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Parent Empowerment Newsletter

Teaching Your Preschooler about Hemophilia

by Laureen A. Kelley

Teaching children about hemophilia builds strong family bonds.

A father of a child with hemophilia once asked me, "What should we say when our son asks where his hemophilia came from?" He suggested telling his four-year-old that God gave him his hemophilia. Why? "Because we sure don't want him to think we gave it to him!"

Your preschooler with hemophilia may go from *Sesame Street* and playtime to hospitals, pain and shots–all in one day. It's an honor, duty, and a feeling of deep satisfaction for parents to offer loving comfort and emotional support during trying times of hemophilia. Yet it's daunting to try to help children make sense of hemophilia. Should we only comfort our children? Should we explain hemophilia? *How* do we explain it to so young a child?

You can offer solace during difficult times, but you can also use infusions, hospital visits and comfort time as opportunities to learn how your child understands his world–and then, to better explain what's happening to him.

Think of teaching your child this way: when you first learned he had hemophilia, you may have been scared, overwhelmed, and emotional. You wanted and needed comfort. After a time, however, you were ready to learn more about this disorder, and craved information. It might have been scary if a doctor had taught you a full "Hemophilia 101" on the first day of clinic. On the other hand, it may have been frustrating *not* to understand at all what was going on with your son, after several clinic visits. Somewhere in between, knowledge and emotional support go hand in hand, to make you feel you have

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inside PEN

The Appletons: America's "First Family" With Hemophilia?

by Richard J. Atwood and Sara P. Evangelos

New England, 1639. Imagine that you are standing on the deck of the sailing ship *Jonathan*. You have just glimpsed the shore of your new home, the Massachusetts Bay Colony. Imagine the brilliant New England foliage, the bright chilly wind. Imagine your dream of farming your newly acquired land. Imagine the adventure. Now, imagine that you are the first European with hemophilia to step on the North American shore.

John Oliver (1613–1642) traveled from Bristol, England with his family to settle under the leadership of the Massachusetts Bay Company. He lived for only three years after he reached North America, fathering one child, Mary, and dying young as a consequence of his hemophilia. Not until after 1800 did the medical community begin using the term *hemophilia* to describe his disorder.

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welcome

As the U.S. holiday of Thanksgiving approaches, our thoughts and activities often turn to honoring those who first came to the New World; those who suffered its harsh winters, and who survived to create the cities and country we now enjoy. Living in Massachusetts, birthplace of America and home of the first Thanksgiving in 1621, has always had special significance for me. Imagine my surprise when I learned that the first person with hemophilia diagnosed in the U.S. was born in 1677 (one year after the first "official" Thanksgiving) and lived in Massachusetts–about 15 minutes from my home! In this issue of *PEN*, we tell you about the colonist Oliver Appleton. His story will remind you of the character, fortitude and courage required to succeed in the New World, in spite of an untreated chronic disorder.

Learn more about this fascinating man and his family with the information provided. Then tell his story to your children with hemophilia, as you teach them about the Pilgrims and Native Americans. But first, read our feature article: Learn how to improve communication about hemophilia with your preschooler, while building a deeper relationship with your children through learning and stories. Then start a dialogue–about hemophilia, Thanksgiving, or any other subject your family can share.

Happy Holidays!

[Visit www.wilstar.com/holidays/thasnkstr.htm to learn more about the first Thanksgiving]

letters

We received with pleasure the August issue of *PEN*. Thank you from the Society, and from the hemophilia community of Pakistan. The new design and style of *PEN* is beautiful. We appreciate the efforts of you and your team. It certainly is the magazine "by families and for families."

Saaed Ahmed
The Pakistan Hemophilia Patients Welfare Society,
Karachi, Pakistan

Thanks so much for sending a PDF version of Dick Lipton's article ["Theme Park Summer Fun," *PEN*, August 2002]. It's been a very "interesting" summer, and this article is great for sharing!

Danna Merritt, MSW, CSW
Hemostasis Thrombosis Center,
Children's Hospital of Michigan

PARENT EMPOWERMENT NEWSLETTER NOVEMBER 2002

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PEN is a newsletter for families affected by bleeding disorders that is edited and produced by a parent of a child with hemophilia. It is an unbiased forum that promotes an active exchange of information and support among divergent groups in the national and international hemophilia community.

PEN does not promote individual products or companies, and will use brand product names and company names pertaining only to news and education.

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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!

PEN en Español

PEN is available biannually in a Spanish version, for all your Spanish-speaking clients and patients. Each issue contains an assortment of *PEN* articles and features. Help educate and empower the hemophilia community… in Spanish!

For a free subscription to *PEN en Español,* or to obtain bulk copies for your office or treatment center, contact Karen Gaudioso at karen@kelleycom.com.

by Ruth Andrea Seeler, MD

Good to the Last Drop!

Wastage Potential of Small Volume Factor Concentrates

Are you throwing away up to **25%** of your factor? This is the risk of using what I call "super concentrated factor concentrates–those with volumes of only 2.5 and 4.0 ml. You know them by the names ReFacto®, BeneFIX®, Kogenate® FS, and Helixate® FS.

Most members of the hemophilia community are used to infusing with 10cc diluent per bottle of factor; and using BD "scalp" (butterfly) needles and tubing. When you infuse, do you leave a little something in the tubing and bottle? If this extra is only about 1 ml, you've wasted only about 10% of your total volume of factor. However, if you use the smaller volumes (2.5 or 4.0 ml) found in the "super concentrated" factor concentrates, the *same* volume wastage will mean a profound percentage loss–25% to 40%!

"What concerned me was how much *wasn't* being injected! Why were we wasting so much?"

I first became concerned about factor wastage when the smaller volume factor concentrates were in development or new to the market. As a hemophilia camp "Doc" for 27 years, I've had the opportunity to observe first-hand how many teenagers and adult counselors do their own infusions. These "experienced infusers" were trained by numerous hemophilia treatment centers. What concerned me was how much *wasn't* being injected! Why were we wasting so much?

First, if a vein is blown, the infusion set is typically tossed and another set is used. If you use the BD infusion set, there is a tubing volume of 0.25 to 0.3 ml. If you use the Terumo® winged infusion set, there is a volume of 0.4 ml. When you throw away the infusion set, you may throw away between 0.3 and 0.4 ml of factor. If you use a second set to complete the infusion, this is also eventually tossed. Using two infusion sets results in a combined factor concentrate loss of 0.6 to 0.8 ml!

Second, consider the factor concentrate left behind in the bottle. Using TB syringes, I measured the residual volume in discarded factor bottles at camp. The residual volumes ranged from 0.2 to 0.5 ml, with most between 0.3 and 0.35 ml. This means a *total loss of 0.9 to 1.0 ccs.* This wastage wasn't due to any particular style of infusing; I observed at least 20 different campers, and *all* left factor behind in the discarded bottle.[1]

In response to this concern, some manufacturers have revised their package inserts to recommend use of the infusion set provided with the factor, which has a smaller volume of 0.15 ml. However, variations in tubing length, syringe adapter, needle size and needle grasper (the "butterfly") lead some with hemophilia to prefer the BD infusion sets to those supplied by the manufacturer.

How can we stop wasting expensive and valuable factor concentrate? Well, it's easy to correct part of this wastage. First, use the infusion sets provided with the 2.5 or 4.0 ml concentrates.[2] Second, be sure to get *all* of the factor out of the bottle by having the tip of the withdrawal needle just at the top of rubber stopper (not a millimeter or so higher!). Third, a little air (0.5 cc) in the infusion syringe would allow the factor to be infused to the "last drop" at the end of the needle.

Factor is expensive. Its cost has an impact on your insurance; sometimes on your wallet; and on insurance premiums of all Americans. Let's do our part to prevent waste of this life-giving medicine. Let's remember that *every drop is good*, and can be used.

Dr. Seeler is Professor of Pediatrics, Hematology and Oncology at the University of Illinois. In 1973 in Illinois, she founded the second hemophilia summer camp in the U.S. A dedicated fan of summer camps, Dr. Seeler has always stressed physical activity for patients. At age 66, she still competes in triathlons.

Crossing the finish line: Dr.Seeler inspires many people with hemophilia with her triathlons.

[1] I did not measure residual amounts in any bottles reconstituted and infused by experienced RNs and MDs.

