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The Pursuit of More Effective Treatments

I WAS A teenager when I saw the made-for-TV movie “The Boy in the Plastic Bubble” for the first time. The story about the “bubble boy” portrayed David Vetter’s life of isolation and loneliness before succumbing to complications brought about by his severe combined immunodeficiency (SCID). David was not much younger than I was when he died, when despite the age of modern medicine, death was inevitably the tragic consequence of diseases like SCID. Unfortunately for David, he was born with SCID at the time when there was no cure other than a bone marrow transplant from a matched sibling. Fortunately for the hundreds of children born with SCID each year today, that cure is no longer the only option, and they are alive and living active lives.

SCID is probably one of the more striking examples of the strides made in finding treatments and cures for life-threatening diseases. As our scientific expert Keith Berman details in his article “Severe PIs: Cutting-Edge Science Turns Tragedies to Cures,” the prognosis for children born with SCID has vastly improved with a “trifecta” of scientific advances. Not only have the prospects for successful bone marrow transplant improved by eliminating many of the complication risks, but TREC screening is gradually being embraced by states to diagnose SCID early enough to successfully treat it, and gene therapy trials are successfully replacing the missing proteins causing the immunodeficiency disorder. Equally important is that the advances in bone marrow transplants make them a viable cure for other PI disorders such as chronic granulomatous disease, Wiskott-Aldrich syndrome, X-linked lymphoproliferative disease and more.

Specific antibody deficiency (SAD) is another immunodeficiency disorder that for many years has been haunted by the controversy over the most effective treatment. In our article “Specific Antibody Deficiency: The Controversy Over Diagnosis and Treatment,” we explain that SAD is thought to affect some 5 percent to 20 percent of individuals who suffer from severe or recurrent infections. Yet, while some can be effectively treated with the aggressive use of antibiotics, many others continue to suffer frequent recurring illnesses because they fail to receive immune globulin (IG) therapy, which has shown to produce a clinical response. The reasons for denial of IG treatment range from the debate over the validity of the diagnosis, to IG not being on the list of FDA-approved therapies and the “resistance by third-party payers to cover” the treatment. All this, despite the recommendation for IG therapy by many of the major medical organizations.

Perhaps the most frustrating of the disease categories to find effective treatments is autoimmune disorders (ADs). IG is prescribed to treat a number of ADs, but the treatments generally require high doses of this therapy that is both expensive and, in some instances, in short supply. The goal, then, is to find ways to make IG therapy more effective in smaller doses. And, some exciting discoveries have been found. As described in our article “Autoimmune Disease: A More Effective Treatment on the Horizon?” scientists may soon be able to create both an enriched IG product and a recombinant one to treat ADs that can be used in smaller quantities with the same effect and that may reduce our dependence on the plasma supply.

As always, I hope you gain insight from the information presented and enjoy these and the many other articles in this edition of IG Living.
WITH THE MYRIAD of issues that people with chronic illness face each day — balancing illness with work, doctor appointments, frequent mobility issues, etc. — they shouldn’t have to deal with the stress of being denied access to care. Unfortunately, this happens frequently. While many different issues can affect a patient’s access to care, the two main policies identified by many patient advocacy groups are specialty tiers for medication and step therapy (also known as fail first).

Specialty tiering is a cost-saving method employed by payers that place drugs into different tiers. There may be anywhere from three to five tiers depending on the plan. Medications that are placed in any of the lower tiers of a plan have a fixed co-pay (for example, a flat rate of $20 per prescription). However, when medications are placed in the top tier of a plan, the insured is usually switched from the traditional co-pay to co-insurance. With co-insurance, the patient is required to pay a percentage of the cost of the drug instead of a flat rate. This percentage can range from 20 percent to 50 percent of the cost of the drug. A study conducted in 2013 by the Kaiser Family Foundation on employer health benefits found that 81 percent of covered workers are in a plan with three or more tiers, and that co-insurance is the most common form of cost-sharing in the highest tier.

There are many consequences to this policy. But, the most notable is that while the policy reduces the cost to the payer, it can be devastating to the patient. Most of the drugs, if not all of them, that are placed in the top tier are biologics that have no generic equivalents and can cost thousands of dollars. And, many of the patients who are prescribed the drugs that fall into the top tier suffer from chronic illness and are the least able to pay high out-of-pocket expenses. In fact, patients frequently have to choose between getting their medication and their financial wellbeing. Medical debt is one of the leading causes for bankruptcy in the United States.

