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June-July 2014

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INDICATIONS AND USAGE

Flublok is a vaccine indicated for active immunization against disease caused by influenza virus subtypes A and type B contained in the vaccine. Flublok is approved for use in persons 18 through 49 years of age.

DOSAGE AND ADMINISTRATION

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DOSAGE FORMS AND STRENGTHS

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To report SUSPECTED ADVERSE REACTIONS, contact Protein Sciences Corporation at 1-888-855-7871 or VAERS at 1-800-822-7967 or www.vaers.hhs.gov.

USE IN SPECIFIC POPULATIONS

- Safety and effectiveness of Flublok have not been established in pregnant women, nursing mothers, children, or adults 50 years of age and older.
- A pregnancy registry is available for Flublok. Contact: Protein Sciences Corporation by calling 1-888-855-7871.

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Manufactured by:

Protein Sciences Corporation

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About *IG Living*

IG Living is the only magazine dedicated to bringing comprehensive healthcare information, immune globulin information, community and reimbursement news, and resources for successful living directly to immune globulin consumers and their healthcare providers.

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DONA DARR
Mother of a PI Daughter

IG Chronicles: Ready for Back to School? I Hope So...

"I can't help but feel mixed emotions with the start of the new school year inching ever closer."



TERRY O. HARVILLE, MD, PHD
Medical Director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences

Diagnosing an Antibody Deficiency: Case 6, Part 2

"The severity and unusual nature of infections, requiring longer courses or more powerful antibiotics, and, in particular, radiographic evidence of pneumonia and/or sinusitis are more compelling evidence that something is amiss."



MATTHEW D. HANSEN, DPT, MPT, BSPTS
Physical Therapist

Exercise Success

"Some patients living with a PI or autoimmune disease may still be capable of competing in recreational athletics if care is taken not to exhaust the immune system by overtraining."



AMY SCANLIN, MS
Medical Writer

Screening for Severe Combined Immunodeficiency Disease

"TREC screening has a high rate of accuracy, which results in significant cost savings."

Be a Part of IG Living's Blog and Facebook Discussions!

IG Living isn't just a magazine; it's an interactive community of people with interesting stories. And, we want you to be a part of it! Our blog, which has a new entry posted every Friday, provides interesting perspectives on topics such as living with IG, people to be admired, support and encouragement, current news of interest and things you just need to know. We encourage you to comment with your opinions and stories. And, better yet, if you have a blog you'd like to post, send it our way!

IG Living's Facebook page has hundreds of fans who respond to our questions that are posted every Monday through Friday. Together, these fans share their life stories and thoughts. What's more, they are making a connection with one another that otherwise wouldn't be possible.

So, be a part of it now at www.igliving.com/blogengine and www.Facebook.com/IGLivingMagazine.



Connect with Other IG Living Readers through Monthly Teleconferences!



IGL's Readers Group Teleconferences allow readers to connect with others to share their experiences living with chronic diseases. Here's how you can participate:

- Email IG Living to be added to our email invitation list for the teleconferences.
- IG Living will send you invitations to let you know when the monthly, hosted, toll-free teleconferences will be held, as well as what topic relevant to the IG community will be discussed.
- The moderated, hour-long calls will be filled on a first-come, first-served basis and participant numbers will be limited.

In addition to connecting with others, IG Living's patient advocate can help you determine if there's a patient organization support group in your area.

Sign up for the Teleconferences now by emailing patientadvocate@IGLiving.com or calling (800) 843-7477, ext. 1366.



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Making Gains

Over the years, we have received many emails from our readers expressing dissatisfaction with their lives post-diagnosis. The common theme: a desire to be able to do the things they used to do. But, coming to terms with a “new normal” means learning to accept gains even if they don’t measure up to what could previously be accomplished.

This issue, we feature an extraordinary man, Brandon Dillon, who in his 20s began experiencing recurrent sinus infections and pneumonias. An avid cyclist and runner, he was struggling with his energy and stamina. After his diagnosis of common variable immunodeficiency (CVID) at age 37 and a successful response to intravenous immune globulin therapy, Brandon followed his immunologist’s advice to continue to participate in the activities he loves. He has now completed his first marathon, two half-marathons and several triathlons, and he hopes that others who think their life has ended with a primary immunodeficiency (PI) diagnosis will also realize that life as they once knew it doesn’t have to be over.

Brandon, most certainly, is one of the exceptions in the PI community. As our article “Understanding and Treating Common Variable Immunodeficiency” explains, the disease is anything but “common,” and it is most certainly “variable”; no two patients experience the same degree of disease symptoms. Some, like Brandon, respond well to treatment and continue to flourish in life. Others continue to be plagued with infections and are unable to actively participate in their lives as they once did.

But, that doesn’t mean that those who are often ill despite treatment can’t

aspire to make gains in their lives; it’s all a matter of perspective. Matthew Hanson, author of the article “Exercise Success,” has a personal philosophy: “Life is a competition of endurance, made up of individual battles, not so much with outside opponents as with our own expectations of ourselves.” From this perspective, he outlines how chronically ill patients who used to be athletically competitive can continue to do so by understanding that their successes are measured by whether or not they do the very best they can with what they have. This is a philosophy that can be used not just in athletic competition, but in all aspects of life.

Fortunately, as diseases like CVID become more well-known, the long-term outcome of PI patients and the risk of severe complications are improving due to earlier diagnoses, resulting in healthier lives and patients’ ability to accomplish more than they could previously. This is true for many PIs, including severe combined immunodeficiency (SCID). Since 2010, 18 states have added SCID screening to their newborn screening programs, and many more are following suit. What this screening is and how it saves the lives of so many infants nationwide each year is discussed in our article “Screening for Severe Combined Immunodeficiency Disease.”

I hope you gain insight from the information presented and enjoy the many other articles in this edition of *IG Living*.

Ronale Tucker Rhodes, MS

Ask the Experts



Reader: I infuse immune globulin (IG) subcutaneously with Hizentra weekly. How much flexibility do I have between doses? How close is too close together?

Amy: There's not a hard rule regarding dosing. In fact, Hizentra was recently approved for biweekly (every two weeks) dosing. One of the benefits of subcutaneous IG therapy over intravenous IG therapy is flexibility in dosing, as well as more consistent IgG trough levels due to smaller doses of IG administered more frequently. Therefore, the most important thing is to infuse the drug, regardless of the timing.

If different sites are being used, the weekly doses could be infused on consecutive days if needed. If the same sites are going to be used, I would not recommend infusing until the previous dose has been absorbed.

Reader: How long can an open bottle of immune globulin (IG) be used? I infuse at home and want to use a very slow rate due to migraines, but it's time-consuming. Can I draw half the dose into my pump to infuse on Wednesday and then infuse the other half on Thursday?

Amy: All IG products are preservative-free, so they should be used immediately. If the dose is prepared in a pharmacy in an IV room, there is more flexibility for longer-term storage. But, in the home, immediate use is recommended. If this is an intravenous IG infusion, you should speak with your pharmacist about sending smaller vial sizes to split the dose over two days. For example, if the dose is Privigen 40 grams once a month, instead of sending a 40-gram vial, the pharmacist could send two 20-gram vials or even four 10-gram vials. I would also suggest you let the pharmacist and physician know about the migraines to see if another adjustment in therapy can be made (addition/change of pre- or post-infusion medications, hydration or possibly a change of brand), in addition to infusing the dose over two days.

Reader: I experience severe skin reactions to the tape used to hold the needles in place during subcutaneous

immune globulin (SCIG) infusions. What can you suggest?

Amy: There are several options to secure SCIG needles. If you are using paper tape (Micropore), you could use plastic tape (Transpore) or silk tape (Durapore) instead. Occlusive dressings such as Tegaderm, Opsite or Primapore may also be used.

Reader: I have common variable immune deficiency. My doctor ordered 500 mLs of normal saline prior to and after my infusions of 40 grams of Privigen. Is normal saline compatible with Privigen?

Leslie: Intravenous immune globulin (IVIG) products have limited compatibility with normal saline. If IVIG requires dilution to a lower concentration, the manufacturer of Privigen suggests dextrose 5% in water solution as a compatible solution. For flushing of IV lines, it is acceptable to use normal saline to flush before or after IVIG. For adding fluid as extra hydration, there are a couple of things to consider. First, it is not recommended to run normal saline and IVIG at the same time through the same IV line, unless there is a physical separation. For example, some people have an IV access line that has more than one line — called a double or triple lumen catheter. In this situation, the IVIG and hydration solution are being infused into two separate lines within what looks like a single line. Second, when adding normal saline as a hydration solution, infuse it before or after the IVIG, as your physician has ordered. Or, consider using dextrose 5% in water as the hydration solution. The option your physician selected, to infuse normal saline before and after your IVIG, is fine. ■

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LESLIE J. VAUGHAN, RPh, is senior vice president of clinical services at NuFACTOR Specialty Pharmacy.

Immunology 101:

Diagnosing an Antibody Deficiency: Case 6, Part 2: A 2-Year-Old Boy with Chronic Respiratory Symptoms

By Terry O. Harville, MD, PhD

LAST ISSUE, we began the discussion of a 2-year-old boy, who began an apparent respiratory infection before 1 month of age, which required antibiotic treatment. He continued with apparent respiratory infections year-round, although they were worse during the winter months. Multiple courses of antibiotics were required for treatment.

When considering the significance of infections, several factors have to be evaluated. Did X-rays or other diagnostic testing demonstrate evidence for an infection? For example, pneumonia can be more or less confirmed by specific features found on X-rays of the chest. If pneumonia is diagnosed, but X-ray evidence is not present, then is it truly pneumonia? Or, is it asthma misdiagnosed as pneumonia? If green mucous is found in nasal secretions, does this indicate sinusitis, even if X-rays or CT scan are not performed to indicate sinusitis is present? Commonly in children, the diagnostic procedures are not performed, but antibiotics are given. In these cases, objective data for a true bacterial or fungal infection is lacking. And, an apparent beneficial effect of the antibiotic may be a misinterpretation of the viral illness running its course.

It's also necessary to consider whether the patient required more powerful or even intravenous antibiotics to clear the illness. When the typical course of oral antibiotics did not work, were more powerful antibiotics required? Were multiple courses or repeat courses required to cure a single infection? If cultures were obtained, were unusual infectious organisms present?

The severity and unusual nature of infections, requiring longer courses or more powerful antibiotics, and, in particular, radiographic evidence of pneumonia and/or sinusitis are more compelling evidence that something is amiss — specifically, the immune system may not be functioning at its full capacity.

After the 2-year-old's first infection, he seemed to be fine for several weeks, but then fevers began to occur. First, an ear infection was diagnosed, which seemed to respond to antibiotics. Subsequently, infections were diagnosed as ear infections, but fever and symptoms continued through the initial course of antibiotics. After a few days, the diagnosis was changed to sinusitis or



pneumonia. At first, the boy was switched to a more powerful antibiotic for a somewhat longer course. Subsequently, his physician began with the more powerful antibiotic at the onset. On a couple of occasions, the boy received shots of antibiotics. During each of his two winters, he spent a few days in the hospital, with a diagnosis of asthmatic bronchitis, but he did not receive IV antibiotics. The X-rays were not interpreted as pneumonia.

In light of this history, there is evidence that the immune system may not be fully functioning normally, but his physician was interpreting the situation through a different set of filters. An older sibling had been sickly as an infant, but outgrew the problem. Both parents and other relatives have allergic disease, and asthma is present in the family. Therefore, his physician interpreted these issues as an indicator that the patient is likely more sickly than other children at the same age due to risk for allergies and asthma. And, despite having illnesses year-round and requiring extensive use of antibiotics, the patient was expected to outgrow the problem.

We will continue with the discussion of this case in the next issue. ■

TERRY HARVILLE, MD, PhD, is medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, and a consultant for immunodeficiencies, autoimmunities and transplantation.

Disclaimer: This case report is intended as an illustration for education purposes only. It does not necessarily represent an individual or precise information from patient files.

Diseases

More States Are Screening for SCID

Eighteen states are now screening for severe combined immunodeficiency (SCID), and another dozen states are slated to start screening by the end of 2014. States currently screening for SCID include California, Colorado, Connecticut, Delaware, Florida, Iowa, Massachusetts, Michigan, Minnesota, Mississippi, New York, Ohio, Pennsylvania, Texas, Utah, Washington, Wisconsin and Wyoming. Combined, these states are screening for more than 50 percent of babies born in the U.S. Slated to begin screening for SCID are Illinois, Maine, Missouri, Nebraska, North Dakota, Oklahoma, Oregon, Puerto Rico, Rhode Island, South Carolina, South Dakota and West Virginia. In addition, advocacy efforts are currently targeting states where advisory committees have approved



adding SCID, but the timeline for implementation remains unclear. These states include the District of Columbia, Georgia, Maryland, New Jersey, North Carolina and Virginia. And, the Jeffrey Modell Foundation is continuing to pursue funding to combine with their own funds that would be provided as seed money for states that agree to screen. SCID screening received more attention after former

Secretary of Health of Human Services Kathleen Sebelius added it to the panel of 29 genetic disorders to be screened for at birth.

SCID is a primary immunodeficiency that results in the onset of one or more serious infections such as pneumonia, meningitis and bloodstream infections within the first few months of life. Left untreated, infants typically will not survive one year. Babies born with SCID appear to be healthy, which is why SCID screening is critical. If a baby is diagnosed with SCID and receives a bone marrow transplant within the first three-and-a-half months of life, the survival rate can reach 94 percent. The survival rate for infants who receive a bone marrow transplant after this age drops to less than 70 percent. ■

Vaccines

IDF Issues Vaccine Guidelines for PI Patients



The Medical Advisory Committee of the Immune Deficiency Foundation (IDF) has issued recommendations concerning the use of live viral or bacterial vaccines for immunodeficient patients. Based on current literature, the com-

mittee recommends that clinicians educate parents and physicians about the need for maintaining herd immunity in the population at large; avoid live viral and bacterial vaccines in all patients with significant T- and beta-cell deficiencies; determine the degree of immune reconstitution in patients treated with hematopoietic stem cell transplantation (HCT), enzyme therapy or gene therapy before live vaccine treatment; and balance the exposure to infection from live vaccines and close contact-transmitted vaccine-derived infection in the immunoreconstituted child.

“These recommendations address the concern for immunodeficient

patients acquiring infections from healthy subjects who have not been immunized or who are shedding live vaccine-derived viral or bacterial organisms,” William T. Shearer, MD, PhD, of Baylor College of Medicine and Texas Children’s hospital, and colleagues wrote. “Immunodeficient children who have attained full immune reconstitution after bone marrow, blood or cord blood stem cell transplantation might have sufficient T-cell responses to protect against exposures to horizontal viral infection, but careful evaluation of the degree of immune reconstitution of an HCT-treated immunodeficient patient must be made before live viral vaccines are administered.” ■

AutoimmuneCorner

Research

New Study Links Autism and Autoimmunity



A new large-scale study of more than 2,700 mothers of children with autism shows that about one in 10

mothers have antibodies in their bloodstream that react with proteins in the brain of the babies. The study, led by Dr. Betty Diamond, head of the Center for Autoimmune and Musculoskeletal Disorders at The Feinstein Institute for Medical Research in Long Island, N.Y., shows that while the blood-brain barrier in adult women prevents them from being harmed by the antibodies, that same filter in the fetuses is not devel-

oped well enough and may allow the anti-brain antibodies to pass through the babies' brains, possibly causing autism. According to Dr. Diamond, the large sample size in the study "gives a clearer impression of the prevalence of these antibodies."

Approximately 50 million Americans live and cope with autoimmune disease, 75 percent of whom are women. The study was published in the Aug. 20 edition of *Molecular Psychology*. ■

Research

Autoimmune Disorders Associated with High Rates of Depression

Two recent studies show that autoimmune conditions such as rheumatoid arthritis and psoriasis are associated with high rates of depression. The findings suggest the impact on mental health, as well as the chronic pain and fatigue association with the conditions, could be much larger than previously estimated.