[2] Baxter BioScience does not currently include infusion sets with its products. The new BAXJECT® device is designed to eliminate residual factor loss in the bottle.

by Paul Clement

Needlestick Hazard REDUCED!

Have you, or has your child, ever been stuck by a needle while infusing? Are you aware of the risks of being stuck with someone else's needle? An estimated 600,000 to 800,000 healthcare workers–primarily nurses–annually suffer needlestick injuries (NSIs) and other "through the skin" injuries. These injuries put workers at risk of contracting blood borne viruses.[1] As many as one-third of all sharps injuries are related to the disposal process. The Centers for Disease Control and Prevention (CDC) estimates that 62% to 88% of sharps injuries can be prevented by using safer medical devices.

Many more needlestick injuries occur to individuals in the home than to healthcare workers in the workplace.

In response to this situation, Congress passed into law the *Needlestick Safety and Prevention Act* on November 6, 2000. To meet the requirements of the act, OSHA revised its Bloodborne Pathogen Standard. The revisions recommend that employers select safer needle devices when available, and involve employees in identifying and choosing the devices.

Still, many more needlestick injuries occur to individuals in the home than to healthcare workers in the workplace. In a market survey of 78 patients with hemophilia A from two HTCs, 30% had stuck themselves with a transfer needle in the past, and 77% were concerned about transfer needlesticks.

Factor concentrate manufacturers know that needlesticks worry many parents. Companies have been researching safer, more convenient methods of delivering factor, such as self-contained syringes already loaded with factor and diluent. Yet until recently, people with hemophilia have seen few notable improvements in needlestick safety in the home. Since this summer, users of Baxter BioScience's products can enjoy a new level of needlestick safety with the introduction of the BAXJECT®[2] device, the first needless transfer device. The BAXJECT device replaces the metal double-ended transfer needle and the filter needle, thus eliminating needle sticks. In addition to being safer, using the BAXJECT device is faster than using needles because some of the steps in the reconstitution process are eliminated. The plastic spikes in the device are also perfectly positioned to allow withdrawal of *all* the factor in the bottle. This reduces wastage due to improper withdrawal of the factor–which often happens with a filter needle [see "As I See It", page 3].

Reconstituting factor using the BAXJECT device is simple:

1) Peel away the package lid.

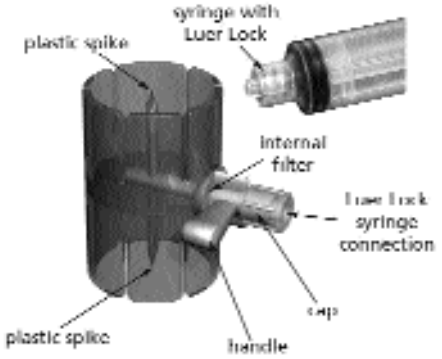
2) Push the BAXJECT device, with the plastic package still attached to one side, onto the cleaned diluent bottle.

3) Remove the plastic package.

4) Invert the unit (diluent bottle with BAXJECT device attached), and push it onto the cleaned factor concentrate bottle.

continued on next page

BAXJECT® Device Features



[1] According to the U.S. Department of Labor, Occupational Safety and Health Administration (OSHA)

[2] "BAXJECT" is a registered trademark of Baxter International, Inc.

Needlestick... continued from page 4

The diluent is automatically drawn into the factor bottle, and the factor is mixed. Then, just turn the handle down and attach a syringe. Inject 10 cc of air into the mixed factor, flip the system, and withdraw the factor with the syringe–you use no transfer or filter needle.

You can learn more from your HTC, or by visiting the Baxter website at www.hemophiliagalaxy.com/4_PRODUCTS/baxject/index.html.

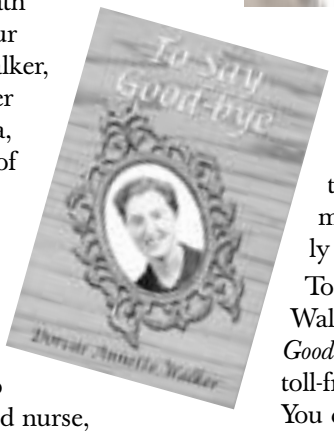
The downside? The BAXJECT device is available only if you use Baxter's products, like Recombinate® or Hemofil® M. If you use a different product, however, you should know that other manufacturers are always trying to create safer infusion delivery systems. Until that happens, be sure to follow standard procedures for safe infusions: wash carefully before infusing, don't reuse needles, always use a sharps disposal system, and clean up immediately after an infusion.

To Say Good-bye

A new book by Dorcas Walker, mother of a child with hemophilia

To Say Good-bye is a gripping, first-person account that begins with the paralyzing news that Dorcas Walker's mother, Margaret, is HIV positive.

The story ends with Margaret's death from AIDS four years later. Walker, whose daughter has hemophilia, tells the story of her efforts to support her mother's courageous fight against AIDS. Margaret, who was a registered nurse, contracted the HIV virus from her husband, who had severe hemophilia and died of AIDS in 1985. This is a tragic story of pain and despair. Only Walker's fierce love for her mother carried her through the last painful months, as she personally took care of Margaret.



To receive a copy of Ms. Walker's book, *To Say Good-bye*, call PublishAmerica toll-free at 1-877-333-7422. You can also order it online at www.publishamerica.com or write to PublishAmerica, Inc., P.O. Box 51, Frederick, MD 21705.

Thomas Joseph Fahey, Jr.
January 18, 1953 -
September 26, 2002

It is with a sad and heavy heart that I inform the hemophilia community of the death of Tom Fahey.

Tom was my dear friend and mentor, and cofounder of the Committee of Ten Thousand. COTT represents people with hemophilia and HIV, and was instrumental in achieving settlement payments to people infected with HIV in the U.S. Tom died Thursday, September 26 in Pittsburgh, awaiting a liver transplant with his family.

Tom leaves behind his wife of 22 years, Fran Fahey, a large and loving family, and hundreds of friends. Known as a sensitive and loving "teddy bear," Tom counseled and supported many within the HIV/AIDS world, and the hemophilia community. He was widely respected by the public health community in Massachusetts and nationwide. Tom was also involved with the global AIDS and hemophilia community. He was an activist who personally embodied COTT's credo, "action = life." He enjoyed birding, nature, music and good parties.

Tom will be deeply missed by many who were blessed to have known him and been touched by his life. No words will ever be adequate to pay tribute to such a fine friend and wonderful human being.

John P. Rider
Committee of Ten Thousand, National HIV/AIDS Advocate

Will CJD Donor Restrictions Affect Blood Supply?

Effective November 2002, the **American Red Cross** will adhere to the FDA's donor deferral guidelines, instead of its own more stringent criteria. Currently, Red Cross blood donors must defer blood donations if they spent three months or more in the U.K. from 1980 to the present. The FDA requires only the time frame 1980 to 1996. Why the switch? An "impending supply crisis" in its service area, says a representative of the New York Blood Center. *Source: International Blood Plasma News, Sept 02* [Visit www.kelleycom.com for more information.]

West Nile Virus: Threat to Blood Safety?

West Nile Virus (WNV) is a mosquito-borne disease never before reported in the Western hemisphere until 1999. It causes encephalitis (brain inflammation) and meningitis. This summer three people developed WNV infection from blood or blood products. The FDA and CDC have determined that organs, blood and blood products (packed cells or plasma) can transmit WNV. Statistically, your risk of contracting WNV is low. Most people who are infected have no symptoms. At highest risk are the elderly and people with weakened immune systems. WNV does not remain in the blood stream, as do HCV and HIV.

While no specific studies have been performed, the FDA calls the risk of WNV transmission through plasma derivatives (like factor concentrates) "extremely low." WNV is a lipid-enveloped virus, likely to be killed by viral inactivation processes. Yet one FDA official commented, "It is reasonable to do some serological surveillance in the hemophilia community." A WNV vaccine may be ready for human testing this fall. *Source: FDA & CDC* [Visit www.kelleycom.com or www.hemophilia.org for more information.]

ReFacto® Available in New Size

In response to consumer requests, **Wyeth BioPharma** has begun deliveries of ReFacto® Antihemophilic Factor (Recombinant) in a new 2,000 IU bottle, still with only 4 ml reconstituted volume. This larger size, the **first** in the industry, is especially convenient for large individuals, and those on Immune Tolerance Therapy, who require larger doses of factor concentrate.