Step therapy, also known as fail first, is another cost-saving method used by insurers. Step therapy mandates patients fail on a less-expensive medication before they can be prescribed a different drug that is more expensive but that may be more appropriate to their condition or that has a higher rate of effectiveness. Patient advocacy groups feel this policy is detrimental to patients in many ways. First, this cost-cutting policy doesn’t take into account an individual patient’s needs of which their treating physician is aware when determining a course of action. Therefore, it takes away the physician’s ability to prescribe what he or she feels may be the best medication for the patient, thereby undermining the physician-patient relationship. Second, the policy has been shown to increase the overall cost of healthcare by generating increased emergency room visits and hospital admissions. The key terminology in this policy is “fail first,” which means the patient must fail on one medication to be prescribed a more expensive drug. Failure means that the patient remains ill, suffers a relapse or has serious side effects.

Specialty tiering and step therapy have been identified as having the most impact on patients with chronic illness by patient advocacy groups throughout the country. It is very important that patients are aware of these issues that affect their access to care. If you have any questions about these policies, you can contact me on IG Living’s Ask the Experts page (www.IGLiving.com/AskTheExperts.aspx) or by emailing me at patientadvocate@IGLiving.com.

ABBIE CORNETT is the patient advocate for IG Living magazine.
FACES OF IG

Join the conversation! Connect with other immune globulin patients through IG Living’s Facebook page at www.facebook.com/IGLivingMagazine. Each day, we post interesting articles and facts, as well as thought-provoking questions that you can weigh in on. These are some snapshots of what’s being discussed.

Stress is part of life, especially when you live with chronic illness.

It does get so frustrating at times as you just want to have a normal life (describe normal) and people and doctors who actually truly care about you. And, yes, there [are] multiple stresses, and stress lowers the immune system. Consistent merry-go-round!

— Bella W.

No, it does not ever get any easier. That being said, you have to learn to adapt and realize that this is your new normal, and the alternative is no longer being alive.

— Rachel D.

Learning to live does. As you live with it, you learn new ways of taking care of yourself and coping, which makes it easier.

— Jamie S.

I agree with Jamie. I am going through a rough patch right now, possibly a relapse of my cancer. But finding something positive to focus on each day is what keeps me going. [I’m] counting my blessings.

— Debbie K.

How do you keep your home germ-free?

I had a germicidal UV lamp installed in the air-conditioning system to kill germs and mold, and I have a professional portable ozone generator I use when someone is sick. Ozone is a respiratory irritant, so you can’t be in the house when it’s on. Also, [I] have two large HEPA filters, one in the bedroom and one in the living room. Also the usual sweeping, mopping and laundering the bedding.

— Don F.

Constantly cleaning. When someone is sick, I send them to their room. After they are better, they clean up their beds. … Everyone is [an] adult in my home. This saves me from infections.

— Suzan B.R.

Have you ever felt discriminated against because of your illness?

Someone told a friend of mine [that] if I was as sick as I “pretended” to be, I should just go ahead and die.

— Kay P.B.

I have had someone call the police on me because I used a handicap parking spot, and I have a handicap placard. This woman said, “Oh, she’s not disabled; she must be using someone else’s placard.”

— Rachel D.
Matt » Short-term effects of the right amount of exercise should leave your mind and body feeling more invigorated, not completely worn out. Feelings of tiredness may catch up with you later in the day, but you should be able to get a good night’s sleep (at least seven hours) and wake up feeling refreshed, possibly looking forward to doing more yoga and/or taking another walk. If you are wiped out for the rest of the day after your exercise activity, or much more tired than you would have been had you not performed it, you are either overdoing it, or you aren’t getting enough rest or nutrition.

I like to use a fatigue scale of zero to 10 (similar to the pain scale, but in reverse). Imagine that a zero means absolutely no “gas in the tank.” A zero would mean that you couldn’t even sit up in bed on your own — any movement takes a lot of effort; a 10 would represent a “roaring engine” and a feeling that you could run to the top of the nearest mountain. Most of us don’t spend much time (if any) on either extreme of the scale. Record your feelings in an exercise journal. And, notice how the amount of sleep and exercise affects your fatigue scale number.