In one observational study (published in the journal *Arthritis Care and Research*) of 322 patients with severe rheumatoid arthritis who were waiting to go on biologic therapy, researchers investigated the impact of psychological factors upon each of the different parts of the current measure of disease, the DAS28. The DAS28 score takes into account the number of tender and swollen joints and the level of inflammation in the body and also includes a subjective, patient-reported measure based on how well the patient is feeling. The researchers found that subjective measures of

response were more likely to be influenced by psychological factors such as mood or beliefs about their illness and the therapies used.

"This may seem obvious, but it has not been reported before and is important because without treating the depression, the patient's DAS28 score might not improve as much as it should on a biological drug, and doctors may assume the drug is ineffective," explained Dr. Lis Cordingley, a health psychologist who was the lead author of the study. As a result of this study, researchers at the Arthritis Research UK Center for Genetics and Genomics at the University of Manchester in the United Kingdom say that patients with severe active disease who are waiting to go onto a biological therapy should be routinely screened for depression.

Research has also shown that those with psoriasis have a 39 percent increased risk of depression. In a study published in the *Journal of*

Dermatological Treatment, researchers looked at how certain biological therapies may also have an impact on depression in psoriasis patients. They reviewed various studies correlating increased levels of cytokines to depression and concluded that TNF inhibitors, frequently used in the treatment of psoriasis, may be helpful in directly reducing depressive symptoms for patients with psoriasis and other chronic inflammatory conditions.

Individuals with psoriasis are now being asked to take part in a new study at St. Vincent's University Hospital that is looking at the effects of psychological intervention known as mindfulness, which aims to develop awareness of the present moment through the practice of a form of meditation. Mindfulness has been found effective for improving mental health by reducing the symptoms of depression, anxiety and stress, as well as for improving health-related quality of life. ■

Research

TB Vaccine May Stall MS Progression

A recent study shows that a vaccine typically used to prevent tuberculosis in countries outside of the U.S. could also prevent multiple sclerosis (MS) in people who are in the beginning stages of the disease. In the study, researchers looked at 73 patients who showed early signs of MS, 33 of whom received one injection of the Bacille Calmette-Cuerin (BCG) vaccine, while the others received a placebo. After six months of brain scans, all the participants received

another MS drug called interferon beta-1a for one year, followed by whatever MS drug their neurologist prescribed. Immediately following the BCG vaccine, all patients were evaluated for definite MS for five years. Six months into the study, patients who received the vaccine had a lower-than-average number of brain lesions (three) that are indicative of MS compared with the placebo group that had seven lesions. No major differences in side effects were noticed

between the two groups by the end of the study. All together, 58 percent of the vaccinated group hadn't developed MS, which was almost twice that of the placebo group (30 percent). Typically, half of all patients in the early stage of MS, known as the clinically isolated syndrome, develop a clinically definite form of MS within two years of diagnosis, while 10 percent remain unchanged. The study was reported on in the Dec. 4 issue of *Neurology*. ■

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Research

Gene Therapy May Lead to a Long-Term Cure for SCID-X1



An international consortium of scientists has perfected gene therapy in promising clinical trials that they say may lead to an eventual long-term cure for X-linked severe combined immunodeficiency (SCID-X1). The mechanism used to deliver the gene therapy is designed to prevent the serious complication of leukemia that developed in one-quarter of boys treated a decade ago in a similar trial in Europe.

In the new trial, eight of nine boys (all between the ages of 9 months and 36 months) are alive and well with functioning immune systems and free of infections associated with SCID-X1 following treatment. Of the

eight patients, seven are actively producing T cells, and six of seven have met the trial's primary endpoint: a T-cell count greater than 300 cells per microliter of blood and T-cell proliferation in response to stimulation with phytohemagglutinin. The one patient among the seven who is producing T cells but has not yet achieved 300 cells per microliter was scheduled to receive a second round of gene therapy in January. The investigators are continuing to monitor all of the children for signs of treatment-associated leukemia, which developed three to five years post-treatment in the prior trial.

At the heart of the trial is a self-inactivating vector used to ferry the genes for the IL-2 receptor gamma subunit (IL2RG) into a patient's hematopoietic (blood-forming) stem cells. Once the gene is inserted, the cells are returned to the patient. IL2RG fuels the development and growth of the immune cells and is a key component of normal immune system development. In children born

Did You Know?

Washington added severe combined immune deficiency disease (SCID) to its list of conditions that all newborns in the state are screened for at birth beginning January 1.

with SCID-X1, the gene carries a mutation that renders it inactive. The viral vector used in the study is a modified gammaretrovirus, a member of a family of viruses able to insert genetic cargo into the genome of mammalian cells and drive expression of the inserted genes. The vector has been engineered to avoid the leukemia that halted the previous SCID-X1 gene therapy effort. Analyses of T cells from the participants in the trial suggest that the new vector avoided genomic sites known to contribute to leukemia development.

"These results show that the new vector appears to retain efficacy and, at least in preliminary studies, may be safer," said David A. Williams, MD, a leader of Dana-Farber/Boston Children's, who is chief of the Division of Hematology/Oncology at Boston Children's Hospital, associate chair of pediatric oncology at Dana-Farber Cancer Institute and principal investigator for the gene therapy trial's U.S. sites. The study's U.S. sites are supported by the National Institute of Allergy and Infectious Diseases and the National Heart Lung and Blood Institute's Product Assistance for Cellular Therapies Program. The findings were presented at the 55th annual meeting of the American Society of Hematology on behalf of the Transatlantic Gene Therapy Consortium. ■

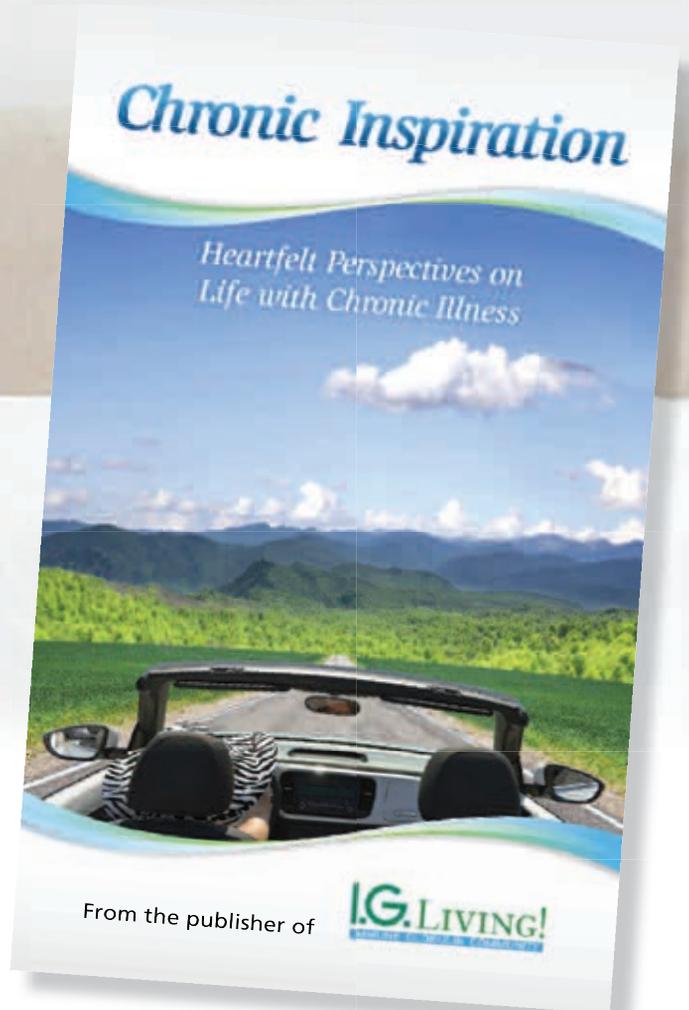
People & Places

Idera Pharmaceuticals Inc. has been issued a patent by the U.S. Patent and Trademark Office for IMO-8400, the company's lead drug candidate for treating autoimmune diseases. IMO-8400, which is an antagonist of Toll-like receptors 7, 8 and 9, is being evaluated in a Phase II trial in patients with moderate-to-severe plaque psoriasis. Patent number 8,486,908 protects

both composition of matter and method of use for IMO-8400.

UCB and Sanofi have entered into a collaboration for the discovery and development of innovative anti-inflammatory small molecules that have the potential to treat a wide range of immune-mediated diseases in areas such as gastroenterology and arthritis.

Discover the “must read” new eBook from the publisher of *IG LIVING* magazine



*“You can lament what is lost to you, whether it’s opportunity, a person or your health, but clinging to anger is no way to experience life.” – Rebecca Zook in “Life Lessons,” excerpted from **Chronic Inspiration**.*

Download a daily dose of inspiration with this heartfelt compilation of writings on life with chronic illness. From coping strategies and parenting tips to “from the trenches” advice on dealing with family and friends who simply don’t get it, these personal stories are sure to uplift, challenge and inspire. Honest and candid, *Chronic Inspiration: Heartfelt Perspectives on Life with Chronic Illness* gives voice to those who refuse to let their diagnosis define who they are or what they can accomplish.

“I encourage all in our community to read this and pass along to anyone who needs to better understand the emotions and difficulties of the chronic care community.”

— Dominick Spatafora
President, Neuropathy Action Foundation

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Making Kids TinySuperheroes

One woman is leading a crusade to empower sick kids, and she's doing it with help from all over the world.

By Ronale Tucker Rhodes, MS

KIDS WHO ARE ill need a lot of things. They need hugs and compassion. But, most importantly, they need to feel good about themselves. And, that's the mission of Robyn Rosenberger's TinySuperheroes, which seeks to "empower extraordinary kids — one cape at a time." Through her efforts, Robyn is helping thousands of kids around the world who "exemplify strength and determination by overcoming illness and disability." But, she doesn't do it alone; she needs and receives a lot of help.

What Is TinySuperheroes?

TinySuperheroes was launched by chance in January 2013. It all started in August 2012, when Robyn decided to sew capes for her nephew's birthday. During the same period, Robyn had been following a blog written by a mother of a child, Brenna, who suffers from a rare genetic skin disorder. After seeing how cute her son, nephew and dog looked in the capes, Robyn decided to make one for Brenna, too. "I just wanted to send them something to let them know how we saw her," explains Robyn. "The way we saw her was extraordinary. We were amazed with her strength. In the beginning, it was just a token. I wanted them to feel recognized and [to let them know] that it's really amazing what they've been through." She then reached out to a few other kids through their

Facebook pages and sent them capes, and TinySuperheroes was born. "These kids took an identity from [the cape]. They wear it to school, to doctor visits, etc. It resonated with me that this was something that could empower them," she says.

Robyn refers to TinySuperheroes as "a tiny company with a *super* big mission!" These kids "spend so much of their life doing things that aren't typical for kids their age. They're in the hospital instead of in school or recovering from surgery instead of playing T-ball," says Robyn. "So, they spend life experiencing what's different about them. Everyone wants to identify with something bigger than themselves. These kids want to identify with something that is normal. My goal is to turn things that are different about them into what makes them amazing. [These capes] give them confidence and strength and pride that their lifestyle doesn't automatically give to them."

Connor Wobbe felt just that way when he received his TinySuperheroes cape last year. Connor, who is 14 and has a primary immunodeficiency disease, says "It made me feel like someone actually cared that I was sick." According to Connor's mother, Nicole, the cape "touched his heart and made him feel very appreciated." Because Connor doesn't appear to be sick, it made him feel that someone outside of his family recognized his



Connor Wobbe, who suffers from a primary immunodeficiency, received a TinySuperheroes cape, which inspired him to raise enough money to buy capes for 18 other ill children.

illness. In fact, the cape made Connor feel so good that it inspired him to raise money for capes for other kids. "I just thought other kids would enjoy having a cape. I raised money for my birthday, and we also did a blood drive to raise money," explains Connor. "At his birthday party, he invited his whole class and asked them to bring a donation instead of bringing presents," adds Nicole. Connor raised enough money to purchase capes for 18 kids. "Connor is such an amazing young man who never lets any obstacles get in his way," says Nicole. "He has missed out on so much and may get a little disappointed, but truly just rolls with the punches. Connor is our Superhero; he not only fights a daily battle, but he does so with a smile on his face. Being able to surprise Connor with the cape and seeing the smile brought to his face brought tears to my eyes."

How Can People Help?

By August 2013, TinySuperheroes had sent more than 2,000 capes to children in 15 countries and all 50 states. But, they're expensive. That's why TinySuperheroes is a "socially conscious company," says Robyn. It's a for-profit business that covers the cost of the cape and the cost of running the business. Individuals can purchase a cape for a special child they know who has overcome adversity. They also can sponsor a cape to go to any child, or they can purchase a cape for one of the children on the TinySuperheroes wait list.

In addition to empowering kids with capes, TinySuperheroes seeks to spread awareness of the many serious illnesses that many kids face. It does this with what Robyn calls the company's "missions." "My goal was to create a community for these families," explains Robyn. "When kids get a cape, they become a part of the TinySuperheroes Squad. I didn't want the cape to be the end. This is something they're a part of, and it's active. The missions are an effort to keep the community alive and to bring in people who don't have a TinySuperhero." In 2013, TinySuperheroes held two missions. The first introduced people to the TinySuperheroes Squad. The second, held during the holidays, was to "send good cheer" by creating and sending a card of encouragement to kids in the TinySuperheroes Squad. "For the holidays, we had 200 people apply for [the mission], and they all made greeting cards for our Superheroes to let them know that people care and are following their story, and that we're all cheering for them," says Robyn.

In 2014, there will be a mission every month, and as of this writing,

TinySuperheroes was conducting its third mission during the month of January titled Ichthyosis Awareness: Teaching that Different Is Beautiful! Ichthyosis is the disease that Brenna suffers from. Completing the mission requires four tasks: 1) signing up for the mission on the TinySuperheroes website, 2) giving stickers supplied by TinySuperheroes to local heroes in the community, and telling them about TinySuperheroes and how it is taking action to help those with ichthyosis, 3) taking a photo with each hero, and posting it on the TinySuperheroes Facebook page that includes a sentence about why they are a hero, and 4) copying and pasting the www.tinysuperheroes/missions link into one's Facebook page to help the company spread the word about how the TinySuperheroes Squad is taking action to raise awareness and how others can help.

experience with this community. And, having a kid myself, I feel so blessed to get a look at this community. We're all fragile, and we're all facing death, and we live in denial of that. And, these families have found a way to face that reality and have learned to appreciate what they have. These are the happiest families I've ever met. A lot of stories are sad. People ask me all the time: 'How do you deal with hearing these stories every day?' That's not my experience at all. These families aren't putting out sadness; they're putting out faith and joy. It's taught me that life is short, and what we have today is what we have. And, they've learned that, and it's something we all could learn from."

Where is TinySuperheroes headed in the future? "I don't know," says Robyn. "I never expected to be where we are right now. I'm just trying to keep up with it. It grew exponentially

In addition to empowering kids with capes, TinySuperheroes seeks to spread awareness of the many serious illnesses that many kids face.

Faith Will Lead the Way

Robyn has now empowered many thousands of ill children through her TinySuperheroes company. But, Robyn has also been empowered: "It's changed my life in every way. Practically speaking, it's changed what I do. I left my job last June to keep this going. But, really what it's changed is my heart and my perspective. Prior to this, I didn't have a lot of

in a way we were not prepared for. I feel really blessed to be the vehicle that is driving it. But, I'm putting faith in that it will lead its way and take whatever direction it needs to take." ■

RONALE TUCKER RHODES, MS, is the editor of IG Living magazine.

Editor's note: To get involved with TinySuperheroes, visit the website at www.TinySuperheroes.com.