New Recombinant Factor XIII in Trials

Avecia, a U.K. based company, has delivered the first batches of recombinant human factor XIII (rhFXIII) to **ZymoGenetics**. ZymoGenetics now hopes to file with the FDA to conduct a Phase I patient trial before the end of 2002. Current annual worldwide sales of factor XIII total about $35 million.

Source: International Blood Plasma News, Sept 02 [Visit www.kelleycom.com for more information.]

Gene Therapy Trial Reports Success

GenStar reports that the first hemophilia A patient treated with its MAX-AD Factor VIII gene therapy protocol has produced factor VIII at approximately a 1% level for seven months following treatment. There were side effects from the single treatment, which GenStar believes can be reduced with anti-inflammatory agents and encapsulation of the vector. A Phase I clinical trial will be initiated in Europe in the second half of 2002. *Source: www.genstar-rx.com/news/mainnews_frame.html*

College Step-by-Step

Make Yourself A Winning Candidate

The first step on the road to college is all about you: the courses you take during high school, the grades you earn, the activities you enjoy. It's never too early to start thinking about the future.

Here's what you need to do:

1. Start taking the right courses

The first thing colleges look at is your high school record. A strong college-prep course schedule should include the following mix of subjects, which means getting started in your freshman year.

- English: four years
- Science: three or four years (two lab sciences)
- Math: three or four years
- Foreign Language: two or three years
- Social Studies: three or four years

Learn more about…

- How to build a solid record
- Why you should take calculus
- Why honors courses do help

2. Build a solid transcript

What do admissions officers look for when they review your transcript?

- Grades: an upward trend
- High School: its academic quality
- Rank: in class or GPA
- Test Scores: SAT I, ACT, SAT II
- Senior Year: make it a solid one

Learn more about…

- Insider admissions strategies
- Upping the odds of an "in"
- Stay on top of your GPA

3. Choose your extracurricular activities carefully

Passion for extracurriculars must come from within. If you don't care about a particular sport or hobby, don't force it just to get into college. Consider these factors:

- Sports: gaining an admissions edge depends on the school, team, and make-up of the squad
- Talent: trilingual students, gifted musicians, and artists are always in demand
- Grades: a star athlete's grades and recommendations still count very heavily

4. Get to know yourself

A little introspection goes a long way in helping you make decisions about college. Talk with a counselor and take time to talk with yourself, too. Start by pondering the following questions:

- Why do I want to go to college?
- What is my learning style?
- How do I respond to pressure?
- What activities matter to me?
- What majors interest me?

Reported in *Newsline Eight & Nine*, Fall 2001. Reprinted from www.usnews.com/usnews/edu/college/stepbystep/index.htm

Tips: Don't worry!

Experts agree that students who plan their high school years *in advance*–and who decide carefully where to apply–will almost certainly be accepted by fine institutions.

An additional tip from *PEN*: Involvement in community service shows a well-rounded applicant, and balances a student who may otherwise appear self-occupied. Volunteering for clothing drives, fundraisers, and other charitable events demonstrates your compassion, leadership and teamwork.

The Appletons... continued from cover

John's daughter, Mary Oliver (1640–1698), was likely the first hemophilia carrier of European descent born in the colonies. With her husband, Major Samuel Appleton, Jr. (1625–1696), Mary had three daughters and five sons. One of these sons, Oliver Appleton (1677–1759), was the first American colonist born with hemophilia.

Beautiful vistas of Appleton Farms evoke the past: At right, a view of Farm Avenue, the main entrance to the farm. Below, looking toward the site of the "New House," a family homestead that stood from the 1880s to 1960s.

Photos: Appleton Farms

Early Ipswich Roots

Mary and Major Appleton lived in a settlement known to native Americans as Agawam, but re-christened by the English in 1633 as the town of Ipswich. What would life in Ipswich have offered their son, Oliver Appleton? Thirty miles north of Boston on the Atlantic shore, Ipswich was owned by the Massachusetts Bay Colony; it was purchased earlier in the century from Native Americans for 20 British pounds. By the mid-1600s, Ipswich ranked second only to Boston in population and wealth.

The Appletons were a wealthy colonial family. Major Samuel Appleton, Jr., Oliver's father, was the son of Samuel Appleton Sr., one of the "landed gentry," and a good friend of John Winthrop, the first governor of the Massachusetts Bay Colony. Appleton's fertile 460 acres of farmland had been granted to him by the Colony in 1638, and left to his son, Major Appleton, around 1670. Major Appleton, who served as a judge at the infamous Salem Witch Trials in 1692, died in 1696. He left his now nearly 600 acres, split into four parcels, to his four sons: Oliver, Isaac, Samuel and John. Oliver's 100-plus acre inheritance included his father's sawmill, ox pasture, and farmland bordering his brothers' parcels.

In 1701, Oliver married Sarah Perkins. Well-to-do millers, farmers and traders, Oliver and Sarah possessed numerous household and farm goods. They were involved in local politics, church affairs and business. Together they raised fourteen children; several sons and their descendants would become fine cabinetmakers.

At the turn of the eighteenth century, Oliver and his three brothers were working their adjoining farms in a loosely communal style. Each brother might grow a crop that the other brothers could use. Yet each brother farmed separately, produced his own goods for trade (like basket hoops), and kept his own business ledger. The brothers owned cattle, sheep, turkeys and hogs, and traded goods with family and friends in Ipswich.

A Dangerous Occupation?

To the British, the lure of Massachusetts Bay lay in its fertile farmland, forests and fish. The name "Agawam" refers to the abundant sturgeon, cod and salmon in Ipswich's tidal marshes. On their "new" land (already cleared and cultivated by Native Americans), the Appletons cut and milled timber, raised livestock and worked the farm. Today, farming is still one of the most dangerous occupations. In the seventeenth and eighteenth centuries, its hazards were surely compounded by Oliver's hemophilia, and the harsh New England winters.

Yet Oliver lived to be 82–a considerable age in any century. Imagine the taxing and dangerous farm chores, the lack of treatment. Was his hemophilia mild? What kind of treatment would he have received?

Unfortunately, few health records exist to shed any light. We know that Oliver suffered from "rheumatism," now called arthropathy, and probably used a rheumatism medicine. External bleeds were likely treated with styptics, such as bark tea. The Native Americans may have supplied their own styptics to the colonists: cobwebs, sassafras root, witch-hazel and eagle feather down. Some medicinal plants were brought from Europe, and grown in medicinal herb gardens next to the vegetable gardens. Internal bleeds were probably treated with sulphate of soda.

Late in life, Oliver was confined to his bed and developed bedsores on his hips. At age 82, his cause of death is recorded as bleeding from his bedsores and his urethra. Oliver appears to have been a generous and fair man, dividing his estate equitably among his children and his wife Sarah.

Making Medical History

Oliver and Sarah had six daughters and eight sons. Two of the daughters, Sarah and Hannah, had sons with hemophilia. Interestingly, Hannah's sons, Oliver and Thomas Swaim, were doctors. What would they have thought of their family's disorder? Without letters or other documents, we can only guess.

Yet it was the Swaim branch of the Appleton family that attracted the attention of the medical community. Based on his personal connection with the Swaim family, Dr. John Hay, a Massachusetts physician, published an article on the Appletons in a New

England medical journal in 1813. Following this publication, the Appleton family history appeared in numerous medical journals, at least as late as 1962. By then, the family had been traced through 350 years and 11 generations: 25 males with hemophilia, and 27 carrier females. In 1961 a blood sample, drawn from the last known living carrier in the family tree, revealed factor VIII deficiency, or hemophilia A.

"Jimmy's Barn:" A post and beam structure built c.1850, it was named in memory of James Appleton (1899-1915). Today, the barn serves as the distribution center for Appleton Farms' Community Supported Agriculture Program.

After 1800, a diagnosis of hemophilia was based on personal history of bleeds–prolonged bleeding from minor cuts, "rheumatism" in joints, deaths from bleeding. It was also based on family history over several generations. So in retrospective, Oliver is known to have hemophilia because of his bleeding history, the later reconstruction of the Appleton family history, and the 1961 blood sample.

The Appleton Legacy

For three centuries, generations of Appletons worked the farm in Ipswich. Homes and farm buildings were constructed on the land, then demolished, or destroyed by fire. But in 1901, an Appleton house built in 1794 was joined with an early farm building from Oliver's family, relocated across a road dating from 1637, and named "Applefield." It still stands today as a private residence, still flanked by the 1637 road.