If you aren’t getting at least seven hours of sleep, I would recommend starting there and seeing how you feel afterward. If you are sleeping well, but still feel worn out, try cutting your exercise routine in half (i.e., half as long performing yoga and walking half the distance), and then see how you feel. If you’re just as fatigued, try halving it again and gradually work your way up. If your activity level doesn’t seem to have an impact on your fatigue, wipe you out for the rest of the day or make you more prone to infection, I would keep it up at your current level. Research has shown that exercising, but not to complete fatigue, can boost the immune system.

Lastly, it’s all right to exercise less or even take days off on those days that seem more challenging or impossible. Don’t be ashamed. Feelings of fatigue are the body’s way of telling a person that he or she needs more or less of something. Listen to it, but don’t allow yourself too many days of inactivity. The body also has a tendency to decondition quickly, and the inactivity/fatigue cycle can become self-perpetuating.

Michelle » GBS and CIDP are essentially the same condition, but GBS is acute and CIDP is chronic. As such, the symptoms of GBS occur over a period of days and weeks, and in CIDP, they occur over a period of greater than two months.

Doctors can typically diagnose GBS quickly because most patients go to the doctor or even to the emergency room right away because of the rapidly progressing symptoms. Once diagnosed, patients are treated one time with intravenous immune globulin (IVIG) or plasma exchange, and then begin physical rehabilitation.

CIDP takes a little longer to diagnose because the symptoms progress slowly, and people may or may not identify the neuropathy as CIDP. Treatment can include steroids, immunosuppressants and IVIG. Because CIDP is chronic, treatments continue for as long as it takes for the patient to respond, which means the patient’s sensation returns, balance and walking improve and strength improves.

More information about GBS and CIDP, as well as available links to organizations that can provide additional resources, can be found at www.nufactor.com/DS_CIDP.aspx and www.nufactor.com/DS_Guillain-BarreSyndrome.aspx.

Have a question? Email us at editor@IGLiving.com.
Your information will remain confidential unless permission is given.

MATTHEW D. HANSON, DPT, is a physical therapist and president of Freedom2Move (dba of SOMA Health, LLC).

MICHELLE GREER, RN, is senior vice president of sales at NuFACTOR Specialty Pharmacy.
How to Make a Diagnosis of an Antibody Deficiency,
Summarization Part 1
By Terry O. Harville, MD, PhD

IN THE PAST several issues, specific case scenarios of antibody deficiencies have been presented. These cases described the patient’s clinical features, patient and family histories, and the laboratory studies conducted for diagnosing an antibody deficiency. Here, we will summarize the important concepts that were developed in those vignettes.

Even though the disease manifestations, history and laboratory studies are somewhat unique for each person, important generalizations can be made. Five items, two clinical and three laboratory, must be considered for categorizing patients. These items can help us understand more about the disease and why certain situations make it easier or more difficult to make a specific diagnosis, and determine which therapeutic approaches may be of potential greater benefit.

The clinical categorizations include infections and autoimmune manifestations, which can range from non-existent to the most severe. Physicians will look at the number, severity, how common or uncommon the infections are, as well as the pattern of infections, which are unique to each patient. The typical infections of antibody deficiencies are described as sinopulmonary involving sinusitis, otitis, pharyngitis, bronchitis and pneumonia, although all of these usually do not occur in the same patient. Unusual or opportunistic infections (those not typically expected to occur) can also be helpful in indicating the presence of an immune system problem.

By definition, we expect someone with an immunodeficiency to be having problems with chronic and/or recurrent infection. But there can be rare occasions when the initial presentation of symptoms is autoimmune in nature. Autoimmune manifestations such as hemolytic anemia due to the production of anti-RBC autoantibodies or a diagnosis of autoimmune thyroid disease are easy to recognize as autoimmunity, but they may be difficult to sort out. This is because when infections occur, the immune system releases many chemicals and messengers (commonly, cytokines and chemokines) within the body, some of which result in fevers that generally cause inflammation. But, they can also cause achiness, fatigue, malaise and the general feeling of being unwell. When autoimmunity occurs, many of these same cytokines and chemokines are released by the immune system, also causing inflammation. Therefore, it can be difficult to know if the inflammation is being driven as a result of chronic infections or autoimmunity. After therapy begins, if the infections are brought under control but achiness and low-grade fevers persist, then there is a greater chance that some form of autoimmunity is present.