Exercise Success



While illness places limits on athletic competition, success should truly be measured by doing the best we can with what we have.

By Matthew D. Hansen, DPT, MPT, BSPTS

Competition, to varying degrees, is an innate drive that is found in all of us. As we age and mature, many of us learn to control the drive appropriately, so that for the benefit of society, as well as our own sanity, not everything becomes a competition. For instance, I don't grab for the last dinner roll or piece of pizza, pushing my children's and wife's hands aside as I used to do with my teenage brothers. It wouldn't do me much good if I did; at the very least, I could count dessert as a forgone possibility.

In some arenas of life, however, including athletics, structured competition can be beneficial to our personal progress and well-being. Competition drives us to perform and can boost self-confidence and bring us a sense of accomplishment. Many patients with a primary immunodeficiency (PI) or autoimmune disease may have considered athletic competition to be an integral part of their lifestyle prior to diagnosis, and now feel discouraged thinking that they may never be able to compete again.

Defining Success

Before presenting any suggestions or resources for participating in athletic competition, I ask you to indulge me in a moment of personal philosophy. Life is a competition of endurance, made up of individual battles, not so much with outside opponents as with our own expectations of ourselves. Worthy opponents frequently extend our own tape measures of success, albeit sometimes equivocally, but true success is a matter of perspective. When everything is said and done in life, I believe that our success will be measured by whether or not we did the very best we could with what we had. Not many people are born with perfect health or a 170 IQ, but we're all blessed with various attributes that can be used to better ourselves and the world around us.

I was a pretty good basketball player by most standards, but my dad told me that there would always be someone, somewhere, who was bigger, stronger and more naturally talented than I was. "The important thing," he would say, "is that you always give your best effort, and that even when you lose, you're not left beaten." We all have "losses," but we can't allow them to beat us. I still enjoy playing basketball. Although I'm no longer able to play at the same competitive level that I once did due to the natural effects that time takes on the body, I still do my best. The difference is that I no longer measure my success by the competition (I can't, because I'm not quick enough to keep up with many young high school players who I

would have easily bested in my prime). Instead, I measure my success by my effort and what I know I am capable of doing; I've also adapted my game to become a better outside shooter instead of driving to the basket as much as I used to.

I understand that some patients reading this article are thinking, "I wish that I could play basketball at all," but the principle is the same. Success should take into consideration our current abilities and our best efforts. The famous sports broadcaster, Howard Cosell, stated, "Ultimate victory in competition is derived from the inner satisfaction of knowing that you have done your best and that you have gotten the most out of what you had to give." My sentiments exactly. Now, it's time to offer some encouraging news.

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Competing with One's Best

Some patients living with a PI or autoimmune disease may still be capable of competing in recreational athletics if care is taken not to exhaust the immune system by over-training. Days of running half marathons may be over (don't be too discouraged, most people never see those days), but a 5K or 10K race may be very achievable. The key is progressive training and understanding that there may be some minor setbacks along the way. Physical therapists and/or athletic trainers can help to develop a flexible, personalized training program. Just be sure they understand the diagnosis! Patients aren't looking for a boot camp instructor. Whatever activities patients are training for (e.g., jogging, biking, walking, swimming, tennis), progress should be gradual. For example, if they were typically able to jog for seven minutes without exhausting themselves or their immune system, they may want to try jogging for seven-and-a-half minutes the next time. If they do too much, their body will let them know.

They shouldn't get discouraged; they've just set a new goal to work toward.

Patients should be encouraged to find joy in the journey and not to make the race itself the end-all and only measure of success. Too many people with a PI have trained for months, only to have their plans for a big race crushed by a late respiratory infection. Others have started the race, only to discover that they aren't able to finish. We all "finish" what we are doing; the question is how we finish. The effort that we give is sometimes more important than where we end up. Who is to say that the person who finished their race on a given day by running to the

Patients should be encouraged to find joy in the journey and not to make the race itself the end-all and only measure of success.

10th oak tree on the left wasn't more successful than the Ironman who finished at the tape line after running a "lazy" race with an average time of seven minutes per mile?! Even if we had set out for the tape line, goals change. I wanted to be an NBA basketball player more than anything in the world (my tape line), but that goal changed. It doesn't matter whether it was because I physically couldn't reach the line or because my priorities in life changed, because I'm the best physical therapist that I can be and very happy about it. The same determination and adaptability that helped me to become a great basketball player helped me to become a great physical therapist.

Where to Compete

There are quite a few groups that exist to help people with different physical ailments and/or disabilities to participate in organized sport. It's not too difficult to find a local adaptive basketball wheelchair league or amputee swimming team. Unfortunately, it's much more difficult to

find organizations that focus on less visible diagnoses such as a PI. One resource is the National Center on Health, Physical Activity and Disability, whose website, www.ncpad.org, has a state-by-state directory of different programs, organizations and even personal trainers who "self-report their experience and qualifications in working with clients with disabilities and health conditions."

Another great organization is Disabled Sports USA. The non-profit's goal is to "improve the lives of wounded warriors, youth and adults with disabilities by providing sports and recreation opportunities." Its website, www.disabledsportsusa.org, has a directory of local chapters throughout the U.S. Again, many of these chapters have a focus on some of the more visible physical disabilities; however, some of them have also done work with athletes who live with multiple sclerosis and several other autoimmune or PI conditions.

One of the most unique methods for patients to remain physically competitive is through home technology. Several video game consoles (e.g., Nintendo Wii, Microsoft Xbox 360 and Sony PlayStation 3) now have interactive components that allow the player to participate/compete in a number of athletic and adventure games through motion-controlled, active play. The consoles can detect the physical movements of a person standing in front of the television screen, and move the figure on the video game accordingly. In this manner, players can enjoy competing from the comfort and safety of their own home or other controlled environment. I've participated with patients in virtual track and field events, boxing, basketball, bowling and even Star Wars light saber duels.

The greatest thing about using the optional Internet feature with a video game console is that you can play against people with similar skill levels from anywhere in the world while using an alias for your computer figure. The other players have no way of knowing who you are, and you don't know who they are. Your worthy opponent could be the real Muhammad Ali playing from his home in the U.S., or it could be a 7-year-old girl in Beijing, China. Video games can be used to help train for outside events or used as competition themselves. If you get tired, simply end the game. Just don't pull the cord out of the wall when others are still playing; my brother used to do that when he was losing, and it drove me crazy!

For patients who don't feel they need to be competing against other people, the possibilities for personal competition are endless. I've set a lot of little goals for myself throughout the years and tried to reach beyond the mark



(e.g., seeing how high I could climb a mountain near our home, how many push-ups I could do in a row, how long I could run without stopping). I recommend that patients set the same type of goals, with one additional suggestion: They shouldn't push too hard to beat the mark each time that they compete. They should set their own baseline (e.g., I'm going to climb at least one set of stairs, because I know that I can usually do that easily). If they don't feel up to the baseline, they probably shouldn't be exercising that day. If they do choose to exercise and at least meet the baseline, that's a "win." As patients become more consistent with meeting the baseline, gradually advancing that baseline as they are able, they will see the mark advance as well (it's OK for the baseline to move forward or backward as needed). Patients can keep a log of their accomplishments — not just of the new records, but more importantly, of how many times they are able to meet the baseline.

Life Is a Competition

Life is a competition of endurance, made up of individual battles, not so much with outside opponents as with our own expectations of ourselves. True success is a matter of perspective. It will be measured by whether or not we did the very best that we could with what we had.

On the 30th anniversary of her 1968 Olympic gold medal win, renowned figure skater Peggy Fleming had surgery to remove a malignant tumor in her breast. It's been years since she has skated competitively; however, since the time of her diagnosis, Peggy has dedicated time to tirelessly champion health causes. She is still a fierce competitor in life, striving to better herself and the world around her at every opportunity that she gets. She once said, "I think exercise tests us in so many ways — our skills, our hearts, our ability to bounce back after setbacks. This is the inner beauty of sports and competition, and it can serve us all well as adult athletes."

Patients should be supported in working toward being happy with who they are and with maximizing use of the tools that are at their disposal. As they celebrate their successes, however small they might seem, they will be surprised with how much they really do have to offer to themselves and the world. ■

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Diagnosing and Treating Common Variable Immunodeficiency



While the outcome for patients with this not-so-common disease has improved, it often takes six to eight years to diagnose, putting them at risk for high-risk complications.

By Ronale Tucker Rhodes, MS

When Tami Slaats's doctor calmly told her she had a primary immunodeficiency disease (PID) known as common variable immunodeficiency (CVID), she assumed he would tell her how this disease could be cured because she heard the word "common."¹ Yet, despite its name, CVID is anything but common; it is listed as a "rare disease" by the Office of Rare Diseases (ORD) of the National Institutes of Health (NIH), which means the disease affects fewer than 200,000 people in the U.S.² The true incidence and prevalence of CVID are difficult to ascertain. But, it is estimated that Tami, who lives in the U.S., is one in 10,000 to 50,000 people who are diagnosed each year in Europe and North America — a figure that is thought to underestimate its prevalence, or the estimated population of people who are managing disease at any given time. A nationwide survey in the U.S. suggests a population prevalence of all diagnosed PIs of approximately one in 1,200, 35 percent of whom are supposed to suffer from CVID.³

What Is CVID?

CVID has been recognized as the most common symptomatic form of antibody deficiency diagnosed in adulthood since it was first described in 1953.⁴ It is the second most common PI (second to selective immunoglobulin A [IgA] deficiency),⁵ but clinically the most significant because of its prevalence, complications, hospitalizations and requirement for lifelong therapy.⁶

The one key characteristic of CVID is an impaired ability to produce antibodies (also called immunoglobulins, or Igs, which are proteins in the body that help ward off infection).³ The medical terms for absent or low blood Igs are agammaglobulinemia and hypogammaglobulinemia, respectively. Deficiencies previously known as late-onset hypogammaglobulinemia and adult-onset hypogammaglobulinemia are now considered part of CVID.⁵ All CVID patients have low serum IgG, and often they have low IgA and/or IgM, as well as an impaired or even absent antibody response to vaccines. In comparison with healthy individuals, the number of B cells and in vitro function may be normal or low, and CD4 and/or CD8 T cell numbers may be normal or low.⁷

The disease affects both genders equally. The onset is typically in young adulthood, but the disorder is found in younger children as well.⁸ A diagnosis of CVID is typically not made until the third or fourth decade of life, but about 20 percent of patients have symptoms of the disease or are found to be immunodeficient in childhood.⁹

Causes of CVID

In 75 percent to 80 percent of cases, the cause of CVID is unknown; however, in 10 percent to 20 percent of cases, a genetic cause has been identified.¹⁰ Most cases are classified as sporadic and occur in people with no apparent history of the disorder in their family. It is believed that sporadic cases probably result from a complex interaction of environmental and genetic factors.¹¹

Recently, studies have shown the involvement of a small group of genes in a few patients. These include inducible co-stimulatory (ICOS) and a few other proteins on B cells, which appear to be causes of autosomal recessive CVID.⁹ When CVID is inherited in an autosomal recessive pattern, the patient inherits a copy of the mutated gene from both parents, but typically, the parents don't show signs and symptoms of the condition. In very rare cases, CVID is inherited in an autosomal dominant pattern, which means one copy of the altered gene in each cell is sufficient to cause the disorder. However, not all individuals who inherit a gene mutation associated with CVID will develop the disease. And, in many cases, affected children have an unaffected parent who shares the same mutation.¹¹

In 75 percent to 80 percent of cases, the cause of CVID is unknown.

Approximately 10 percent of affected individuals have a mutation in the TNFRSF13B gene (tumor necrosis factor receptor superfamily, member 13B). The TNFRSF13B gene provides instructions for making a protein called transmembrane activator and calcium modulator and cyclophrenia ligand interactor (TACI). The TACI protein is found on the cell membrane of B cells. When B cells mature, they produce Igs (antibodies) that attach to specific foreign particles and germs, marking them for destruction. More than 25 mutations in the TNFRSF13B gene have been associated with CVID. It is not clear how mutations in the gene prevent normal B cell maturation and antibody production. Some people with TNFRSF13B gene mutations develop CVID, but others do not.¹²

Symptoms of CVID

CVID symptoms vary widely, and they manifest in a range of severity.¹⁰ The degree and type of deficiency of serum Igs, and the clinical course, varies from patient to patient, hence, the word “variable.”⁹

The major characteristics of CVID are bacterial and viral infections. The most common infections are sinopulmonary and include *Streptococcus pneumoniae*, *Haemophilus influenzae*, *Klebsiella pneumoniae* and, sometimes, mycoplasma infections. Recurrent sinopulmonary infections can begin any time, but more commonly, they are preceded by a long period in which infection frequency and severity are normal or near normal.¹³

Individuals may also experience meningitis or other systemic bacterial infections, recurrent eye or skin infections or gastrointestinal (GI) symptoms related to compromised immune/gut homeostasis, including chronic diarrhea, malabsorption or bloating. Twenty percent of CVID patients experience infections and inflammation with associated malabsorptive symptoms. Forty percent of dyspeptic individuals with CVID experience *H. pylori* infection. And, 2 percent to 3 percent of CVID patients experience chronic gut inflammation and associated malabsorption, which is not responsive to a gluten-free diet.¹³

Complications of CVID

In addition to the recurring symptoms caused by CVID, there are many problematic complications that often occur in CVID patients, many of which are due to a diagnostic delay, including chronic lung disease, granuloma formation, lymphoid hyperplasia and infiltrative disease, GI disease, autoimmunity and the development of cancer. Indeed, these causes appear to be the major cause of morbidity and death in patients with CVID.⁶

CVID symptoms vary widely, and they manifest in a range of severity.

Infections of the respiratory tract are the most common symptom of CVID, occurring in up to 73 percent of patients. Continued respiratory tract disease can lead to chronic lung damage, and in the more severe cases, it necessitates continuous oxygen treatment and/or heart or



lung transplantation. The question is whether the damage is due to infections prior to diagnosis, continued low-grade infections that are not adequately addressed by treatment or a combination of the two.⁶

Granuloma formation, caused by lymphoid aggregates within the tissues, occurs in approximately 25 percent of patients with CVID. These granulomata resemble those found in both sarcoidosis and Crohn’s disease, which can affect several organs. Granuloma formation may commonly manifest in skin and subcutaneous tissues, the GI tract and lungs.¹⁴ In some patients with granuloma formation, it is accompanied with an intense lymphoid infiltration, which leads to what is known as granulomatous lymphocytic interstitial lung disease. One study of CVID patients with interstitial lung disease reported a median survival of 13.7 years compared with 28.8 years in those without this complication.⁶

There also is an increased incidence of malignancy. Cervical, mediastinal and abdominal lymphoid hyperplasia and enlarged spleen are found in at least 20 percent of CVID patients. Lymphoid infiltrates occur in lung or other organs such as the liver or kidneys.⁶ These patients are also susceptible to lymphomas. In one study, it was found that nearly 8 percent of patients developed non-Hodgkins lymphoma, another 1 percent to 2 percent had Hodgkins lymphoma, and other individuals had 24 different cancers, including breast cancer, prostate cancer, squamous cell carcinoma, melanoma and basal cell carcinoma.¹³

The GI tract is the largest immune organ in the body, so

it is expected that an immunodeficiency will affect it in some way. GI complications for CVID patients are variable, and tend to mimic known diseases such as celiac, pernicious anemia and inflammatory bowel disease. Many studies confirm a high prevalence of inflammatory, malignant and infectious GI disorders in CVID patients, which are thought to be caused by T-cell-mediated defects.⁵

Up to 25 percent of CVID patients develop autoimmune disorders, mainly immune thrombocytopenia (ITP), autoimmune hemolytic anemia (AIHA) or both (Evans syndrome), and more rarely, autoimmune neutropenia. CVID patients with ITP or Evans syndrome tend to be younger than those who develop AIHA. Other autoimmune diseases that occur in CVID patients include pernicious anemia, rheumatoid arthritis, Sjögren's syndrome, vasculitis, thyroiditis, alopecia, vitiligo, hepatitis, primary biliary cirrhosis, uveitis, sicca syndrome and systemic lupus erythematosus.⁶

Diagnosing CVID

Currently, there is a six- to eight-year delay in diagnosing CVID, which puts patients at considerable risk since the prevention and control of long-term complications such as bronchiectasis critically depend on early diagnosis and treatment.³ Most patients are diagnosed with CVID between the ages of 20 years and 40 years, but approximately 20 percent are diagnosed under age 20. Generally, physicians are reluctant to diagnose CVID in young children (under age 4) because physiologic immaturity can mimic the disease in the early years; also, this period of delay allows other diagnostic considerations to be explored.¹⁵

Physicians should suspect a PI in children or adults who have a history of recurrent bacterial infections involving ears, sinuses, bronchi and lungs.⁹ CVID is primarily diagnosed by testing for low serum IgG concentration, which ranges from profoundly reduced (less than 100 mg/dL) to just below the normal range in adults (800 mg/dL to 1,200 mg/dL). Those with the disease also don't mount an antibody response to the pneumococcal vaccine. Other abnormal laboratory studies that would suggest a CVID diagnosis include reduced serum concentrations of other Igs, especially IgA or IgM, and reduced numbers of switched memory B cells as assessed by peripheral B-cell immunophenotyping. For some inherited causes of CVID, testing for loss of protein expression and/or molecular genetic testing to identify causative mutations is possible.¹³

The diagnosis of CVID requires low serum IgG, low serum IgA and/or IgM and decreased response to the pneumococcal vaccination. A normal response to the pneumococcal vaccine is greater than 50 percent for children 2 years old to 5 years old and greater than 70 percent for individuals 6 years old to 65 years old.