The Appletons managed (despite several sales outside the family) to hold on to what is now nearly 1,000 acres. Today, the farm is a patchwork of rolling fields, meadows, pastures, stone walls and tree-lined lanes. White Park cattle, a British breed that Oliver and Sarah might have owned, graze the fields. Thanks to gifts, in 1970 and 1998, from Colonel Francis R. Appleton, Jr. and his wife Joan, the farm was donated to the state of Massachusetts. It is now owned and managed by the Trustees of Reservations, and is open to the public.

Today, when you experience the peace and beauty of Appleton Farms, then stand in the center of Ipswich, you can almost believe that you have returned to Oliver Appleton's era. The cemetery, where many Appleton descendants are buried, graces a hillside. The library, town hall, and historic buildings all speak of earlier times.

Are the Appletons America's "First Family" with hemophilia? Perhaps, in the sense that our knowledge of hemophilia has been enriched by the study of this large and long-lived colonial family. Thanks to our American Revolution, we have no "royal family" with hemophilia. Yet we can still honor and remember the Appleton family. This Thanksgiving, we can recall the challenges faced by earlier generations with hemophilia–people who contributed to our heritage as Americans, and as a hemophilia community. To understand ourselves, and create our vision for the future, we must always remember the past.

***Richard J. Atwood MA, MPH**, works at Wake Forest University School of Medicine in Winston-Salem, North Carolina. He is Regional Coordinator for the federal grants in Region IV North. He has worked at the Wake Forest HTC for sixteen years.*

***Sara P. Evangelos** is a freelance writer, editor and poet. She is editor for LA Kelley Communications, Inc., and a frequent contributor to* PEN *and* Hemophilia Leader.

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Teaching... continued from cover

control, and are capable of handling this disorder.

Like you, your child also needs comfort, and eventually information. But you don't want to give *too* much information when he's not ready, or use inappropriate terminology or concepts. Luckily, we've learned a lot about how preschoolers understand hemophilia. Keep reading to find out how you can better help your child understand his disorder. The right information, presented in the right way, may help calm him emotionally, build his confidence and sense of security, and encourage him to learn even more!

How Preschoolers Think

To learn how your preschooler understands hemophilia, it helps to know how preschoolers process their world in general. There are five major thinking tools that preschoolers use:

Magical thinking means that a preschooler cannot logically figure out a missing "causal" step in a step-by-step process. For example, if he sees one thing happen (Event A), then another (Event C), he automatically concludes that A caused C. He may be right, but he can also be very wrong! And he has trouble figuring out the missing step, Event B, that actually helped to cause the sequence of actions. To him, someone catches a cold (Event C) because she goes out without a coat (Event A). No mention of germs, coughing on someone, or getting sick!

Perceptual thinking means that everything the preschooler knows about his world comes from his senses: what he sees, hears, smells and feels. So he is very externally oriented; that is, focused on what's happening *outside* his body. This is one reason that preschoolers love magic–they believe what they see!

Absolute thinking means that a child perceives his world in black and white, with no in-betweens or shades of gray. Now you know why many preschoolers are attracted to superheroes–good guys and bad guys. To the preschooler, people are either sick or healthy, good or bad, old or young… there's no relativity for these kids!

Egocentric thinking means that the preschooler sees the world from his vantage point only. He has a difficult time putting himself in other people's shoes. What happens to him explains what happens to everyone. When it is daytime where he lives, it is daytime the world over. If you ask a preschooler to face you (try it!), then ask him to show you your right hand, he will point to your left hand–the same side that his right hand is on.

Immediate thinking means that the preschooler lives in the present. He rarely perceives a past, and thinks little of the future. His brain is hard-wired to focus on the moment. Ever take a long car ride with a preschooler, and five minutes into the ride hear him ask, "How much longer?" Five minutes is often the same as five days or five weeks to him.

You can use your knowledge of these thinking tools to help you teach your child about hemophilia. Where to begin? Well, hemophilia is a tough subject for so little a person. To avoid overloading your child, both cognitively (thinking) and emotionally (feeling), it helps to *know what your child thinks **first***. Often, the best way to begin teaching is to ask questions. Make them "open-ended" questions, like "What do you think hemophilia means?" as opposed to "Hemophilia is a blood disorder, right?" See the difference? Don't make your questions biased with any extra information. Just ask "How does…" or "Why is…" or "What happens next…" and see what he says!

"My port comes from God."

by Diane Horbacz

Did you ever wonder what your child thinks of his port? My husband and I used to discuss with our sons only how the port receives factor or draws blood. We focused on the *process* of getting factor, not on the port's origins. I assumed that my son Matthew, who is advanced in his thinking, would understand a lot about his port just from living with it daily. But we learned that parents can't assume anything when it comes to how our children understand hemophilia or ports. And it's important to understand how they perceive having a port, having it moved, or having it removed.

To a preschooler, having a port is like having a nose or a finger. It's an integral part of him, especially if he had it implanted as an infant. He may not remember what it is like to not have a port. And because a preschooler learns about his world predominantly by what he sees and hears, making him **perceptual** and **externally focused**, he may believe that his port is *external* to his body–not inside it!

My younger son, Justin, believes that his port is external. He debates me, pointing to the bump on his chest to prove I am wrong. When I ask, "How did it get there?" he can't answer. He doesn't remember his surgery. Yet he can *see* a bump, which we call a port–therefore, it's external!

When asked where their port comes from, preschoolers may say, "My port comes from God," or "I was born with it." Some preschoolers even think that their ports *give* them hemophilia: "I have a port. I have hemophilia. Therefore…" One causes the other. And if he believes that his port is external, a child is going to be in for a big surprise when it has to be removed!

What do you need to know if your child must have a port moved? First, think of it as an opportunity for learning. Ask questions. Learn what he thinks first, then engage him in a discussion about the port; you might be able to introduce new–and perhaps more accurate–answers. Since preschoolers are perceptual and concrete, an analogy may help them understand. Try this to help you understand your preschooler:

Imagine what you would think if a doctor told you that she had moved your belly button and put it on your nose! But your belly button has *always* been in the same place, since you were born. This is what you *believe* to be true. Now tell a five-year-old, whose port has been in his chest since he was ten months old, that his port has been moved to his belly! What will he feel? What will he think? Here is what my seven-year-old son Matthew said, after we first learned what he thought about his port, then taught him at his level of understanding:

I used to think that you are just born with it, but then I had to get a new one when I was five. So, now I know that a doctor puts it in. You have to go for surgery. They put you to sleep, take out the old one and put in a new one. Sometimes they even have to move it to a new place. When my mom first told me that my port was moved to a new spot, I didn't believe her. I didn't think that they could do that!

Helping our children understand the sometimes invasive procedures that come with hemophilia means taking the time to understand *how* they think. Do you focus only on the process of getting factor? Do you explore ideas with your child? Don't assume that your preschooler knows all about his port the way *you* understand it. Ask questions, like (pointing to bump) "What is that?" "Where did it come from?" "Why do you have that?" "How does it work?" Explore possible explanations. Let your child first attempt to figure things out and conjure answers. Finally, offer age-appropriate explanations to help him learn. Your efforts will support his burgeoning emotions and ever-developing thinking, and foster a better relationship between you and your child.

Diane Horbacz is the author of I Have Hemophilia and I am Special, Infusion Time, My Brother is Getting a New Port *and* Factor Fun. *She earned a masters degree in Special Education, and worked for five years as a special education teacher with emotionally and behaviorally challenged children. Diane currently works as an Education Specialist for Hemophilia Resources of America, where she has created and directs a pediatric program called "Karing for Kids." She lives in New Jersey with her husband, Gary, and her three children: Matthew, age eight; Justin, age four; and Erin, age one. Matthew and Justin have severe hemophilia.*

How Preschoolers Understand Blood

To teach your preschooler about hemophilia, try to learn first what he thinks about **blood**. This can be a fun task! Whereas hemophilia is a rather vague concept, blood is something you can see and touch–"It's red and goopy"–and preschoolers love anything involving the senses, because they are so perceptual.

To a preschooler, blood is a single thing–a red liquid. Because of their absolute thinking, preschoolers don't see the whole and its parts. Blood is *not* composed of little parts, or microscopic cells. There are no abstract proteins, and certainly no factor. Blood is just blood.

