia. Currently, IVF and PGD are still controversial, expensive, and not often covered by medical insurance.

While gender selection aims to select a sex or prevent transmission of a disorder, many families opt instead for prenatal genetic testing. Tests such as *amniocentesis* and *chorionic villus sampling* are used to diagnose hemophilia in the uterus after conception. Although knowing whether an unborn child has hemophilia may be helpful to the physician delivering the baby, it can also be used in making the decision to terminate a pregnancy based on a hemophilia diagnosis.

Consider Thoroughly, Then Decide Carefully

For families with hemophilia, the decision to have another child is rarely an easy one. Yet the time spent considering the options, and taking into account how another child with hemophilia will affect the family, is time well spent. Families that openly share their feelings and consider all ramifications are more likely to confront hemophilia with a positive attitude–that not only lowers the stress level of the family, but results in a well-adjusted boy with positive self esteem.

News Notes... continued from page 16

New! Handheld Electronic Patient Diary

Bayer Biological Products has developed EZ-Log, a unique handheld electronic diary that offers patients a new way to record and communicate accurate treatment and bleeding information. EZ-Log lets patients record home treatments and bleeding episodes, track product supply, and submit refill requests. Features include user-friendly menus and bar code scanners. Information from EZ-Log can be transmitted securely over phone lines to the patient's HTC, where it can be viewed over the Internet or downloaded into an existing clinical database. Only the patient's HTC has access to the data.

For more information contact your local HTC or visit www.bayer.com
Source: Bayer HealthCare

Bayer Donates Kogenate® FS to WFH

Bayer HealthCare LLC, Biological Products Division (Bayer BP), will donate more than 12.2 million units of Kogenate® FS to the World Federation of Hemophilia (WFH) for distribution in developing countries. The donated product has an approximate market value of $9.3 million US, and will be used to assist the WFH with hemophilia care in 27 countries. The product will be used in emergency lifesaving situations, or for patients awaiting necessary surgical and dental procedures.

For more information visit www.bayer.com
Source: Bayer HealthCare

Breakthrough Clinical Trial Underway

Opperbas Holding BV is conducting a phase II clinical trial using its proprietary PEGlated liposomes to prolong the half-life and hemostatic efficacy of recombinant factor VIII. This technology could potentially reduce the number of infusions needed by hemophilia A patients by 33%, and enable more patients to use prophylactic treatment.

For more information visit www.hemophilia.org
Sources: Opperbas Holding BV and National Hemophilia Foundation

Letters... continued from page 2

I SINCERELY APPRECIATED the information on potential bleeding problems in women who are hemophilia carriers. ["What Your Doctor Doesn't Know Might Hurt You," *PEN*, May 2004] I was diagnosed as a symptomatic carrier. I always run a lower-than-normal factor VIII level. This led to many problems throughout my life–but I wasn't diagnosed until the mid-1970s, when I participated in a study conducted by Dr. Oscar Ratnoff at the Cleveland Clinic; and after I gave birth to a son with severe factor VIII deficiency. I am now a registered patient at our local HTC, and receive treatment with Stimate™ or factor VIII concentrates when necessary. If I had been diagnosed and treated properly in my youth, I would not have encountered situations leading to high-risk bleeding episodes, including a tonsillectomy that almost resulted in my death at age four. Another episode was an unnecessary hysterectomy–which I might have avoided by using Stimate every month.

What's interesting is that not every carrier of severe factor VIII has the same problems. Several of my female relatives, who are known carriers, do not experience levels low enough to need treatment; and several have problems needing intervention. I've always wondered how this could happen, and the information in *PEN* was enlightening.

I'm now in my mid-fifties, and experience much discomfort with arthritic conditions in my knees, lower back, shoulders and hips. Could this be related to lower-than-normal clotting factor levels? Do other symptomatic carriers experience this problem later in life? I recall from biographies that Queen Victoria of England and Empress Alexandra of Russia had back pain and took bed rest often.

Women who are symptomatic carriers have clinically mild hemophilia. Do people with mild hemophilia have a higher incidence of arthritic conditions later in life? If this is true, then perhaps they should receive treatment [with Stimate or factor concentrate] for traumatic episodes to the joints. When I broke my leg, and later my shoulder, I never received treatment–maybe I should have, although there was no obvious swelling. Perhaps my question could result in a survey or study.

PRISCILLA OREN
Pennsylvania

PAUL CLEMENT *replies:*

THERE ARE DIFFERENT FORMS of arthritis, but the type that you describe is probably the most common: "osteoarthritis" or "degenerative joint disease" (DJD). The pain associated with this type of arthritis is caused by damage to, or loss of, cartilage on the ends of the bone in the joint.

Osteoarthritis is usually the result of aging, and there may be a genetic link. It can also develop because of injury, such as broken bones; intense physical activity, as in athletics; or a defect in the protein that composes the cartilage.

People with bleeding disorders who suffer from joint bleeds are especially susceptible to osteoarthritis because every joint bleed results in some damage to the cartilage. People with severe hemophilia who have repeated bleeds into the same joint, and are not adequately treated, may also develop a form of arthritis called "hemophilic arthropathy." This kind of arthritis involves an inflammatory response that resembles another kind of arthritis, the autoimmune disease "rheumatoid arthritis."

Do people with mild hemophilia have a higher incidence of arthritic conditions later in life? With our current knowledge, we can't make this generalization because the term "mild hemophilia" encompasses such a wide variation in factor levels. However, it may be safe to say that the more joint bleeds you have, the greater your chance of having arthritic complications later in life.

As a symptomatic carrier, you should be treated with Stimate or factor if you suffer joint trauma or broken bones. This will help prevent immediate complications due to bleeding, and long-term complications such as arthritis.

Visit your doctor: joint pain can be caused by other ailments, such as bursitis, tendonitis or peripheral nerve compression. Your doctor can suggest treatment options to control pain. In addition, several alternative therapies exist: many people report that products containing glucosamine help reduce the pain associated with arthritis. But be aware that many alternative therapies have no proven benefits.

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