Treating CVID

To prevent recurrent sinopulmonary infections and chronic lung disease, immune globulin (IG) replacement therapy is given either intravenously (IVIG) or subcutaneously (SCIG). Generally, IVIG is dosed at 500 mg/kg every three to four weeks. SCIG also can now be performed with a variety of dosing schedules, from every two weeks, weekly and even semi-weekly, to suit the preference of the patient.¹⁶

Currently, there is a six- to eight-year delay in diagnosing CVID.

Prophylactic antibiotics may help patients with chronic sinusitis or chronic lung disease. Antibiotics also may help control GI tract complications. For those with bronchiectasis, physical therapy and postural drainage will remove the secretions from the lungs and bronchi.¹⁶

Other complications of CVID are most often prescribed the standard treatments. However, higher doses of IG therapy are often prescribed to treat chronic lung disease, lymphoid interstitial disease, granuloma and some autoimmune disorders.⁶

For all patients, regularly scheduled follow-up is mandatory since new problems may arise and evolve over time. Stable patients should be seen at least yearly, while those with complications should be seen at shorter intervals such as every three to six months.⁶ During these visits, several surveillance methods can be performed, including periodic complete blood cell counts and differential white blood cell counts to detect lymphoma, annual pulmonary function testing (beginning in children at age 8 years to 10 years), high-resolution CT scans every two to three years to follow progression of lung disease, biopsy of enlarged lymphoid tissue, and other imaging

techniques for the assessment of granulomatous disease and GI complications.¹³

Allogeneic stem cell transplantation (ASCT) has occasionally been reported as treatment in cases of occurring malignancy in CVID patients, but the outcome of ASCT as a potentially curative approach in CVID with poor prognosis is unknown. In a study on the feasibility and outcome of ASCT in four patients suffering from severe CVID, one patient died after three months due to overwhelming hemorrhagic pneumonia. However, at the time the study was reported on, the other three patients had survived between 4.5 years and 7 years. One patient is cured from his lymphoma and the underlying CVID; he is free of recurrent infections, has normalized his serum Ig levels (except IgA) and responds for the first time to vaccines. In the other two patients, ASCT reached the end point of resolving the lymphoproliferative disease in one, and reducing the steroid dose with stabilization of pulmonary disease in the second, but had little effect on the underlying immunodeficiency. The researchers note that the importance of the study lies in the evidence that CVID can be cured in selected patients.¹⁷

IVIG in high doses can be a very effective treatment.

Improving CVID Outcomes

The outcome for CVID patients has greatly improved over the years, and the majority of patients can expect to have stable lifestyles. However, diagnosis is still delayed six to eight years after characteristic symptoms, which poses a high risk of long-term and severe complications from CVID. Therefore, one of the most important goals is to reduce the time it takes to diagnose the disease. Certainly a greater awareness about the disease can help. For the low percentage of at-risk individuals who have relatives with CVID, clinical surveillance may allow timely intervention and improve outcome. On a case-by-case basis, molecular genetic testing for early identification of at-risk family members may improve diagnostic certainty.¹³ ■

RONALE TUCKER RHODES, MS, is the editor of *IG Living magazine*.

References

1. Mitschang, T. Let's Talk. *IG Living*, April-May 2014. Accessed at www.igliving.com/Assets/IGL/eMagazine/CurrentIssue.
2. Prevalence and Incidence of Common Variable Immunodeficiency. RightDiagnosis.com. Accessed at www.rightdiagnosis.com/c/common_variable_immunodeficiency/prevalence.htm.
3. Bergbreiter, A, and Salzer, U. Common Variable Immunodeficiency: A Multifaceted and Puzzling Disorder. *Expert Review of Clinical Immunology*, 2009; 5(2): 167-180. Accessed at www.medscape.com/viewarticle/589907_2.
4. Myrtek, D, and Salzer, U. Common Variable Immunodeficiency: Genetic Insights Into a Complex and Baffling Disease. *The Rheumatologist*, June 2008. Accessed at www.the-rheumatologist.org/details/article/980299/Common_Variable_Immunodeficiency.html.
5. Kalha, I, and Sellin, JH. Common Variable Immunodeficiency and the Gastrointestinal Tract. *Current Gastroenterology Reports*, 2004, 6:377-383. Accessed at gastro.ucsd.edu/fellowship/materials/Documents/CVID.pdf.
6. Cunningham-Rundles, C. How I Treat Common Variable Immune Deficiency. *Blood*, July 8, 2010, vol. 116, no. 1 7-15. Accessed at bloodjournal.hematologylibrary.org/content/116/1/7.full.
7. Bonilla, FA, and Geha, RS. Common Variable Immunodeficiency. *Pediatric Research*, (2009) 65, 13R-19R. Accessed at www.nature.com/pr/journal/v65/n5-2/full/pr2009117a.html.
8. Yazdani, S, McGhee, S, and Stiehm, R. *Chronic Complex Diseases of Childhood: A Practical Guide for Clinicians*. Brown Walker Press, 2011.
9. Immune Deficiency Foundation. Common Variable Immune Deficiency. Accessed at primaryimmune.org/about-primary-immunodeficiencies/specific-disease-types/common-variable-immune-deficiency.
10. WebMD. Common Variable Immune Deficiency. Accessed at www.webmd.com/a-to-z-guides/common-variable-immunodeficiency.
11. Genetics Home Reference. Common Variable Immune Deficiency. Accessed at ghr.nlm.nih.gov/condition/common-variable-immune-deficiency.
12. Genetics Home Reference. TNFRSF13B. Accessed at ghr.nlm.nih.gov/gene/TNFRSF13B.
13. Scharenberg, AM, Hannibal, MC, Torgerson, T, Ochs, HD, and Rawlings, DJ. Common Variable Immune Deficiency Overview. In: Pagon, RA, Adam, MP, Bird, TD, Dolan, CR, Fong, CT, Smith, RJH, and Stephens, K (editors). *SourceGeneReviews* [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2014. 2006 Jul 05. Accessed at www.ncbi.nlm.nih.gov/pubmed/20301476.
14. Medscape. Chronic Granulomatous Disease. Accessed at emedicine.medscape.com/article/1116022-overview.
15. Cunningham-Rundles, C. The Many Faces of Common Variable Immunodeficiency. *American Society of Hematology Education Book*, Dec. 8, 2012, vol. 2012, no. 1 301-305. Accessed at asheducationbook.hematologylibrary.org/content/2012/1/301.full.
16. Rhodes, RT. Understanding and Treating Common Variable Immunodeficiency. *IG Living*, April-May 2012. Accessed at www.igliving.com/Assets/IGL/Articles/IGL_2012-04_AR_Understanding-and-Treating-Common-Variable-Immunodeficiency.pdf.
17. Transplantation in Common Variable Immunodeficiency — A New Therapeutic Option? *American Academy of Allergy, Asthma & Immunology*, Sept. 20, 2011. Accessed at www.aaaai.org/global/latest-research-summaries/Current-JACI-Research/transplantation-common-variable-immuno.aspx.



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Screening for Severe Combined Immunodeficiency Disease

By Amy Scanlin, MS

Screening for SCID saves thousands of newborns' lives each year, costs far less than screening later and has resulted in the identification of new types of this deadly disease.

Severe combined immunodeficiency disease (SCID) is a pediatric emergency caused by genetic defects that create havoc on the immune system. Babies born with SCID (about one in every 30,000 to 50,000) have little to no T cell function to defend them against infections, and if not treated during their first months of life, they will die. Those who do receive treatment have a 94 percent chance of survival.

Thankfully, about 55 percent of infants born in the U.S. are now screened for SCID (more than two million so far nationwide) and other conditions that cause low T cell counts. Screening is conducted using T cell receptor excision circles (TRECs) screening. The TREC assay allows for diagnosis of newborns before symptoms have developed, which results in lifesaving treatment.

TREC screening “is so important because this is the only way to determine if a child has SCID,” says Fred Modell who founded the Jeffrey Modell Foundation (JMF) with his wife Vicki in memory of their son who at age 15 died of an infection brought about by a primary immunodeficiency disease (PI). “These newborns look great; they have rosy cheeks and bright eyes, and without this testing, they will begin very early on to develop overwhelming infections, and they won’t survive. But, when we can screen for and identify SCID, we can do a bone marrow transplant very early on, and these kids don’t just improve, they are cured!”

What Is TREC Screening?

TRECs are DNA molecules formed within maturing T cells that develop in the thymus and are measured by a technique called polymerase chain reaction (PCR). Normal blood samples have one TREC per 10 T cells (a good result indicating a high rate of T cell development), while babies with SCID and other related conditions either have very low T cell numbers or lack T cells altogether.¹

TREC testing is conducted via dried blood that is collected onto filter paper from a heel stick. From this dried blood spot (DBS), a very small sample is punched out and tested. Testing is so efficient, that public health laboratories can actually test thousands of DBSs at one time.

If the result of the TREC assay indicates low levels of T cells or PCR failure, blood samples are drawn and a process called flow cytometry is performed to look at the total lymphocyte numbers and subsets of T, B and natural killer cells, as well as naïve and memory T cells.¹ Infants may also go through genetic testing.

Because early testing is conducted on such a large scale, the vast majority of tests will have a negative result. Babies who test positive, however, now have a very good chance of survival. It should be noted that premature babies have the highest incidence of false positives.

What Does the TREC Assay Identify?

“The TREC newborn screening has revealed a wide spectrum of non-SCID disorders and led to better understanding and treatment of these conditions,” says Emily Hovermale, senior public policy manager for the Immune Deficiency Foundation (IDF). “It has led to the discovery of new types of SCID that were not previously known, now being classified as variant SCID.”

The most widely recognized condition identified by TREC screening, and the most deadly, is SCID. There are several forms of SCID with the most common linked to the X

chromosome that mostly affects males as they only have one X chromosome, while females have two and are more commonly carriers. A TREC screen will also identify conditions such as Omenn syndrome (also known as leaky SCID), DOCK8 deficient hyper-IgE syndrome (also a severe PI), Trisomy 21 (Down syndrome), DiGeorge syndrome, CHARGE syndrome, Jacobsen syndrome and cartilage-hair hypoplasia, as well as low T cell counts brought on by other conditions such as neonatal cardiac surgery and leukemia.

About 55 percent of infants born in the U.S. are now screened for SCID (more than two million so far nationwide) and other conditions that cause low T cell counts.

SCID is a PI that at first appears to be persistent infections that don’t resolve with normal treatments. Symptoms including severe respiratory infections, rashes that look like eczema, persistent diarrhea, thrush and pneumocystis pneumonia, which is the real red flag infection that should be an indicator a baby should be tested for SCID. Babies with SCID have severely impaired immune systems that cannot fight off infections and may have multiple hospitalizations before they are diagnosed, assuming they are not identified early through the TREC assay.

Omenn syndrome (leaky SCID) is caused by a mutation in the RAG1 or RAG2 genes and is characterized by peeling and red skin, diarrhea, enlarged lymph nodes, as well as elevated IgE levels and T cells (though the T cells are not functioning properly). Low IgA, IgG and IgM levels and virtually no B cells are also indicators. Babies with Omenn syndrome develop similar infections to other types of SCID and have the same prognosis if not identified and treated early.

Dock8 deficient hyper-IgE syndrome (HIES) is a rare PI characterized by skin conditions such as eczema and abscesses, as well as high levels of IgE. Mutations and

deletions in the DOCK8 gene are found to be the cause of most of the autosomal recessive (type 2) type, while the genes encoding the transcription factor STAT3 (not identified by the TREC assay) cause most of the more common autosomal dominant (type 1) type. Dock8 HIES is an inherited trait in which 95 percent of those affected have an error in chromosome number 9.

Trisomy 21 (Down syndrome) is a genetic condition in which one has 47 chromosomes instead of 46. In most cases, chromosome 21 is the error. It is one of the most common birth defects and may include additional conditions such as cataracts, dementia, hearing problems, breathing problems causing sleep apnea, as well as an underactive thyroid.

The most widely recognized condition identified by TREC screening, and the most deadly, is SCID.

DiGeorge syndrome (22q11.2 deletion syndrome) is one of the most common conditions identified, and is a syndrome with more than just SCID, according to Michael S. Watson, PhD, FACMG, executive director of the American College of Medical Genetics and Genomics. It may first become evident through a variety of types and severity of issues such as a heart defect, cleft palate and impaired immune function, as well as behavioral problems. Babies appear weak with poor muscle tone and have trouble gaining weight due to difficulty feeding and frequent infections.

CHARGE (coloboma, heart defect, choanal atresia, retarded growth and development, genital abnormality and ear abnormality) syndrome often results in multiple life-threatening conditions such as a hole in an area of the eye called the coloboma, narrowed (choanal stenosis) or completely blocked (choanal atresia) nasal passages, and cranial nerve damage that diminishes sense of smell. It can cause facial paralysis and hearing loss.

Jacobsen syndrome (11q terminal deletion disorder) is caused by loss of genetic material from chromosome 11. While symptoms vary considerably, most have delayed developmental growth and possible learning disabilities. As children develop, they may have heart defects, a blood

platelet disorder called Paris-Trousseau syndrome that causes abnormal bleeding and easy bruising, difficulty feeding and skeletal issues. Jacobsen syndrome in most cases is not inheritable, but it is caused by random deletions during early reproductive cell development. Those who have the deletion may pass it on to their children.

Cartilage-hair hypoplasia is an autosomal recessive genetically inherited bone growth disorder resulting in dwarfism, impaired immune function and infections due to bacteria and viruses.

There are some conditions TREC testing will not diagnose because the circulating T cell levels are above the cutoff. Examples are major histocompatibility complex (MHC) class II deficiency, a rare autosomal recessive form of combined immunodeficiency with similar symptoms to SCID,² and a rare autosomal recessive form of SCID. Alternative testing strategies need to be developed to identify those with these type of immune deficiencies.