Although preschoolers are externally oriented, most will know that blood comes from inside. They cannot really tell you *where* from inside it comes, and most can't say what blood actually does; you get no step-by-step description of how blood carries nutrients and oxygen, or helps you heal. For a preschooler, blood is simply a red liquid that "goes around your body."

But, without being able to explain how or why, most preschoolers *do* realize that we need blood to stay alive, and that this is blood's primary function. "It helps you be alive. It helps us breathe."

You might think that all children with hemophilia would associate blood with hemophilia–but not true! Usually, *hemophilia isn't even mentioned* when preschoolers with hemophilia are asked about blood. Some may comment, "It helps with infusing" or "It's where you put the shot." But beyond that, blood has little connection with hemophilia. Since preschoolers are externally oriented, they may never link blood with hemophilia because they may never *see* blood when they need a shot–many bleeds are internal, out of sight.

Because your preschooler may believe that blood's function is to keep you alive, *be careful how you explain infusions.* Understand that preschoolers may fear an infusion–not only because of the pain of the needlestick, but because *they may lose their blood.* Reassure your preschooler, and explain what is happening during each step of the infusion.

How Preschoolers Understand Hemophilia

Since preschoolers don't usually mention hemophilia when you ask them about blood, they typically don't even consider hemophilia a blood disorder!

- Hemophilia is a **specific activity** that *the child* has personally experienced. This is due to his egocentric thinking. Hemophilia is "…when *I* get hurt and need a needle, or have to go to the hospital." "If you jumped on a hard board you could hurt your knee. You'd have to go to the hospital or something."
- Hemophilia is **perceptual**, and externally oriented. The preschooler focuses on tangible things he can see and feel outside his body. "You have to wear a bracelet and take factor when you get hurt." "I get a bad bruise and a needle helps it."
- Hemophilia is **immediate**, in the here and now. "When somebody punches you, you can get hurt real easily and have blood everywhere."

Look at the preschool thinking tools: perceptual, external and egocentric. Bruise, bracelet, and "when you get hurt." Hemophilia is something that you can see or feel, or that happens to you *when* you fall down or get hurt–*at that particular moment*. It's not necessarily something "in" your body that's always present.

When you talk to your preschooler, ask him questions first: "What is hemophilia?" "What happens?" "Then what?" Listen carefully. Does he use external symptoms to explain what hemophilia is? Is it an external, one-time situation (like a fall) that happens to him? Is it "when" he has to do something or go somewhere? Don't contradict him, or use words he's not ready for, like "blood disorder." Appreciate that he's using the best tools he has for the job. Ask questions, listen, and then praise him! Encourage him to open up, and you'll give him the confidence needed to explore his thoughts.

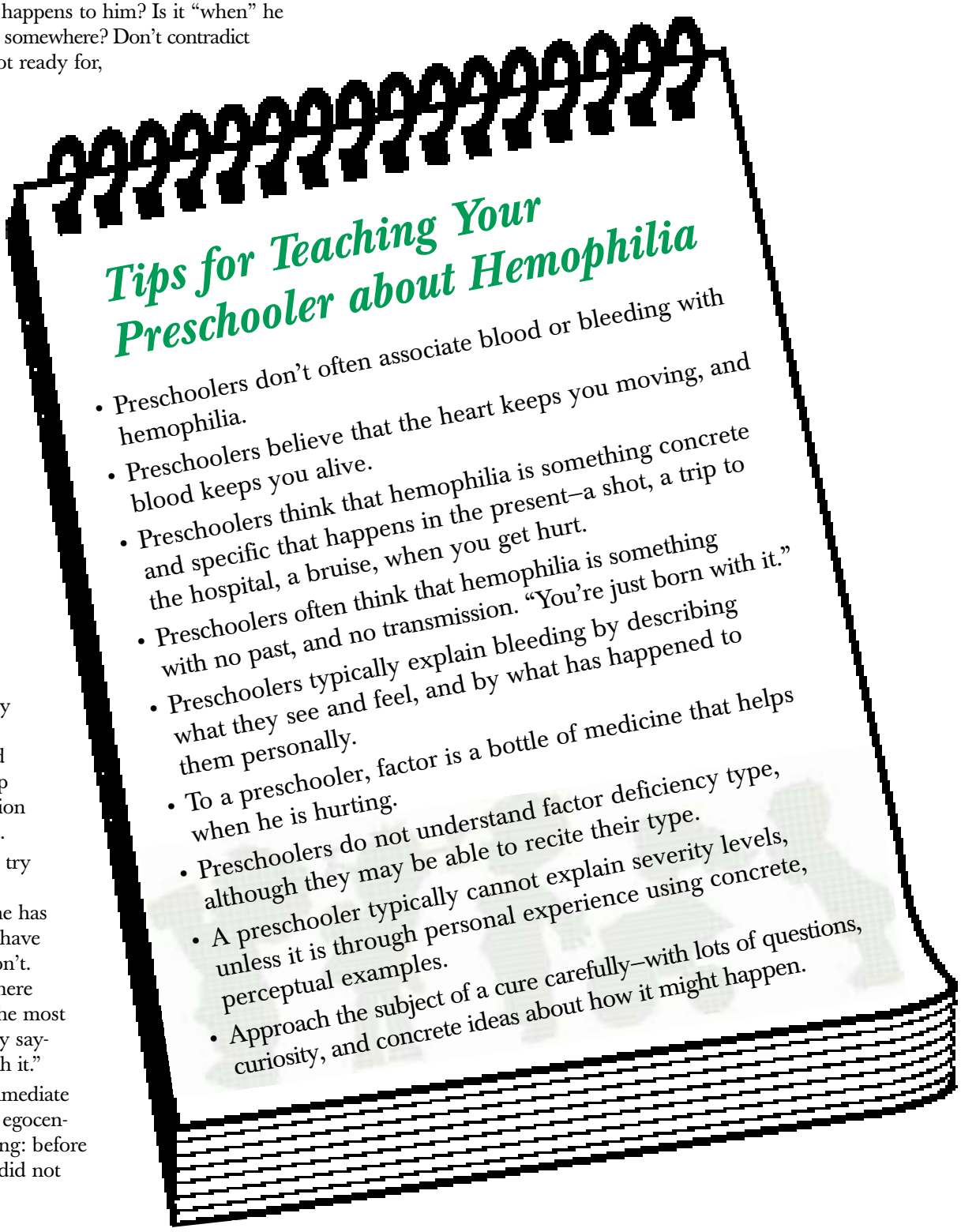
How Preschoolers Understand Genetics

If you think genetics is a tough subject to understand, imagine what it's like for a magical-thinking preschooler! Since preschoolers generally think in the here and now, discussing deceased ancestors and step-by-step links in genetic transmission will *not* make much sense.

A preschooler may still try to explain where he got hemophilia, or ask why he has to go to the hospital and have shots when his friends don't. Yet when you ask him where hemophilia comes from, he most likely will summarize it by saying, "You're just born with it."

This answer reflects immediate thinking (here and now), egocentricity and magical thinking: before *he* was born, hemophilia did not exist in your family or in anyone else. To him, there is no link between the fact that he was born (Event A) and the fact that he has hemophilia (Event C). (Okay parents, what's the missing Event B?)

Let's take it a step further. Most preschoolers *don't even mention their parents*, especially their mothers, as the source of hemophilia! Why? Magical thinking, with little cause and effect. A child has hemophilia, and he was born. Therefore, he was born with it. There is no past, and no transmission



agent. There is one exception: some children raised in actively religious households may say "God" when asked where hemophilia came from. This may be seen as Event B, the missing link. But keep asking. The key question is *how*. "How did God give you that hemophilia?" Your child might be stumped, reverting to, "I was just born with it."

So, when you ask your child where his hemophilia came from, relax! It's okay if your child offers this simple explanation. It's an answer that makes sense to him, and will keep him satisfied. And if he asks *you* where his hemophilia came from, don't feel obliged to spew out a confession of your defective genes, or exercise your overactive guilt: "Why us?" or "It wasn't supposed to happen this way!" Believe me, your preschooler does not want to hear this, mostly because these comments simply don't make sense to him. Instead, keep the doors of communication open by saying, "Well, what do you think?" or "I don't know. I guess you were just born with it."[1]

How Preschoolers Understand Bleeds

How do preschoolers explain bleeds? Bleeding is a step-by-step process, something that they are not yet equipped to handle. So to explain bleeds, they consistently rely on perceptual, absolute thinking, and immediate thinking tools.