TREC Screening Economics

TREC screening has a high rate of accuracy, which results in significant cost savings. According to Dr. Watson, the TREC assay's "clinical accuracy varies a bit since there are many forms of SCID. Screening for the most severe forms of T cell lymphopenia is very accurate, while some of those with partial or variant forms of the disease can be harder since they may have partial function of the process."

However, Dr. Mei Baker, co-director of the Newborn Screening Laboratory at the Wisconsin State Laboratory of Hygiene, reports that the state of Wisconsin has a 99.98 percent specificity in about 400,000 screenings since the adoption of TREC screening in 2008. This is not only a great percentage but a great percentage at a very low cost. For example, when adopted in Wisconsin, the TREC test cost about \$6.50, in part because the state already had infrastructure in place. The cost of the screenings overall depend largely on a state's birth rate. States with lower birth rates will have lower costs because they need fewer machines, smaller labs and fewer staff. Modell suggests costs can even be as low as \$4.25 per test. However, adds Dr. Baker, any cost of newborn screening is far less than the costs of ruling out a condition later, both financially and emotionally. "This is a positive predictive screening," she says.

According to Hovermale, in approximately one million babies in two years screened in California (the largest number of screened cases in any state), one in 19,900 babies had a significant T cell lymphopenia. For all forms of SCID (typical SCID, leaky SCID and variant SCID), there was a combined incidence of one in 49,700. This compares

with earlier estimates of SCID incidence at one in 100,000 and no real known estimate of the incidence of all T cell lymphopenias.

“We ran a full economic analysis and decision tree,” adds Modell, “of what the costs of testing are versus not testing. When we don’t screen, and just treat as we go, it costs about \$2 million to let the baby go through intensive care treatments and try to keep them alive. If they don’t receive a bone marrow transplant within six months, their bodies may reject the transplant due to overwhelming infections. But if we screen and identify early, we can manage it, treat it, and the baby will live a normal life. That treatment will cost about 25 percent of the costs of a SCID baby not screened.”

Hovermale also provides an economic analysis for the state of Washington: “Recently, Washington state developed a cost-benefit study in conjuncture with their argument to add SCID, which found the benefit–cost ratio of screening for SCID was 4.36, meaning that for every dollar of costs to provide SCID screening, there will be \$4.36 worth of benefits.”

Grass-Roots Campaigns

In 2010, then-Health and Human Services Secretary Kathleen Sebelius added SCID to the list of 29 disorders recommended for population-based universal newborn screening. This addition was the first to be recommended by the Advisory Committee on Heritable Disorders in Newborns and Children since its founding in 2003. However, because this is a recommendation, it is up to each state to decide whether to add TREC screening for SCID to their newborn screening program.

Wisconsin was the first state to add TREC screening for SCID in 2008, followed by Massachusetts, California and New York. Many other states have also added TREC screening, including Colorado, Connecticut, Delaware, Florida, Iowa, Michigan, Minnesota, Mississippi, Ohio, Pennsylvania, Texas, Utah, Washington, Wyoming and the territories of the Navajo Nation. States that are set to add the testing to their panel include Missouri, Nebraska, North Dakota, Rhode Island, South Dakota and West Virginia. Still others have approved the testing but have a longer timetable for implementation. These include the

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District of Columbia, Georgia, Illinois, Maryland, Maine, North Carolina, New Jersey, Oklahoma and Virginia. JMF was instrumental in getting state-level screening off the ground by committing funds to help states get started.

According to Dr. Baker, all states are interested in adding SCID to their newborn screening programs. "Implementing it is the right thing to do," she says. Why some states have already adopted and others have not depends largely on time and funding. "They've got to get the budget and figure out where the money is coming from," she explains. "It's a new area for many, and we have a lot of states in different stages of adoption. We plan to see a lot of new states adding the screening in 2014 and 2015."

In 2010, then-Health and Human Services Secretary Kathleen Sebelius added SCID to the list of 29 disorders recommended for population-based universal newborn screening.

"There may also be issues of available equipment and space," explains Dr. Watson. "The TRECs assay is among the first molecular tests for many of the labs, so training to use it may be needed. This isn't as big an impediment for TREC since it isn't a gene but is, rather, a product of a normal process that results in small pieces of DNA that get clipped out of sequences that create immune responses to different types of exposures. Some labs may even have space issues for adding more tests to their menus."

Unfortunately, at present, the Affordable Care Act (ACA) does not offer much assistance to parents with regard to covering testing. "The ACA is quite thin with regard to genetics. While it does include prevention, which is what newborn screening is all about, there is little specificity as to what it applies to," says Dr. Watson. What the ACA does cover are essential health benefits. These "mostly address the diagnosis and treatment of the condi-

tion rather than the screening, which is a state function that is paid for by state funds, Title 5 funds from the feds, or through fees that are passed on to those being screened," adds Dr. Watson. "We are working to have the diagnosis and treatment of conditions identified in newborn screening added to the essential health benefits since, without them, some kids could fall through the cracks of coverage."

Grass-roots efforts are underway to not only keep the pressure on states to add SCID screening but also to provide educational and advocacy materials to families. "The newborn screening system is not just a lab test," says Dr. Baker. "Education plays a huge role from primary care physicians to family members." IDF says now that screening is on the table, the real work of education across the board can begin. "Since the addition of SCID to the Recommended Uniform Screening Panel, we have had about 50 grass-roots volunteers working in 30 states to get SCID added to their state newborn screening panel," says Hovermale. "One of the most impactful things that these advocates do is to participate in the newborn screening advisory committee meetings that states have to recommend which conditions get added to the state newborn screening panels. When these advocates, who are usually parents of patients (some of whom have passed away), give their testimony and share their stories, they put a face on this disease. It is hard to tell a parent who has lost a child that this test is not worth doing."

"The newborn screening program as a whole is one of the best examples of a public health program," adds Jan Klawitter, public affairs manager at the Wisconsin State Laboratory of Hygiene. "From labs to follow-ups, to genetic counselors to nutritionists, this is prevention at its most basic. Patients who are identified can start on a treatment, and they can hopefully have a much better outcome."

"This is just a great story that was thought to be impossible only a few years ago!" adds Modell. ■

AMY SCANLIN, MS, is a freelance writer specializing in medical and fitness writing.

References

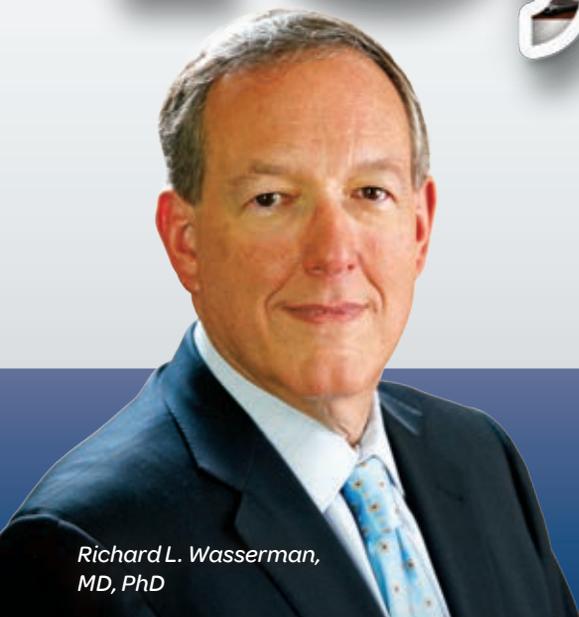
1. Immune Deficiency Foundation. *IDF Patient and Family Handbook*, Chapter 26: Newborn Screening. Accessed at primaryimmune.org/wp-content/uploads/2013/06/Chapter-26-Newborn-Screening.pdf.
2. American Academy of Asthma Allergy & Immunology. SCID Newborn Screening — Not Quite Perfect Yet, Feb. 28, 2013. Accessed at www.aaaai.org/global/latest-research-summaries/Current-JACI-Research/SCID-newborn-screening-not-quite-perfect-yet.aspx.



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*Richard L. Wasserman,
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Important Safety Information

Hizentra treats various forms of primary immunodeficiency (PI) in patients age 2 and over.

WARNING: Thrombosis (blood clotting) can occur with immune globulin products, including Hizentra. Risk factors can include: advanced age, prolonged immobilization, a history of blood clotting or hyperviscosity (blood thickness), use of estrogens, installed vascular catheters, and cardiovascular risk factors.

If you are at high risk of thrombosis, your doctor will prescribe Hizentra at the minimum dose and infusion rate practicable and will monitor you for signs of thrombosis and hyperviscosity. Always drink sufficient fluids before administration.

Please see additional Important Safety Information on reverse side and brief summary of full prescribing information for Hizentra, including boxed warning, on adjacent page.

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*Based on an equivalent dose in grams.



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Important Safety Information (continued)

Tell your doctor if you have had a serious reaction to other immune globulin medicines or have been told you also have a deficiency of the immunoglobulin called IgA, as you might not be able to take Hizentra. You should not take Hizentra if you know you have hyperprolinemia (too much proline in your blood).

Infuse Hizentra under your skin *only*; do not inject into a blood vessel.

Allergic reactions can occur with Hizentra. If your doctor suspects you are having a bad allergic reaction or are going into shock, treatment will be discontinued. Immediately tell your doctor or go to the emergency room if you have signs of such a reaction, including hives, trouble breathing, wheezing, dizziness, or fainting.

Tell your doctor about any side effects that concern you. Immediately report symptoms that could indicate a blood clot, including pain and/or swelling of an arm or leg, with warmth over affected area; discoloration in arm or leg; unexplained shortness of breath; chest pain or discomfort that worsens with deep breathing; unexplained rapid pulse; and numbness or weakness on one side of the body. Your doctor will also monitor symptoms that could indicate hemolysis (destruction of red blood cells), and other potentially serious reactions that have been seen with Ig treatment, including aseptic meningitis syndrome (brain swelling); kidney problems; and transfusion-related acute lung injury.

Reference: 1. Data on File. Available from CSL Behring as DOF HIZ-003

The most common drug-related adverse reactions in the clinical trial for Hizentra were swelling, pain, redness, heat or itching at the site of injection; headache; back pain; diarrhea; tiredness; cough; rash; itching; nausea and vomiting.

Hizentra is made from components of human blood. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent, cannot be completely eliminated.

Before being treated with Hizentra, inform your doctor if you are pregnant, nursing or plan to become pregnant. Vaccines (such as measles, mumps and rubella) might not work well if you are using Hizentra. Before receiving any vaccine, tell the healthcare professional you are being treated with Hizentra.

Please see brief summary of full prescribing information for Hizentra, including boxed warning, on adjacent page.

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.

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Initial U.S. Approval: 2010

BRIEF SUMMARY OF PRESCRIBING INFORMATION

These highlights do not include all the information needed to use HIZENTRA safely and effectively. See full prescribing information for HIZENTRA.

WARNING: THROMBOSIS

See full prescribing information for complete boxed warning.

- **Thrombosis may occur with immune globulin products, including Hizentra. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.**
- **For patients at risk of thrombosis, administer Hizentra at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.**

INDICATIONS AND USAGE

Hizentra is an Immune Globulin Subcutaneous (Human) (IGSC), 20% Liquid indicated for the treatment of primary immunodeficiency (PI) in adults and pediatric patients 2 years of age and older.

DOSAGE AND ADMINISTRATION

For subcutaneous infusion only. Do not inject into a blood vessel.

Administer weekly or biweekly (every two weeks).

Dosage

Before switching to Hizentra, obtain the patient's serum IgG trough level to guide subsequent dose adjustments.

Weekly: Start Hizentra 1 week after last IGIV infusion

$$\text{Initial weekly dose} = \frac{\text{Previous IGIV dose (in grams)}}{\text{No. of weeks between IGIV doses}} \times 1.53$$

- Biweekly: Start Hizentra 1 or 2 weeks after the last IGIV infusion or 1 week after the last weekly Hizentra infusion. Administer twice the calculated weekly dose.
- Adjust the dose based on clinical response and serum IgG trough levels (see *Dose Adjustment*).

Administration

- Infusion sites – 1 to 4 injection sites simultaneously, with at least 2 inches between sites.
- Infusion volume – First infusion, up to 15 mL per site. Fifth infusion, up to 20 mL per site, then to 25 mL per site as tolerated.
- Infusion rate – Up to 15 mL per hr per site. Increase to 25 mL per hr per site as tolerated.

DOSAGE FORMS AND STRENGTHS

0.2 g per mL (20%) protein solution for subcutaneous injection

CONTRAINDICATIONS

- Anaphylactic or severe systemic reaction to human immune globulin or components of Hizentra, such as polysorbate 80
- Hyperprolinemia (type I or II) (Hizentra contains the stabilizer L-proline)
- IgA-deficient patients with antibodies against IgA and a history of hypersensitivity

WARNINGS AND PRECAUTIONS

- IgA-deficient patients with anti-IgA antibodies are at greater risk of severe hypersensitivity and anaphylactic reactions.
- Thrombosis may occur following treatment with immune globulin products, including Hizentra.
- Aseptic meningitis syndrome has been reported with IGIV or IGSC treatment.
- Monitor renal function, including blood urea nitrogen, serum creatinine, and urine output in patients at risk of acute renal failure.
- Monitor for clinical signs and symptoms of hemolysis.
- Monitor for pulmonary adverse reactions (transfusion-related acute lung injury [TRALI]).
- May carry a risk of transmitting infectious agents, e.g., viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.

ADVERSE REACTIONS

The most common adverse reactions observed in ≥5% of study subjects were local reactions (i.e., swelling, redness, heat, pain, and itching at the injection site), headache, diarrhea, fatigue, back pain, nausea, pain in extremity, cough, rash, pruritus, vomiting, abdominal pain (upper), migraine, and pain.

To report SUSPECTED ADVERSE REACTIONS, contact CSL Behring Pharmacovigilance at 1-866-915-6958 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

DRUG INTERACTIONS

The passive transfer of antibodies may interfere with the response to live virus vaccines, and lead to misinterpretation of the results of serological testing.

USE IN SPECIFIC POPULATIONS

- Pregnancy: No human or animal data. Use only if clearly needed.
- Pediatric: No specific dose requirements are necessary to achieve the desired serum IgG levels.

Based on September 2013 version

Let's Talk!

By Trudie Mitschang

Six months after lying in a hospital bed with severe pneumonia and being diagnosed with common variable immune deficiency (CVID), Brandon Dillon completed his first full marathon, coming in under his goal time. This active 41-year-old is determined to live a full, active life despite his diagnosis, and is passionate about inspiring others to pursue their own personal best.

Trudie: You were diagnosed with CVID at the age of 37. Tell us about your life and/or symptoms prior to diagnosis.

Brandon: Growing up, I hardly ever got sick. I had what I would consider a pretty healthy childhood. Prior to my diagnosis in October 2010, I battled the usual sinus infections that most of us with a primary immunodeficiency (PI) do, usually four or five per year. It seemed that every time I caught a cold, it would

turn into an infection. This started happening when I was in my mid-20s. Years later, I was in the hospital being treated for a very severe case of pneumonia when I was diagnosed with CVID; this was my third time with pneumonia and by far my worst case.

Trudie: You were an active athlete prior to your diagnosis. Did you ever struggle with keeping your stamina up?

Brandon: I've been an avid cyclist and runner since I was in my 20s. Before I was diagnosed, I never really struggled with having energy to do these things or keeping up my stamina. I did always fear that I would get a cold or a sinus infection that would keep me out of an event that I wanted to participate in. However, I never felt my abilities were limited due to having any illness.

Trudie: What is your current treatment plan, and how are you responding to it?

Brandon: I am currently being treated with intravenous immune globulin every four weeks. I have my



After a relatively healthy childhood, Brandon Dillon began getting multiple colds, including three cases of pneumonia, before he was diagnosed with CVID at age 37.