Based on what he sees, the preschooler typically answers that when people get cut, "they bleed." The injured person gets better if he "puts a Band-Aid® on it" or "goes to the hospital." What's missing? Information about what happens *inside* the body, since preschoolers are so externally focused. Your preschooler cannot explain *how* the body reacts after a cut, and during a bleed, in a step-by-step fashion. And forget the time involved in healing–none of the preschoolers I have ever met has mentioned any length of time involved in healing.[2]

Now, what happens when *he*–the child with hemophilia–gets a cut? Pretty much the same response (perceptual, external), but with the added tools of egocentric and absolute thinking. First, either you have hemophilia, or you don't; it's black and white to him. And if you have hemophilia, and you get cut, you "get factor" or "go to the hospital." Some preschoolers will tell you that if you *don't* have hemophilia, you *don't* bleed when you get cut! This is absolutism and egocentrism combined, as the child explains the world from his own viewpoint.

[1] With preschoolers, avoid using words like "genes" or "chromosomes." These are too abstract, and involve too many step-by-step concepts.

[2] Be careful to *not* ask "loaded" questions, which imply an answer. Consider these two questions: "How long does your cut take to get better?" and "What happens after you put on a Band-Aid?" The first introduces the child to the concept that time is involved, and pre-empts his original thinking. The second encourages the child to express his own thinking–to figure out the problem on his own.

Preschool Logic: I Think, Therefore...

Cody takes factor on demand. One week he had three ankle bleeds in eight days. All three bleeds happened after he had done something fun. He became apprehensive and wouldn't let his mother out of his sight for three weeks, for fear he would have a bleed again if he was having fun. He just stayed where his mother was doing housework, and did nothing. For Cody, "I had fun, therefore, I bleed." In time, his mother was able to understand how he was thinking, and helped him see that it was Event B—the specific activity—and not the fun that had caused his bleeds! Cody's preschooler thinking directly affected his emotional well-being. Becoming informed about how Cody processes information helped his mother encourage him to handle his hemophilia and make better decisions.

Most preschoolers realize that they may need factor when they get cut, *but they can't explain why*. Why not? Magical thinking! They lack the ability to explain hemophilia in a step-by-step manner. "I get cut (Event A), I need factor (Event C)." But where's Event B? When asked why he would need factor while other boys wouldn't, your preschooler may respond, "Because *they* don't have hemophilia" (perhaps with some preschooler eye-rolling!).

When asked about internal bleeding, like joint bleeds, a preschooler will again resort to external, perceptual cues. He will describe the external symptoms of a bleed: "I bleed. It swells up. It hurts. You put a Band-Aid on it and you get an infusion. Maybe you can't walk or run."

Before you explain bleeds to your preschooler, think about how *he* processes incoming information. Ask him questions first: "What happens when you get a cut?" "Then what?" If he shows that he is curious and ready, provide some information. Or continue with your questions: "Well, why do *you* think it bleeds?" "How does it get better?" To uncover answers, the key word is *how*. "*How* does the Band-Aid make it get better, I wonder?" To help children feel empowered, I often pretend that I am pretty ignorant, and am sincerely trying to figure this out. (Of course, my teenager doesn't believe any more that I am pretending.) Children always seem more eager to explore answers when an adult is floundering, too!

Hands-on play is essential for children's cognitive development and understanding of hemophilia.

How Preschoolers Understand Factor

This might be the easiest concept for a preschooler with hemophilia to handle. What is factor? It's his bottle of medicine. He can see it, and all his explanations are perceptual: "It looks white, like water." "It's white powder." "It's stuff you get when you're hurting."

Factor is *not* protein that replaces the missing protein in his blood–he doesn't even know he lacks protein in his blood. Some children define factor according to what it does: "It helps my knee stop aching" or "makes my foot get better." A preschooler doesn't say he needs factor "to *stop* a bleed." Rather, he needs factor because he *has* a bleed. This is a big difference! It's magical thinking. The child observes event A (I got a bleed), then event B (I get factor), and reasons that, "I get factor *when* I get a bleed." But he sees no causal link (the factor stops the bleed).

How Preschoolers Understand How Factor Works

To describe logically how anything works, children must have the ability to explain a process in a step-by-step fashion. This is a skill beyond the reach of most preschoolers. So they often resort to magical thinking to explain a concept.

Preschoolers *see* factor being infused, and, being perceptually and externally focused, they often rely on this as their sole explanation: "It goes in your veins. It goes deep inside you." Or, "It makes you better. You get a shot and you don't bleed no more."

To a preschooler, factor works by "going through your body." While he generally perceives some internal organs (usually only the heart or stomach–can you guess why?), he doesn't understand the system of veins and arteries. Still, he may mention that factor goes into the vein or blood–that much he can see. After that, factor's role is a mystery!

How Preschoolers Understand Factor Deficiency Type

Preschoolers generally have no comprehension of factor deficiency type. Because preschoolers can label things, or mimic what they are told, it's no surprise that when asked, "Do you know what type of hemophilia you have?" some say, "Factor eight." But continue: ask your preschooler what *that* means. No preschool child can define "factor eight," except with the perceptual, "It's the medicine I get."

Replying "factor eight" is reciting words without real comprehension. But we're not looking for "right" answers from our children; we are seeking to learn *how* they process their world. It's really less important for a child to memorize what factor deficiency type he has than it is for him simply to understand, "Hemophilia means I have to get shots."

Preschooler Thinking Tools

Preschoolers exhibit **five** thinking traits that help them understand their world.

1. Magical When explaining *why* things happen (causality), preschoolers skip a step in logical thinking. They often explains things chronologically: Event A (counting to three) and Event C (traffic light changing) means that A *caused* C. "My counting caused the traffic light to change!" What's missing is Event B–the thing that actually, logically causes Event C.

2. Perceptual Thinking is completely dominated by what a preschooler sees, hears, feels or smells. Because thinking is perceptual, it is almost always externally oriented–outside his body.

3. Egocentric A preschooler is incapable of seeing the world from another person's point of view.

4. Immediate Preschoolers exist in the here and now, with little understanding of the past or future. They have difficulty understanding the concept of measuring time.

5. Absolute Preschoolers think in polarized extremes: all or nothing, good or bad, sick or healthy. They see no in-betweens, no gray areas, no overlapping. They have difficulty seeing a relationship between objects, particularly between a whole and its parts.

The only benefit of memorizing factor deficiency type is that under some unusual circumstance, a child may need to know this information for medical reasons–just as preschoolers should know their phone number or street address. So feel free to tell your preschooler his factor deficiency type, but don't try to explain what that means.

How Preschoolers Understand Severity Levels

Mild, moderate, severe. Understanding severity levels means being able to see variations, gradations, shades of gray, parts of a whole. Preschoolers are absolute thinkers, usually seeing only "black and white." They don't understand parts and wholes. To a preschooler, either you have hemophilia or you don't. Either you bleed or you don't bleed. There's no middle ground!

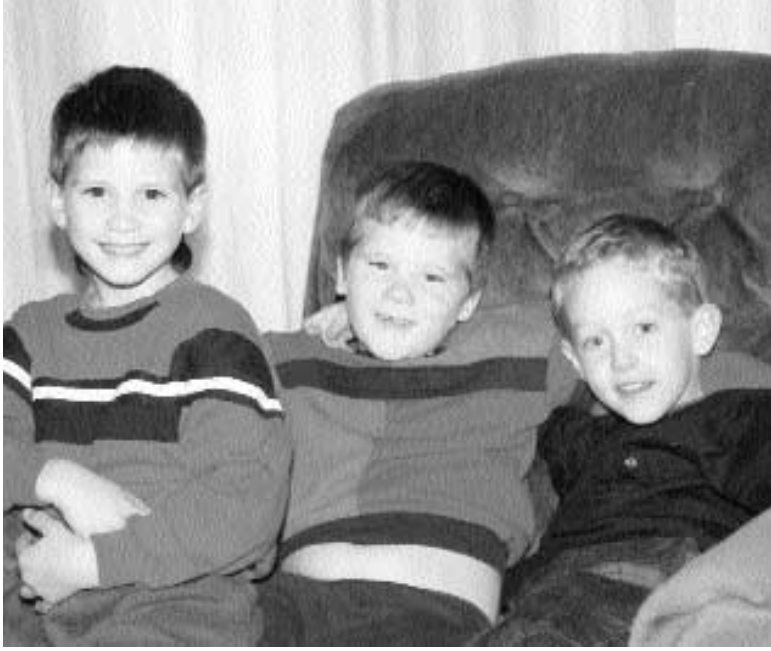
How do preschoolers explain why some children with hemophilia bleed more than others, even when both children were doing the *same* activity? Some preschoolers with hemophilia are aware that certain children get factor more often than others. But they explain this using preschool thinking tools, relying on perceptual and concrete thinking: if one child happens to bleed harder than another, then he must have *hit his knee harder*. Or, preschoolers may rely on egocentric thinking and personal experience. For example, a child may know someone personally with hemophilia, like him, whom he observes getting more shots than he gets. "Trevor has the hemophilia that bleeds a lot. I just bleed a little bit."