My hope is that somebody reading this who has CVID or another PI, who thinks that life as they once knew it before diagnosis is over, can clearly see that it doesn't have to be.

turn into an infection. This started happening when I was in my mid-20s. Years later, I was in the hospital being treated for a very severe case of pneu-

monia when I was diagnosed with CVID; this was my third time with pneumonia and by far my worst case. treatments done in an infusion clinic, and they usually take about three hours to complete. I've been fortunate that my body responds very well

to the treatments, usually with little or no side effects.

Trudie: You decided to continue to pursue your fitness goals, even participating in your first full marathon, despite CVID. Tell us about that experience.

Brandon: After my diagnosis, I was at a follow-up visit with my immunologist, and he told me that the goal with my treatment is to die with CVID and not from CVID, and that there was no reason that I could



After his diagnosis, Brandon was determined to continue his active lifestyle, including finishing his first marathon and competing in triathlons. He hopes he can be an example that life doesn't have to be over after being diagnosed with CVID.

not continue to participate in the activities I love. I took that literally, so when a friend asked me to run a full marathon with him that spring, I said yes. Prior to that, I had run a half marathon and a number of shorter-distance races. I decided that since my body was responding well to the treatments that I was going to go for it. Six months to the day of lying in a hospital bed with pneumonia and being diagnosed with CVID, I completed my first full marathon (26.2 miles) in three hours 42 minutes, under my goal time of four hours. Physically, this is one of the toughest things I have ever done and perhaps one of the greatest accomplishments of my life. When I told my doctor about this, he was ecstatic and said

that was the best thing he had heard from a patient in a long time.

Trudie: Last year, you made the brave decision to start competing in triathlons. Where do you get your strength and motivation?

Brandon: Prior to my diagnosis, I had competed in a couple of short-distance triathlons. In time, I felt the motivation to pick them up again. Last year alone, I competed in three sprint- and one Olympic-distance race, two half marathons, one team relay covering 200 miles, and the Bike MS fundraiser riding 150 miles in two days.

Trudie: What are your goals for the rest of this year?

Brandon: This year, I've decided to challenge myself even more and compete in a half-distance triathlon (1.2 mile open-water swim, followed by a 56-mile bike ride, finishing off with a half marathon — 70.3 miles in all). That race is in September, and I have plenty of races lined up beforehand to prepare for that. Having CVID doesn't discourage me from doing these things; if anything, it gives me more motivation.

Trudie: You mentioned a desire to motivate others living with chronic illness. What advice do you have for

those who feel limited because of their diagnosis?

Brandon: My hope is that somebody reading this who has CVID or another PI, who thinks that life as they once knew it before diagnosis is over, can clearly see that it doesn't have to be. I choose to be active and do these things to help me stay healthy. Running marathons and competing in triathlons isn't for everybody, but if your goal is to be more active, then you should find an activity that you like to do and get out there and do it. Even if it's just a short walk around the block, you've got to start somewhere. If your goal is to participate in a 5K fun run, find one and register! Sometimes just signing up is all the motivation you need. Having an immune deficiency isn't the end of the world, and it shouldn't be used as a reason to hold you back.



TRUDIE MITSCHANG is a staff writer for IG Living magazine.

If your life depends on immune globulin and you have a unique experience to share, we want to feature you in this column! Email us at editor@IGLiving.com.

A Garden of Humanity

By Stacy Oliver

AFTER SIX YEARS of getting immune globulin (IG) infusions, you'd think I'd have a more casual attitude about seeing the bottles of IG that arrive every month. It should be old hat by now; I shouldn't even blink an eyelash anymore. And, yet, the arrival of the magic liquid still utterly fascinates me. The concept that I receive healing from a product that is made from thousands of donor contributions still blows my mind. When infusion time rolls around, I'm like a flower that's wilting. I need that special nutrient to perk my petals up. As far as I know, IG has yet to be recreated as a synthesized product. So, it's as organic as it gets. I like to think of it as a garden of humanity.

Have you ever really looked at a bottle of IG? It says that it's a human product. Right next to the words "Immune Globulin" is the word "HUMAN." I guess I should feel kind of like a vampire sucking on the life force of others to survive, but it's not that grim of a picture in my mind. Picturing the thousands of people giving their blood to extract the miracle IG makes me feel closer to humanity. Their life gives me life, and maybe in some mystical way, I also get their good wishes, positive feelings, strength and courage to face my autoimmune challenges.

I don't have fairy dust in my eyes. I know that IG donors get paid for their service. My husband checked to see if blood banks are located in our area, and there were none close by. He and I wanted to visit one of the donation centers and also ask our

friends to give blood to ensure the supply doesn't deplete. I know that sometimes there are IG shortages. It hasn't affected me yet, but what a terrifying thought. In fact, the blood centers we found were far away, and most were near college campuses. I'll never know the true intentions of the people who take the time and give blood to make IG for me. Maybe some people do it just to get the money, or maybe they know how lifesaving their donation is.

I'm sure there are people in labs working on creating an artificial substance to mimic what the natural form of IG can do. It would ensure there are no shortages. Perhaps in the future, there will be better ways to administer IG besides infusions. Instead of getting two nine-hour doses each month, I'm hoping for administration innovations. Wouldn't it be great if there were IG lotion to slather on and get absorbed into your

skin? Or IG nail polish? I'd like to think there is a scientist with a Willy Wonka spirit at heart who would invent IG bubble gum one day. Chew it all day, and the IG is released into your body via your tongue. Hey, a girl can dream. If our society can put a man on the moon, anything should be possible.

Until that day comes, I am grateful to my fellow human beings for helping me, quite frankly, live. We are all in this garden of life together — a wondrous variety — and we can all help each other bloom and grow. ■



STACY OLIVER was diagnosed in 2008 with multifocal motor neuropathy (MMN). She is the assistant director of the Center for the Writing Arts at Northwestern University, and she is working on her supersecret identity as *Neuropathy Girl*, who will one day save the world after her infusion and a nap.



Life as a 20-Something



Financing Your Illness: Five Financial Mistakes I've Made

By Ilana Jacqueline

"IT'S HARD OUT there for a PI patient," I told my fiancé as we flipped through a giant binder I'd compiled in preparation for filing my taxes. "I'm just not making as much as I thought I was."

"Or you're spending more than you think you can," he replied.

"Well, I'm not out buying Louis Vuittons!" I countered.

In fact, for the record, I have exactly zero red-soled shoes. My monthly credit card statements are saturated with copays, medications and hospital bills. For those keeping score, here's what a monthly breakdown of my medical bills look like (and this doesn't include the insurance premiums that my parents pay):

- An average of two doctor appointments a week at \$25 each equals about \$200 a month.
- The 10 base medications I take every month equal about \$15 each for a total of \$150 per month.
- Financed hospital bills also equal about \$50 a month.

So to start out my monthly budget, I immediately take out a cool \$400.

Then, I'm also human, so I need things like shelter (rent), food, Internet, gas and car insurance.

The older I get, the more and more of my financial medical responsibilities I have to take on — and I'm determined, just as I am with all the other parts of my life, to bridge the gap from sick to normal. So to start off my financial responsibility party, I'm going to count down the top-five money mistakes I made in handling my disease:

1. I didn't factor my disease into my financial equation. As a high school student, I hardly acknowledged the realities of my disease. So, the realities of how they financially impacted my family didn't quite register with me. What does it even mean to factor in the financial realities of your disease? Start by getting a scrap of paper, and as you go through your day, write down what your disease is costing you. Did you take 10 pills this morning? Did you drive half an hour to a regular doctor's appointment and pay a copay? Did you run out of energy to cook the food you already have in your fridge and instead decide to just pick up a pre-made dinner again? Cha-ching.

2. I didn't go generic. Did you know that you can always ask your doctor to write you a prescription for the generic version of your medication? The only difference is the advertising. Generic drugs are safe, FDA-approved and, as a general rule, cost much less than the brand-name drugs.

3. I didn't always do enough research on doctors. The other day, I was looking up a specific specialist for a biopsy I needed done. I asked his secretary if this doctor did that kind of work, and she said yes. I got to the office, and guess what? He didn't do that kind of biopsy. Copay wasted. When it comes to finding the right doctor, ask twice. Three times if you need to — just to make sure they're the right one for you. Check online reviews; search their career history. Ask other doctors who would be a good fit.

4. I didn't search out assistance programs. Did you know that many drug manufacturers have assistance programs to help patients afford their medications? By signing up with their program, you could qualify to save hundreds on your treatment. Ask your doctor, or Google your prescription to see if your medication has a patient assistance program.

5. I was in the mindset of allowance, because I was in need. The reality is that any way you slice it, my needs are greater than the average 24-year-old's. I'm not buying new shoes every week. I'm buying the medication I need to live, the doctors I need to manage my treatment, and handling the medical debt that's layered from years of chronic illness. But just because you *have* to spend your money, doesn't mean you can't put yourself on a budget. You have to cut costs somewhere, so live with financial awareness. Ask for help when you need it, and allocate money to your savings when you have it.

Take it one step at a time. Just being aware of where your money is going can be the perfect start to helping you make better decisions for your medical and financial future. ■

ILANA JACQUELINE is a 24-year-old dysautonomia and primary immune deficiency disease patient from South Florida. She's been writing professionally since 2004 on everything from health and wellness to celebrities and beauty. Her blog www.letsfeelbetter.com is both a personal collection of anecdotes about life with chronic illness, as well as a resource for patients of all ages.

Sub-Q Made Me Do It

By Cheryl L. Haggard



I SUFFER FROM an autoimmune disease that is playing my favorite childhood board game, Operation, with my spine and sacroiliac joints. When my immune system doesn't hold steady, it mistakenly buzzes the delicate sides of the game pieces (in real life and not the game), and my joints go "buzz, buzz" (read: agony, suffering and inconceivable pain).

On the bright side, my chronic illness has given my husband, Mark, and me a rare and unique opportunity to teach our primary immunodeficiency (PI) teens — like in the board game Operation — to hold steady with a fundamental moral code not to give into peer pressure. Every choice we

make has a consequence, and it's up to us whether the consequence is positive or negative. We have also taught our PI teens to find purpose in their pain and live victorious lives. However, little did I know that at the kids' next subcutaneous immune globulin ("sub-q") infusion I was going to revisit puberty and have my PI teen so-called theories put to the test!

It was a particularly barometrically challenging day for me and my spine. You see, I can rely on my joints to accurately predict storms: two jolts of pain in my L2 equals some lightning activity and rain; arthritis-type torture in my knees accompanied with shooting pain originating from my

left hip equals moderate rain, wind and lightning strikes; and finally, "don't speak to me" and "she's gonna blow!" discomfort surging through my right sacroiliac equals hail the size of badminton birdies and gale-force winds that pull your facial skin so taut you look like an alien. It was also the day my two PI teens, Caleb and Molly, had to have their sub-q infusions.

"Hey guys, get your numbing cream going, it's sub-q day," I hollered from the kitchen sink. It felt good to wash my hands, as the warm water somehow soothed the aches of arthritis, not to mention I needed to get my digits moving because it was my turn on the put-sub-q-infusion-stuff-together rotation. "C'mon, guys! I've gotta go get dinner started!" I begged.

Two minutes later, two PI teens finally got their "spots" numbed up, readying them for the tiny needles that will facilitate their infusions. We've done this routine so many times, we could probably put the delicate sets of tubing and pump together in our sleep, but this time it was different for me. It wasn't so simple to take the Mini-Spike, insert into vial of immune globulin (IG) and push the precious liquid through the tubing. I was having a storm-three day, and I didn't know if I was going to be able to have the strength to prep their meds. Something bigger than myself was holding me back, tempting me.

I recently read a study proving the positive effect IG is having on patients suffering with aggressive autoimmune disease. Some patients

experienced complete relief from their symptoms and were clinically showing damage reversal. The article had me imagining complete relief of my symptoms, and that was good enough for me.

On our kitchen table lay an ocean blue chuck pad (protective/sterile lining) dotted with Caleb's and Molly's sub-q supplies: Mini-Spikes, tubing, two Freedom pumps, just to name a few. The off-putting stench of alcohol wipes normally offended my nares, but for some odd reason, the pungent disinfectant intrigued my senses. So much so, I wiped the bottle of IG a smidge closer to my left nostril so it wasn't so obvious.

"Mom, what are you doing?" Molly interrupted, disgusted by my overt public display of affection.

I was getting in deep by whatever it was pulling me into this deep abyss; I had to find self-control and some level of human decency. "I'm sorry, Molly," I half-heartedly begged. "I don't know what's gotten into me, Sweets," I cooed.

"I know your hip has been bugging you lately, but we all agreed that we'd rather wipe our infusion sites with lamb and rice dog food than swab with alcohol," Molly gently reminded me. "Does your hip hurt so bad, you're making poor decisions?" my teenager inquired with a tone similar to my own. "Who's the parent now?" Molly muttered and bullied my bi-weekly job right out from under me. But, I didn't care. As she drew up the crystal clear medicinal elixir from the tiny vial, its healing power weakened every last drop of maturity and good sense I had left. I wanted it bad, real bad.

"I'm ready to go, Mom," Molly

announced, temporarily saving me from my IG trance.

As I wiped every last bit of Emla cream off Molly's belly, I did all I could to shoo away thoughts I never had before. You aren't capable of stealing your kids' meds, are you? I asked myself. Do you think you won't get caught? What happens if you have a serious reaction, how are you gonna explain yourself?

None of it mattered. I was determined as ever to get a chance at freedom. Molly and I recited our infusion prayer, I pinched the butterfly needle and headed toward the "bullseye" on Molly's belly. I squinted, and then, suddenly, my whole body jerked back and forth, writhing and wrenching as if I was playing tug-of-war with myself.

"Uh, Mom? You OK there?" Caleb asked, placing his comforting hand on my shoulder. "You're moving around like you've got snakes in yer skivvies."

"And, she's creeping me out!" Molly cried.

"I can't do this! I just ... can't ... do ... this," I admitted, dropping my arms and shoulders causing my head to flip flop around like fresh-caught mackerel.

After a period of calming myself down and getting back in touch with sane Cheryl, I confessed to the children how much I wanted to try IG. Molly, of course, asked why I couldn't try just one infusion to see if it would help my pain.

"Because it's very illegal and very dangerous," Caleb jumped in to save me from having to utter one (dramatic pause) more (dramatic pause) word (bow, and she's done).

"Well, the side effects aren't too

bad," Molly chimed in. After we read through the laundry list of side effects — migraine headache, blah, blah, vomiting and dehydration, blah, blah, death, blah — I was grateful. I pulled myself together, cleaned up my conscience and wasn't on the floor having a seizure. Lesson learned.

"But ya' know, Mom, I still think you should try an infusion anyway," Caleb said. "I mean, with your crazy immune system, who knows what might happen! You just might grow another limb!"

"Oh, that'd be so cool!" Molly agreed.

I inspected my right foot and thought, I do have a healthy appetite for Jimmy Choo's new line of strappy sandals. But, then I thought better of it. Why would I want to take the risk of borrowing Caleb's and Molly's IG, not to mention having to explain why I needed to drop 1K for the designer shoes? The potential side effects: light-headedness, vomiting, diarrhea, jaundice, blurting out obscenities at bridge club, and that's just for my new Choo's! I weighed the consequences and decided to put the needles into the right patients: Caleb and Molly.

Making the right decision made me feel so good I decided to make another good decision and take a nice long walk on my two legs ... right through the shoe department at the mall. ■



CHERYL L. HAGGARD is a stay-at-home mom and has three children who have common variable immune deficiency.

Parenting

Keeping Connected: Cell Phones and Kids

There are pros and cons to giving kids cell phones, but they can be a useful tool if children are mature enough and are taught to use them responsibly.

By Mark T. Haggard

REMEMBER “THE BRICK”? The cellular phone that was as big as a head? It was a status symbol, so expensive that only a few well-to-do people could use it. Today, cell phones have become a necessity for the family. It’s the way families stay connected, and the number of children who have cell phones is rising. According to a Kaiser Family Foundation survey, 85 percent of adolescents ages 14 to 17 have cell phones. Furthermore, 69 percent of 11- to 14-year-olds have their own cell phones. Most striking is the fact that 31 percent of children ages 8 to 10 have their own cell phones. How young is too young for a cell phone? And, is that answer different for families with children with immune deficiencies?

As with all modern-day conveniences, there are pros and cons. The pros concerning giving cell phones to kids generally revolve around parents having quick access to their kids. Cell phones are a sort of cyber-umbilical cord; it’s good to know that wherever they are, children are only a phone call or a text away.

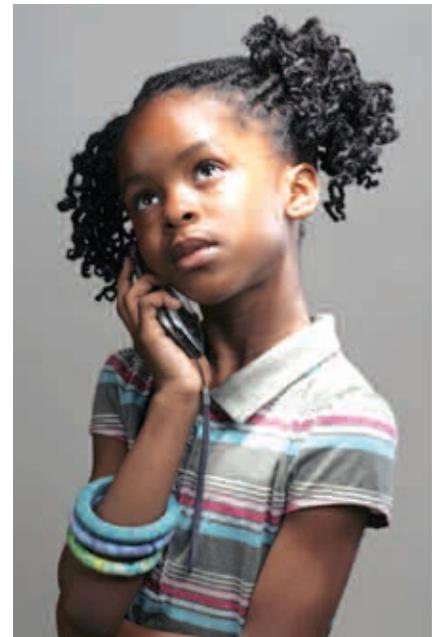
For kids with immune deficiencies, cell phones offer many benefits. For one, a well-timed call can be a reminder that it’s time for a treatment such as subcutaneous immune globulin. For another, it keeps these kids from handling a germ-plagued public

phone. Even so, access to a cell phone should be in direct proportion to a child’s maturity.

“Addiction” and Other Negative Aspects of Cell Phones

There are several potential negatives to children having cell phones. Beyond just sending and receiving phone calls, cell phones now put social media, games, movies, music and videos in the palms of their hands. Do we want children to have access to everything that the Internet has to offer? Being online also exposes kids to the problems of cyberbullying, harassment via texting, instant messaging or other forms of social media. Beyond being a distraction, cell phones can produce seemingly “addictive” behaviors. In my classroom, I have watched students go through what look like withdrawals when I take their phones away; some would rather fail a class than lose their phone. Cell phones with cameras have their own set of problems like using cameras for cheating in school, sexting or using cameras to take pictures of people without their consent (the Massachusetts legislature recently passed laws against “upskirting” and “downblousing”).

A huge concern is the combination of cell phones and driving. A Virginia Tech Transportation Institute study



revealed that using a cell phone is the most distracting task in which a driver can engage. Statistics kept by the National Safety Council indicate that 28 percent of all traffic accidents are caused by drivers using a phone to call or text. More than half of teens who own cell phones say they have talked on the phone while driving, and a third admit to texting while driving.

Cell phones may also lead to physical ailments in children. Early studies suggest that constant texting and emailing can disrupt kids’ concentration and may lead to compulsive behavior since children believe they

must be available 24/7. A recent survey found that four out of five teens who own cell phones sleep with them next to their beds, keeping them close so they can receive texts in the middle of the night. The result is a lack-of-sleep pattern that becomes the norm causing considerable problems with concentration, and in the long term, may lead to obesity, heart disease, diabetes and depression.¹

When to Buy

There is no optimal age for getting a child a cell phone. If a child is responsible enough to complete his or her homework, obey curfews and do household chores, then he or she is likely responsible enough for a cell phone, regardless of age. "Maturity and the ability to be responsible are more important than a child's numerical age," says Caroline Knorr, parenting editor with Common Sense Media. On the other hand, if a child is still working on some of these areas, it may be wise to hold off giving them a phone. Lori Evans, director of training in psychology at the New York University Child Study Center, advises parents to "look for developmental signs. Does your child lose his belongings? Is he generally a responsible kid? Can you trust him? Will he understand how to use the phone safely? The rate at which kids mature varies — it will even be different among siblings."

Parents should also consider whether a child needs a phone or simply wants one. A kid only needs a phone if he or she is travelling alone from place to place. A child who rides the bus or travels in a carpool might not necessarily need a phone. One who walks to school or spends time on his or her

own after school probably does need one. "It's about who they are as individuals — what's going on in their lives, and how much they can handle, not a certain age or grade," Evans says. "The issue is really about educating children how to use cell phones in appropriate ways. Cell phones can definitely be beneficial, as long as you know your individual child."

What to Buy

Once it's determined that a child is mature enough for a cell phone, what kind needs to be considered. "Most parents want to give [their kids] a cell phone to keep them safe," says Dr. Ruth Peters, a child psychologist in Clearwater, Fla. "But that ignores the great majority of uses that kids are using cell phones for." Children very well could get caught up in the negative behaviors noted above. Peters recommends that parents not buy children younger than 13 a phone with a camera and Internet access. "If they don't have access to it, it's just cleaner," she says.

For a long time, my 13-year-old daughter wanted a phone. We broke down this past Christmas and got her one, without Internet access or a camera. It is a prepaid phone for which we buy the hours upfront, and when the minutes run out, she's done. The onus is on her to be responsible for the minutes she uses. For now, she only uses her phone to get hold of her parents or to make very short phone calls to her friends. The more money she is willing to commit, the more she will be able to talk.

Establish Rules

Once a phone is purchased, the rules need to be explained. For

instance, will there be restrictions on texting, how many minutes they are allowed and when and where they can text? (I've noticed teens never talk on their phones.) Also, will there be places where the phone is off limits (such as during dinner)?

Most importantly, kids should not be allowed to play the "invasion of privacy" card. When parents pay for the phone and the plan, they have the right to know who their kids are talking to and for how long. "Kids consider mobile devices to be personal property, and they don't want their parents snooping around," says Knorr from Common Sense Media. "But I think parents are justified in saying, 'I understand this can be used for good, but it also can be misused. So every now and then, I'm going to check to make sure you're using it responsibly and respectfully.' Then, make it an ongoing dialogue: 'Have you gotten weird texts?' 'Any calls that made you uncomfortable?' 'Who are you texting?'"

Like many other technologies in the modern world, cell phones can be the proverbial double-edged sword for children. There are numerous traps that they can fall into if they become tied too closely to their phones. But with good parenting, a cell phone can be an excellent tool in helping kids and parents stay connected in a fast-paced world. ■

MARK T. HAGGARD is a high school teacher and football coach, and has three children who have common variable immune deficiency.

Reference

1. Davis, S. Is Your Child Ready for a Cell Phone? WebMD. Accessed at www.webmd.com/children/features/children-and-cell-phones.

IG Chronicles

Ready for Back to School? I Hope So...

By Dona Darr



Dona Darr is doing all she can to keep her daughter, Emily, who was diagnosed with IgG subclass deficiency and complement deficiency in 2004, healthy.

I CAN'T HELP but feel mixed emotions with the start of the new school year inching ever closer. I am excited to watch Emily get ready for her first day of middle school, yet saddened at the same time that my baby is old enough to be in middle school. I am excited to see her rejoin her friends, some of whom she has been unable to see during the summer break, yet worried at the same time of what that rejoining will bring.

We had a wonderful summer break with sleepovers and a vacation to Kentucky Lake. I'm not ready for it to end. She has stayed relatively well all summer. No major illnesses to deal with other than the occasional asthma attack and one sinus infection. We even got a definitive diagnosis from her immunologist and started a

treatment plan. I'm not ready to send her back to the jungle of germs, but I must.

So with school supply list in hand, off for a day of shopping we go. One by one, the items are checked off the list. Now, we switch to another list, Mom's back-to-school list: medications refilled, hand sanitizer and sanitizing wipes on hand. She is ready for fifth grade, but mom isn't ready.

The last couple of weeks of summer break will be filled with meetings with the teachers and school administrators. We now have documentation from a doctor explaining her disease and a mound of educational supplies provided by the Immune Deficiency Foundation to supply each of the teachers who will have her in their class. I just hope that they actually read it. Armed with my tote bag filled with information, I will meet with her teachers, her principal, her guidance counselor and the school nurse. The nurse and I have come to know each other quite well over the last couple of years. We will develop a 504 Plan and have everything in place to start the new school year. I have even prepared and stocked her school bus with hand sanitizer and sanitizing wipes; it helps that I am the bus driver. I have done all I can do to

prepare Emily and the school, but I'm not prepared. I'm not ready.

Last year, within the first week of school, Emily came down with strep. By the end of September, she had already missed eight days of school, some of which was due to the fact that she ended up having to have surgery to have her tonsils removed. Will this year be a repeat of last? I hope not. She is taking the prophylactic antibiotic to help ward off infections. Will it work? I hope so. Will the school staff actually do everything they said they would do to help keep her healthy? I hope so. Will the year go by with little illness and fewer days missed? I hope so. Have we thought of everything possible that we can think of? I hope so.

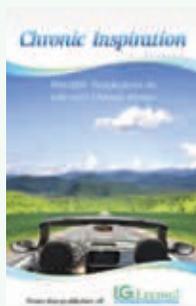
Only time will tell if we have prepared properly. All I can do is hope so. ■

DONA DARR is the mother of Emily who was diagnosed with IgG subclass deficiency and complement deficiency. Dona and Emily have been dealing together with this disease since 2004, when Emily was initially diagnosed. Dona and her support system of family and friends will continue to care for and encourage Emily for the rest her life.

Reprinted with permission from Dona Darr's blog titled Our Journey with Primary Immunodeficiency Disease at donadarr.blogspot.com.

Patients who rely on IG therapy have unique life experiences. If you have a story you'd like to share about your adventures, experiences, relationships, reminiscences, self-portrayals, etc., for publication in this column, submit it to editor@igliving.com. All submissions must be 600 words or fewer and can be accompanied by high-resolution photos.

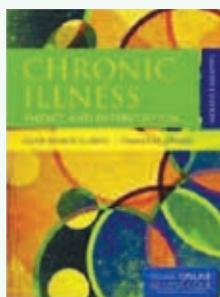
Book Corner



Chronic Inspiration

Author: IG Living Blog Contributors
Publisher: FFF Enterprises

Chronic Inspiration is a compilation of blogs from *IG Living* magazine, the only magazine dedicated to patients who use immune globulin products and to their care providers. The eBook features 58 reflections written by patients, parents and caregivers who offer inspiration and heartfelt perspectives from the frontlines of chronic illness. It is an anthology of *IG Living's* most provocative, creative and inspired submissions that can be downloaded for a quick, on-the-go dose of inspiration, serve as a reminder to readers that they are not alone, and be shared with friends and family who for whatever reason still “don’t get it.” The eBook is separated into eight chapters titled From the Heart, Life as We Know It, Parenting Perspectives, Speaking Our Truth, Encouragement for the Journey, A Walk in My Shoes, Live Like You Were Living and You Don’t Look Sick. Early reviews call it a “must read for any chronically ill patient or caregiver looking for inspiration.” It is available for download and purchase on Amazon.com.

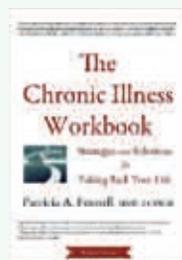


Chronic Illness: Impact and Intervention

Authors: Ilene Morof Lubkin
and Pamala D. Larsen

Publisher: Jones & Bartlett Learning

The newest edition of best-selling *Chronic Illness* continues to focus on the various aspects of chronic illness that influence both patients and their families. Topics include the sociological, psychological, ethical, organizational and financial factors, as well as individual and system outcomes. This book is designed to teach student nurses about the whole client or patient versus the physical status of the client with chronic illness. The study questions at the end of each chapter and the case studies help students apply the information to real life. Evidence-based practice references are included in almost every chapter.

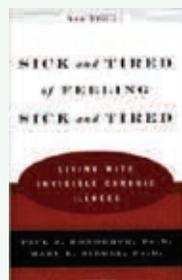


The Chronic Illness Workbook: Strategies and Solutions for Taking Back Your Life

Author: Patricia A Fennell

Publisher: Albany Health Management Publishing

The Chronic Illness Workbook is written to bring clarity and order to what feels like an unmanageable and isolating experience. It shows both those who are ill and those who care for them how to live a full and meaningful life despite undeniable difficulties. Using her extensive experience with chronic illness patients, Patricia Fennell has created an original, comprehensive, research-validated approach that considers not only the physical aspects of chronic illness, but the psychological, social and economic aspects as well.



Sick and Tired of Feeling Sick and Tired: Living with Invisible Chronic Illness

Authors: Paul J. Donoghue
and Mary E. Siegel

Publisher: W. W. Norton & Company

Since its first publication, *Sick and Tired of Feeling Sick and Tired* has offered hope and coping strategies to thousands of people who suffer from invisible chronic illness. Paul Donoghue and Mary Siegel teach readers how to rethink how they themselves view their illness and how to communicate with loved ones and doctors in a way that meets their needs. In this new edition, the authors include a new introduction drawing on the experiences of the many people who have responded to the book and to their lectures and television appearances. They expand the definition of invisible chronic illness to include other ailments such as depression, addiction and obsessive-compulsive disorders. They bring the resource material, including websites, up to the present, and they offer fresh insights on four topics that often emerge: guilt, how invisible chronic illness affects the family, meaningfulness and defining acceptance. ■

Alternative IG Side Effect Treatments

By Carla Schick

PATIENTS WHO INFUSE immune globulin are all too familiar with the side effects. While side effects are not easy to tolerate, most if not all of them can be eased through the use of a few alternative methods.

Body Aches, Chills and Fatigue

Body aches, chills and fatigue are some of the more common side effects of IG therapy. One option that patients can use to manage post-infusion pain is a transcutaneous electrical nerve stimulation (TENS) unit, which is a small, non-invasive device that is accompanied by several electrode pads that attach to the skin. The battery-operated TENS unit delivers gentle currents of electrotherapy to calm agitated nerves and muscles.¹

For those who suffer from body chills, a pair of therapeutic slippers is a way to keep feet warm before, during and after treatment. The slippers can be placed in the microwave for 30 to 90 seconds to provide warmth to sore, cold feet. Another way to ward off chills is to wear clothing that is especially intended for infusion sessions. Full-length zippers that are sewn into sweaters and pants allow nurses to access patients' arms and legs, while still leaving them warm and comfortable. And, a body temperature regulating blanket, which utilizes microcapsules to store, absorb and release heat, can help to prevent chills or overheating.

After IG treatments, extreme fatigue and/or sore infusion sites can make it challenging to find a comfortable sleeping position. Body support pillows are a helpful, non-medicinal solution. Some modern body pillows are available in a special U-shape that cradles the body from head to toe to provide complete and evenly distributed support.

Migraines, Dizziness and Nausea

Also common are headaches, migraines and dizziness post-infusion. In many instances, these adverse reactions can be attributed to dehydration. Registered Nurse Jennifer Richlin recommends patients "start drinking water, juice and power drinks the day before an infusion is scheduled [and] stay away from coffee or alcohol as they can dehydrate the body."² Some patients turn to electrolyte rehydration drinks, which are good to have on hand before, during and after infusions because they provide the body with necessary minerals and salts to function properly.



There are also advantages to consuming ginger and peppermint to treat stomach issues. Dr. Barrie R. Cassileth, chief of the Integrative Medicine Service program at Memorial Sloan-Kettering Cancer Center, touts the benefits of gingerroot tea, ginger ale and ginger cookies made from real ginger to calm a queasy stomach. Additionally, peppermint tea is widely used to relieve stomach problems, aid in digestion and relieve nausea.³

Finally, one of the most debilitating and prevalent side effects is migraine. In addition to being properly hydrated, patients can apply a migraine wrap around their head to help soothe discomfort. The wraps usually feature a removable gel pack that can either be heated or cooled to deliver therapeutic relief.