How Preschoolers Understand Hemophilia as a Lifelong Condition

You'd think that because preschoolers live in the here and now, they would have no concept of permanence, of hemophilia being with them forever. But here's where experience and teaching play a major role in educating our children and shaping how they think: Do preschoolers think about a cure, about a future without hemophilia? Undeniably, they do.

"Do you think that some day doctors will make hemophilia go away forever?" If you ask this question, many preschoolers will respond, "Yes!" Why? Well, it's not because of magical thinking; nor is it due to advanced understanding of the concept of permanence. It's because of their parents. As a parent, you can have a tremendous impact on your child when you discuss with him his "condition," and the fact that "doctors" are working on a way to make it go away forever.

So don't avoid the subject! Yes, preschoolers have a unique way of viewing the world. Yet you can ask questions, probe your child's thoughts, and get the ball rolling, to provide information in a form he can visualize and comprehend. Use simple, concrete language and perceptual imagery to explore the subject of hemophilia: "Maybe some day when you get hurt, you won't need a shot, because doctors will have some new medicine that makes



Listen to your child when he talks about hemophilia. He'll feel loved and valued, and develop confidence.

hemophilia go away forever." But always remember that you're speaking to a preschooler! *Do not build his hopes too high, too fast.* A child this immediate and perceptual might think that his next visit to the clinic, or his next infusion, will provide that "new" medicine.

More than filling a cognitive need to educate your preschooler, your discussions about hemophilia can also fill a need of the heart. Nothing in the world validates a person, making him feel valued and loved, like *listening*–intensely and openly. When you ask your child about hemophilia, and listen, really listen, you are doing more than educating him and learning about him. You are honoring him by showing him that he is loved *now* by you, for exactly who he is. Spend your infusion time wisely; use it as a time for dialogue, and for building a better relationship. Learn a little and teach a little. And reinforce your message physically, concretely, with lots of hugs and kisses!

Laureen A. Kelley is the author of several books about hemophilia, including Raising a Child With Hemophilia. *She is President of LA Kelley Communications, Inc., which is dedicate to improving the lives of hemophilia families worldwide by empowerment through education. Laurie and her husband Kevin, a process scientist for a biotechnology firm, have three children: Tommy, age fifteen, who has moderate factor VIII deficiency, and daughters Tara, twelve, and Mary, eight.*

This article is adapted from the new edition of *How Children Understand Hemophilia*, to be published in early 2003. The book is provided through a generous grant from Aventis Behring. The new edition will include an entire chapter devoted to how children understand prophylaxis. You can place an order for your new copy by visiting **www.kelleycom.com** today!

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In the last issue of *PEN*, Michelle Longsworth, a carrier of hemophilia, wrote that she needs help encouraging her father to get tested for hemophilia. Her father, who lives in Belize, has symptoms of hemophilia. Michelle is concerned that he obtain proper treatment in the event of an emergency. Here are two responses to Michelle's letter.

I am also a carrier, with mild hemophilia. I have an eight-year-old with severe hemophilia. My father, also severe, died when I was very young. Although I did not grow up around hemophilia, I always knew about it, and knew that I had it. I think you are doing the right thing by only telling your father the positive side of this disorder. However, he should be tested. Since he has been very emotional about your sons and your diagnosis, be gentle. Explain that this is a very manageable disorder, and if he is diagnosed with hemophilia, the treatment will make his life a lot less painful. It will dramatically improve his quality of life. If his factor levels are low, he will be better off knowing, being able to treat, and making informed decisions about that treatment.

***Tammy Davenport*, TEXAS**

I am the mother of 10-year-old twins, Evan and Melissa, and my "little bruiser," two and a half-year-old Justin, who has moderate hemophilia A. Let me share my story with you in the hope that it will convince your father to get tested.

Testing saved my father's life. When I was growing up, he had many prolonged bleeding episodes. We were always told he was a "bleeder." He was not diagnosed by a doctor, and we never heard the word "hemophilia." I didn't realize how much I would come to know about this disease, and how it would affect us in the future.

Justin was born in December 1999. After his circumcision, it took two trips to the ER to stop his bleeding. It was terrifying. At that time, his veins were too small to obtain enough blood for testing, so we were told to wait until he was six months old. It was during that waiting period that things started to click. I researched bleeding disorders. Based on the information I obtained, and recollections of my father's bleeding problems, I knew Justin had some kind of bleeding disorder.

When Justin was six months old, he was diagnosed with moderate hemophilia A. Although the signs had pointed toward this diagnosis, it was a shock. My father and I both dealt with our own feelings of guilt and responsibility–mine for not making the connection sooner, and my father's for passing hemophilia to his grandson. It was a very difficult time. My tough, stubborn father cried like a baby. His only soft spot is for his family, especially his grandchildren. They are the only reason he went to a hematologist to get diagnosed. My twins begged him to go, and he listened to them. I had often tried to convince him; but he argued that he had survived this long without seeing a hematologist, so why start now? Grudgingly, he made an appointment, got his blood test, and at 61, was finally diagnosed with moderate hemophilia A.

In November 2001, as we were planning Justin's second birthday party, my father suffered a stroke with significant bleeding in the brain. Had the medical staff *not* known he had hemophilia, he would not have received factor, and would not have survived. Miraculously, he pulled through with only minor memory impairment, which continues to improve. As we plan Justin's third birthday, I watch grandfather and grandson play. They are so alike in so many ways–same eye color, hair color, personality… same MedicAlert® bracelet, same bleeding disorder. I thank God for my father's survival, and for my son, who played a part in saving his life. Michelle, share this story with your father. Tell him that you love him, your grandchildren love him, and that you must do everything in your power to keep him safe and healthy. This might mean telling him things he doesn't want to hear.

***Ellen Najimian*, NEW JERSEY**

Question

How and where do you store your supplies? When both of our boys were infused via ports, we made up baggies with everything they'd need for one infusion, and kept them in a chest set aside for our poke supplies. Now that Sam is doing arm pokes, I need storage suggestions. We don't use the baggies now, because we need less stuff per infusion. Yet I find myself having to running back and forth when I forget the tourniquet, gauze or syringe. Do you have a favorite kind of tourniquet?

How do you draw up your syringes for the poke? Our HTC teaches new parents doing peripheral sticks to poke the butterfly in; let the blood return; fill the line; attach the factor syringe (with a little extra air at the top of the syringe); watch for the air bubble to get close to the needle; then withdraw. [See "As I See It," page 3] We're currently loading the needle with saline, switching to the factor syringe once we're in, then flushing with saline at the end.

***Jill Lathrop*, Wisconsin**

Answers

We make "kits." I love it, and the girls help me, so it's a family thing. We keep adding new things–most recently, the BAXJECT®.

As a nurse, I really like the lab trays we use. They're like those little rectangular buckets with a handle that you can buy to store household cleaning supplies. They have separate compartments.

Before we got my son Noah's port, our HTC had us just prime the butterfly tubing with the factor itself. If you could eliminate the need for saline, it seems that you would have even less to pack in your kits. You could still do little kits, but use small sandwich baggies instead of gallon-sized.

***Stacey Rainer*, Illinois**

I store my son's infusion supplies (minus the factor and everything that comes with it) in a box. Inside the box is a folder for his log sheets, and the papers that come with factor deliveries. I keep all the syringes in one big baggie in the box, and the butterflies in another big baggie. I also keep Tegaderm™ transparent dressings, EMLA® anesthetic cream, gauze pads and alcohol pads in the box. I have one little baggie, which I make up every day after the infusion so I'm ready for the next infusion. In it I put enough supplies for one infusion: alcohol pads, gauze pads, butterflies, a syringe and Band-Aids. Before each infusion, I take the box to the kitchen countertop. I put the baggie on the table where I mix the factor. I infuse my son while he sits on the couch and watches TV, so I just bring the supplies I need to the couch. I don't use a separate saline syringe, and I keep the syringe attached to the butterfly at all times–this is just easier for me. I don't flush with saline at the end.