Relief Is Possible

Although it is not possible to eliminate IG infusion side effects entirely, their intensity can be minimized by incorporating non-medicinal alternative treatments to pre-and post-infusion treatment routines.

CARLA SCHICK is a staff writer for IG Living magazine.

References

1. Mayo Clinic. Immune Globulin (Intramuscular Route, Intravenous Route, Subcutaneous Route). Accessed at www.mayoclinic.org/drugs-supplements/immune-globulin-intramuscular-route-intravenous-route-subcutaneous-route/side-effects/drg-20067720.
2. Richlin, J. IVIG Infusion Guide: Tips to Make Infusion Days Easier. *IG Living*, October 2006. Accessed at www.igliving.com/Assets/IGL/Articles/IGL_2006-10_AR_IVIG-Infusion-Guide-Tips-to-Make-Infusion-Days-Easier.pdf.
3. Cassileth, BR. Complementary and Alternative Cancer Medicine. *Journal of Clinical Oncology*, Nov. 1999. Accessed at jco.ascopubs.org/content/17/suppl_1/44.short#cite-by.

Directory of Alternative IG Side Effect Treatments



Full Size Comfort-U Body Pillow

The Comfort-U Full Body Support Pillow is a U-shaped cushion that cradles the body to evenly support the head, back, knees and ankles. Folded in half, each side of the pillow measures 64 inches long and 10.5 inches wide. The pillow weighs approximately 10 pounds

and is filled with Fusion Fiberfill, a down-like hypoallergenic stuffing. It comes with a 50 percent cotton/50 percent polyester pillowcase.

www.mycomfortu.com/comfort-u.html



DreamTime Pampered Soles Foot Cozys

DreamTime Pampered Soles Foot Cozys offer warm or cold therapeutic relief from aches and pains. The soft fabric

comes in two colors: lavender velvet and sage velvet. The cozys also feature an aromatic blend of cinnamon, clove and eucalyptus to soothe the senses. The foot cozys are available in one size fits most.

relaxanddream.com/dream-time/dreamtime-pampered-soles-foot-cozys?category_id=20



Spa Comforts Welcome Comfort Migraine Wrap

The Spa Comforts Welcome Comfort Migraine Wrap features a removable gel pack that provides localized heat or cold therapy to ease pain and tension. When in place, the adjustable Velcro closure ensures secure relief. The wrap measures 22

inches long by 4 inches wide.

relaxanddream.com/spa-comforts/spa-comforts-welcome-comfort-migraine-wrap?category_id=20



CeraLyte 70-Citrus Ready To Drink

CeraLyte 70 Citrus Ready to Drink is a rice-based electrolyte beverage that is used to prevent and correct dehydration from diarrhea and/or vomiting. It can be served cold, at room temperature or warm. CeraLyte 70 is gluten-free and comes in a

16.9-ounce ready-to-use container.

(888) 237-2598,

www.ceraproductsinc.com/productline/ceralyte.html



Libre Clothing

Libre Clothing is treatment apparel that was specifically designed to provide warmth, comfort and dignity to patients undergoing infusions. The sweatshirt features hidden zippers for left or right arm access and/or left, right or both chest accesses.

The pants are a cotton and polyester blend and offer left or right leg zipper access. Libre infusion wear is available in men's and women's sizes, and is offered in a variety of colors.

(888) 542-7349, www.libreclimbing.com



Temperature Regulating Blanket

The Temperature Regulating Blanket uses patented microcapsule technology developed for NASA to absorb, store and release heat to prevent chills or overheating. The blanket features a chevron weave pattern and

is made from 51 percent soft Merino wool and 49 percent acrylic. It is available in twin, full, queen and king sizes. The blanket is naturally antibacterial, odor-resistant, machine washable and comes in bold blue, silver taupe, Chianti, white, prairie green and natural.

(800) 321-1484, www.hammacher.com/Product/Default.aspx?sku=83742&refsku=84231&xsp=1&promo=xsell

General Resources

These organizations provide information about various disease states, which can be found by conducting a search of the disease state name.

- Advocacy for Patients with Chronic Illness: www.advocacyforpatients.org
- American Autoimmune Related Diseases Association (AARDA): www.aarda.org
- American Chronic Pain Association (ACPA): www.theacpa.org
- Band-Aides and Blackboards: www.lehman.cuny.edu/faculty/jfleitas/bandaides
- eMedicine from WebMD: emedicine.medscape.com
- FamilyDoctor.org: www.familydoctor.org
- Johns Hopkins Medicine: www.hopkinsmedicine.org
- KeepKidsHealthy.com (pediatrician's guide to children health and safety): www.keepkidshealthy.com
- Kids Health (medical and emotional impact of caring for an ill child): www.kidshealth.org/parent/system/ill/seriously_ill.html
- Mayo Clinic: www.mayoclinic.com
- National Committee for Quality Assurance (detailed report cards on health plans, clinical performance, member satisfaction and access to care): www.ncqa.org
- National Heart, Lung and Blood Institute: www.nhlbi.nih.gov/health/health-topics/by-alpha
- National Institute of Neurological Disorders and Stroke (NINDS): www.ninds.nih.gov/disorders/disorder_index.htm
- National Institutes of Health: health.nih.gov/see-all-topics.aspx
- National Organization for Rare Disorders (disease-specific support groups and virtual communities for patients and caregivers): www.rarediseases.org
- Office of Rare Diseases Research: rarediseases.info.nih.gov
- Patient Advocate Foundation (patient access to care, maintenance of employment and financial stability): www.patientadvocate.org



The nonprofit Patient Services Incorporated, www.patient-servicesinc.org, specializes in health insurance premium, pharmacy co-payment and co-payment waiver assistance for people with chronic illnesses. (800) 366-7741

- WebMD (medical reference): www.webmd.com

IG MANUFACTURER WEBSITES

- Baxter: www.baxter.com
- Bio Products Laboratory: www.gammaplex.com
- CSL Behring: www.cslbehring.com
- Grifols: www.grifolsusa.com
- Kedrion: www.kedrionusa.com
- Octapharma: www.octapharma.com

Disease-State Resources

Ataxia Telangiectasia (A-T)

WEBSITES

- A-T Children's Project: www.atcp.org

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

WEBSITES

- GBS/CIDP Foundation International: www.gbs-cidp.org
- The Neuropathy Association: www.neuropathy.org

Evans Syndrome

ONLINE PEER SUPPORT

- Evans Syndrome Research and Support Group: www.evanssyndrome.org

Guillain-Barré Syndrome (GBS)

WEBSITES

- GBS/CIDP Foundation International: www.gbs-cidp.org
- The Neuropathy Association: www.neuropathy.org

ONLINE PEER SUPPORT

- GBS Support Group: www.gbs.org.uk
- GBS/CIDP Foundation International Discussion Forums: www.gbs-cidp.org/forums

Idiopathic Thrombocytopenic Purpura (ITP)

WEBSITES

- ITP Support Association – UK: www.itpsupport.org.uk
- Platelet Disorder Support Association: www.pdsa.org

Kawasaki Disease

WEBSITES

- American Heart Association: www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsofChildhood/Kawasaki-Disease_UCM_308777_Article.jsp#.T1T2boePWE0
- American Academy of Family Physicians: www.aafp.org/afp/2006/1001/p1141.html
- Kawasaki Disease Foundation: www.kdfoundation.org
- KidsHealth: <http://kidshealth.org/parent/medical/heart/kawasaki.html>

Mitochondrial Disease

WEBSITES

- United Mitochondrial Disease Foundation: www.umdf.org
- MitoAction: www.mitoaction.org

Multifocal Motor Neuropathy (MMN)

WEBSITES

- The Neuropathy Association: www.neuropathy.org

Multiple Sclerosis (MS)

WEBSITES

- All About Multiple Sclerosis: www.mult-sclerosis.org/index.html
- Multiple Sclerosis Association of America: www.msaa.com
- Multiple Sclerosis Foundation: www.msfocus.org/learn-about-multiple-sclerosis.aspx
- National Multiple Sclerosis Society: www.nationalmssociety.org

ONLINE PEER SUPPORT

- Friends with MS: www.FriendsWithMS.com
- MSWorld's Chat and Message Board: www.msworld.org

Myasthenia Gravis (MG)

WEBSITES AND CHAT ROOMS

- Myasthenia Gravis Foundation of America (MGFA): www.myasthenia.org

ONLINE PEER SUPPORT

- Genetic Alliance: www.geneticalliance.org

Myositis

WEBSITES



THE MYOSITIS ASSOCIATION

The mission of The Myositis Association, www.myositis.org, is to find a cure for inflammatory and other related myopathies, while serving those affected by these diseases. (202) 887-0088

- International Myositis Assessment and Clinical Studies Group: www.niehs.nih.gov/research/resources/collab/imacs/main.cfm

ONLINE PEER SUPPORT

- Juvenile Myositis Family Support Network: www.curejm.com/family_support/index.htm



The Cure JM Foundation
www.curejm.com
(760) 487-1079

- Michigan Immunodeficiency Foundation: www.facebook.com/groups/108048062584350
- Myositis Association Community Forum: tmacommunityforum.ning.com
- Myositis Support Group: www.myositisupportgroup.org
- Myositis Support Group – UK: www.myositis.org.uk

Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)

WEBSITES

- Guide to P.A.N.D.A.S. Syndrome: www.pandas-syndrome.webs.com
- P.A.N.D.A.S. Network: pandasnetwork.org
- Behavioural Neurotherapy Clinic – Australia: www.adhd.com.au/PANDAS.htm

Pemphigus and Pemphigoid

WEBSITES

- The International Pemphigus and Pemphigoid Foundation: www.pemphigus.org

Peripheral Neuropathy (PN)

WEBSITES

- Neuropathy Action Foundation: www.neuropathyaction.org



The Neuropathy Association,
www.neuropathy.org,

is devoted exclusively to all types of neuropathy, which affects upwards of 20 million Americans. The Association's mission is to increase public awareness of the nature and extent of PN, facilitate information exchanges about the disease, and advocate the need for early intervention and support research into the causes and treatment of neuropathies. (212) 692-0662

- Texas Chapter of the Neuropathy Association: www.handsfeetheart.org

Primary Immune Deficiency Disease (PID)



The Immune Deficiency Foundation (IDF), www.primaryimmune.org, is the national patient organization dedicated to improving the diagnosis, treatment and quality of life of persons with primary immunodeficiency diseases through advocacy, education and research. (800) 296-4433



The Jeffrey Modell Foundation, www.info4pi.org, is dedicated to early and precise diagnosis, meaningful treatments and, ultimately, cures for primary immunodeficiency. (212) 819-0200

WEBSITES

- The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/health/topics/Primary_Immunodeficiency.cfm
- American Academy of Allergy, Asthma & Immunology: www.aaaai.org
- International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: www.ipopi.org
- New England Primary Immunodeficiency Network: www.nepin.org



- Rainbow Allergy-Immunology: www.uhhospitals.org/rainbow/services/allergy-immunology
- Team Hope (for families and patients in New England): www.teamhope.info

ONLINE PEER SUPPORT

- IDF Common Ground: www.idfcommonground.org
- IDF Discussion Forum: <http://idffriends.org/forum>
- IDF Friends: <http://idffriends.org>
- Jeffrey Modell Foundation Message Board: www.info4pi.org

- Michigan Immunodeficiency Foundation: www.facebook.com/groups/108048062584350
- Rhode Island peer group: health.groups.yahoo.com/group/RhodelslandPIDD

Scleroderma

WEBSITES

- Scleroderma Foundation: www.scleroderma.org
- Scleroderma Research Foundation: www.srfcure.org
- Scleroderma Center: www.hopkinsmedicine.org/rheumatology/clinics/scleroderma_center.html

ONLINE PEER SUPPORT

- Scleroderma Support Forum: curezone.com/forums/f.asp?f=404

- International Scleroderma Network: www.sclero.org/support/forums/a-to-z.html

Stiff Person Syndrome (SPS)

WEBSITES

- American Autoimmune Related Diseases Association Inc.: www.aarda.org
- Genetic Alliance: www.geneticalliance.org
- Living with Stiff Person Syndrome (personal account): www.livingwithsps.com
- Stiff Person Syndrome: www.stiffpersonsyndrome.net

Other Resources

EDUCATION AND DISABILITY RESOURCES

- Americans with Disabilities Act of 1990: www.ada.gov Provides protection for people with disabilities from certain types of discrimination, and requires employers to provide some accommodations of the disability.
- Continuation of Health Coverage — Consolidated Omnibus Budget Reconciliation Act (COBRA): www.dol.gov/dol/topic/health-plans/cobra.htm
- Disability.gov: www.disability.gov U.S. federal government's disability-related information and resources.
- Individuals with Disabilities Education Improvement Act of 2004: idea.ed.gov/explore/home
- National Disability Rights Network: www.ndrn.org This website offers a search tool to find resources in your state to assist with school rights and advocacy.
- National Heart, Lung and Blood Institute: www.nhlbi.nih.gov/health/dci/Diseases/Itp/ITP_Whats.html
- Social Security: www.ssa.gov/disability
- U.S. Department of Education Website: www2.ed.gov/parents/landing.jhtml?exp=4 This federal government website offers a parents section titled "My Child's Special Needs."
- U.S. Department of Health and Human Services, Office of Civil Rights: www.hhs.gov/ocr/office/news/2008/discrimdisab.html Spells out civil rights under Section 504 of the Rehabilitation Act.

MEDICAL RESEARCH STUDIES

- ClinicalTrials.com: www.clinicaltrials.com This site has a registration form to request that you be notified about recruitment for future studies.
- ClinicalTrials.gov: www.clinicaltrials.gov A registry of federally and privately supported clinical trials conducted in the United States and around the world.

FOOD ALLERGIES

- Allergic Disorders: Promoting Best Practice: www.aaaai.org
- American Partnership for Eosinophilic Disorders: www.apfed.org
- Food Allergy and Anaphylaxis Network: www.foodallergy.org
- National Institutes of Health, National Institute of Allergy and Infectious Diseases (2004). Food Allergy: An Overview (NIH Publication No. 04-5518): www.niaid.nih.gov/topics/foodallergy/Pages/default.aspx
- Sicherer, S.H. (2006). *The Complete Peanut Allergy Handbook: Understanding and Managing Your Child's Food Allergies*
- World Allergy Organization: www.worldallergy.org

PRODUCT INFORMATION

- Influenza and the influenza vaccine: www.cdc.gov/flu or call (800) CDC-INFO: (800) 232-4636
- IVIG Carimune NF: www.cslbehring-us.com/docs/586/523/CarimuneNF_Oct%2010_PI,1.pdf

- IVIG Flebogamma 5% DIF and 10% DIF: www.grifols.com/portal/en/US/bioscience?div=8883
- IVIG/SCIG Gammagard Liquid: www.gammagardliquid.com
- IVIG Gammagard S/D: www.baxter.com/patients_and_caregivers/products/gammagard_sd_5.html
- IVIG/SCIG Gammaked: www.gammaked.com
- IVIG Gammaplex: www.gammaplex.com
- IVIG/SCIG Gamunex-C: www.gamunex-c.com/html/patient-what-is-gamunex-c.htm
- IVIG Octagam: www.octapharma.com/en/about-octapharma/products-therapies/immunotherapy/octagamr10.html
- IVIG Privigen: www.privigen.com
- SCIG Hizentra: www.hizentra.com

PUMP AND INFUSION SETS WEBSITES

- EMED Technologies: www.emedtc.com
- Marcal Medical Inc.: www.marcalmedical.com
- Intra Pump Infusion Systems: www.intrapump.com
- Micrel Medical Devices: www.micrelmed.com
- Norfolk Medical: www.norfolkmedical.com
- RMS Medical Products: www.rmsmedicalproducts.com
- Smith Medical: www.smiths-medical.com/brands/cadd

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