***Jane H.*, New Jersey**

We have a large toolbox with everything I need, even the needle container. It contains enough supplies to do about ten sticks. In a tray in the upper section, I store the tourniquet, saline, bandage–like a tray in the lab. We bring the toolbox on vacation, filled up and much heavier than usual. I can even lock the box to deter nosey toddlers.

We use a plain-looking green tourniquet. It's not very special, but I've seen some designed and decorated for children, which are very cute but difficult to open with one hand. Once on holiday in France, when we couldn't yet do the sticking ourselves, a French nurse helped us; she liked her own tourniquet, a rubber cord that she wrapped around the arm.

At home we stick with a saline-filled needle, and flush afterwards. I prefer this, perhaps because it makes me feel secure.

***Femke Meijer*, Netherlands**

I bought a toolbox for supplies I use at home. I keep Coban™ (a self-adherent wrap) and IV start stuff, arm boards and 2x2 gauze on the bottom. In the lift-out tray, I keep butterflies and heplock stuff. In the open-up top I keep alcohol, Band-Aids, tape measure, tourniquet and EMLA cream with Tegaderm.

I also have a travel pack from our manufacturer that I keep loaded with one high dose of factor, tourniquet, two butterflies, one syringe, 2x2 gauze, alcohol, one tube EMLA, two Tegaderm; a copy of Caydin's insurance card, a factor box top that has the order label on it, and the assays in the kit (in case the bottles get lost, I will know the lot numbers infused); a 5 cc sealed syringe, and a dose of Tylenol® for pain. This is all zipped up in a baggie. I take it when I travel more than 20 minutes from home, or anywhere Caydin may get hurt, so we can infuse. The factor dose in the travel pack is always the first dose we use to treat a bleed, even if we're home, because our factor is good only six months when stored at room temperature. Caydin also takes the travel kit with him to his dad's house on weekends.

When I infuse Caydin, I prime the butterfly with factor. I have missed the vein and infiltrated multiple times in the last few years with factor–you notice very quickly if it infiltrates, and you stop pushing. Infiltrating doesn't seem to change the bruise size, and I don't have to change syringes. I used to do the air bubble, but have stopped because Caydin pushes his own factor now, and the amount lost in the tubing isn't worth the independent feeling he gets doing it himself. We've used the BAXJECT once and we both love it because it's now much easier for Caydin to mix and draw his own factor.

***Janis Kosak*, Texas**

I keep my supplies in a locked cupboard, and take out what I need at the time. You could post a list of what is needed inside the door. I keep a baggie full of supplies, plus a little extra, inside our small travel factor bag–it goes everywhere with us when we leave the house. In our large travel bag, I keep enough supplies for four or five infusions, and two plastic pencil boxes–a green box for anything I don't want to get wet or that might be damaged (like Betadine® swabs, which occasionally leak); a red box for used needles, so I don't have to carry a sharps container. I transfer them when I get home.

***Mindy Gerdes*, Michigan**

Tracing Hemophilia Families

From ***Dawn Bonney***
MAINE

"As the mother of a 17-year-old-son with hemophilia B, I have been tracing hemophilia in my genealogy. I read an article online by Dr. Thomas Roderick, a geneticist from Maine. Dr. Roderick stated that he was part of a study linking 80% of the hemophilia B in Maine to one family living in Cherryfield, Maine, in the early 1800s.

"*PEN* has the good fortune to be in touch with many families with hemophilia B. Is there a way that *PEN* could help me inform Maine families with hemophilia B that I'm looking for contacts? We might help them find their founding family with hemophilia.

"I've learned that the hemophilia came originally from three daughters of Hannah Plummer and Theodore Leighton. If anyone is interested in this subject, please contact me at sunrise@megalink.net."

Wanted! PEN PAL

From ***David Mordigal***
CONNECTICUT


David and Clover

"I am eight years old and have severe hemophilia A. I get factor three times a week through my port. My infusions have kept me from getting bleeds for a long time. I live with my mom, dad and five-year-old brother, Jared. Jared doesn't have hemophilia, but he likes to watch me get my infusions. I have two guinea pigs named Clover and Buttons, who don't do much, but are fun and easy to care for. I am in the third grade. I am in 'Bear' rank in cub scouts, and have a great time in scouting with my friends and all the great activities!

"My favorite sport is swimming, and I especially like to wear goggles underwater. I also enjoy using the computer, Nintendo, reading, and writing stories and letters. I am very interested in mechanical things like elevators and traffic lights, and I hope to take piano lessons. My favorite books are the *Harry Potter* series, *The Boxcar Children*, the *A to Z* series, and any mystery. I have lost nine teeth, and two more are loose. My mom almost had to give me Amicar® for my teeth, but my infusion took care of the bleeding. I am very good at replying to letters, so I can't wait to get a pen pal!"

At the Movies: *The Doe Boy*

by Richard Atwood

This fall, the film *The Doe Boy* was released on DVD. Written and directed by Randy Redroad, and based on an autobiographical incident, this coming-of-age drama is set in Oklahoma in 1984. Hunter Kirk (played by James Duval) is half Cherokee and has hemophilia. His father is frustrated by the financial burdens, and the hunting limitations, imposed by hemophilia. Hunter's Native American mother, who is a nurse, is both caring and overprotective. Hemophilia is considered a "white man's disease," making Hunter an outsider in a culture obsessed with blood identity. As an 18-year-old, Hunter must find his own freedom separate from his parents, while discovering love and friendship.

The cast includes Kevin Anderson, Jeri Arredondo, Andrew J. Ferchland and Gordon Tootoosis. Filmed on location in Tahlequah, Oklahoma, *The Doe Boy* won Best Actor, Best Actress, Best Director, Best Film and Best Supporting Actress at the 2001 American Indian Film Festival.

The Doe Boy, 2001. Wellspring Media, 87 minutes.

Important Scholarship News!

Applications for The Eric Dostie Memorial Scholarship are now available.

Eight $1,000 scholarships will be awarded to students with hemophilia or a related bleeding disorder, or to their family members. Application deadline is March 1, 2003. The Eric Dostie Memorial Scholarship is made possible through the generous funding of Mr. Patrick M. Schmidt, CEO of FFF Enterprises, and founder of NuFactor, a homecare company and cosponsor of *PEN*. Please visit **www.kelleycom.com** for more information.

Eric Dostie
March 28, 1989 -
August 27, 1994

Corrections

In the August 2002 issue of *PEN*, we failed to acknowledge Aventis Behring as corporate sponsor of the book *A Guide to Living with Von Willebrand Disease*. *PEN* regrets this unintentional omission. Aventis Behring continues to provide groundbreaking resources for patient education in the bleeding disorders community through its many publications and services, which can be reviewed at www.allaboutbleeding.com.

We also regret our failure to indicate that The Eric Dostie Memorial Scholarship is made possible through the generous personal funding of Mr. Patrick M. Schmidt of Fallbrook, California. Mr. Schmidt is CEO of FFF Enterprises, and founder of NuFactor, a homecare company and cosponsor of *PEN*.

The "Hemophilia Statistics and *PEN* Readership" table (page 15, *PEN*, August 2002) overestimated the number of people with hemophilia A and B by a factor of more than two. This error occurred because the prevalence was estimated based on the total population, when it should have been applied to the *male* population only (which, for most states, is very close to half the population). This error also caused the *PEN* readership percentage to be underreported–it is more than twice the percentage listed in the table.

Mike Soucie, Ph.D., of the CDC, reports that the combined prevalence of both hemophilia A and B is about 13.4 cases per 100,000 males, or about 1 per 7,500 males. (The incidence of hemophilia A alone is approximately 1 in 5,000 males; for hemophilia B it is 1 in 40,000 males.) Based on data from the CDC's Hemophilia Surveillance System (HSS), the CDC estimates that there are 400 infants born each year with hemophilia, an estimated 200 deaths per year, and a current estimated hemophilia population of about 18,500 in the U.S.

Visit the LA Kelley Communications, Inc. website (www.kelleycom.com) to view a corrected hemophilia statistics table.

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