The laboratory components are straightforward, mainly because they are objective measurements rather than subjective reports. Serum immunoglobulin measurements, primarily the IgG level, but also IgA, IgM and IgE levels, play an important role in establishing a specific diagnosis. Pre/post-immunization antibody responses to the pneumococcal vaccine is the most critical component for establishing a diagnosis in most cases, since a significant number of patients may not have substantially decreased immunoglobulin levels or reduced B lymphocyte counts. Blood B lymphocyte count, which may not be performed in every patient due to the lack of availability of testing, can be very useful for categorizing the extent and type of immunodeficiency disease present.

Next time, we will examine the patterns these categories create and how specific or different diagnoses may, as a consequence, be classified.

TERRY O. 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.
**Autoimmune Disease: A More Effective Treatment on the Horizon?**

By Elissa Ritt, MAS

**INTRAVENOUS IMMUNE GLOBULIN** (IVIG) is a life-changing and, in many cases, life-saving treatment. IVIG is used to treat a multitude of disease states from the more familiar primary immunodeficiencies and autoimmune neuropathies to the more esoteric pemphigus vulgaris and even recurrent miscarriages. Treatment dosages for autoimmune diseases that range from 500 milligrams to 2 grams per kilogram of patient body weight (usually given monthly) result in a massive amount of IVIG being used by the healthcare system. Not only is IVIG a very expensive treatment, but because it is made from a limited resource, shortages have occurred. Therefore, researchers are driven to find ways to make IG therapy more effective at smaller doses.

**Figure 1. Antibody Structure**

![Antibody Structure](image)

**The Sialic Acid Discovery**

In 2008, Dr. Jeffrey Ravetch and his team at The Rockefeller University made a molecular discovery that could potentially be used to improve the anti-inflammatory effects of IVIG. Dr. Ravetch noted that a small number of the antibodies found in IVIG are different from the others; they exhibit a greater affinity to receptor sites that, when activated, blunt the immune response. This small, distinct subset of antibodies has a molecular entity called a sialic acid group attached to one end.

As shown in figure 1, antibodies are Y-shaped molecules. The stem of the Y is referred to as the Fc region, or heavy chain, which activates the Fc receptors involved in immune response. These Fc receptors appear to have a far greater affinity for the antibodies that have a

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**Sialic Switch Technology**

- Some IgG antibodies carry a sugar called sialic acid on the Fc portions of the molecule.
- Sialic acid is at the root of anti-inflammatory activity.
- A small fraction of IgG antibodies found in IVIG solution carry sialic acid.
- Enriching IVIG with IgG antibodies with sialic acid increases its anti-inflammatory activity by a factor of 10.
- Soon, it may be possible to create a recombinant form of IgG with a sialic acid molecule.
- Using sialic switch technology, researchers could make a form of IVIG or a recombinant drug to treat autoimmune diseases that is more anti-inflammatory.
sialic acid group attached to the Fc region, and when these sialylated antibodies activate the Fc receptors, the inflammatory response ceases (see figure 2).² Prior to this discovery, Dr. Ravetch and his team discovered that antibodies without a sialic acid group might be pathogenic because they actually promote autoimmune disease in mice.³

Dr. Ravetch and his team are now faced with how to turn this knowledge into more effective treatments for autoimmune disease. It has previously been demonstrated that enriching IVIG with sialic acid-linked antibodies results in a greater anti-inflammatory response.⁴ Even more exciting is that Dr. Ravetch and his team are able to create recombinant (laboratory made) sialylated Fc antibody regions that show a similar enhanced anti-inflammatory response.¹ This means that autoimmune diseases could be treated more effectively at smaller doses using “sialic-switch” technology — either sialic acid-enriched IVIG or a drug that makes use of recombinant sialylated Fc antibody regions.⁵ Additionally, the use of a laboratory made molecule instead of a plasma-derived antibody could reduce dependence on plasma supply and even result in less frequent drug shortages.

**Commercial Development**

Sialic switch technology has been licensed to Momenta Pharmaceuticals in hopes of it commercializing an enhanced autoimmune disease treatment. While Momenta intends to continue to study the potential benefits of sialic acid-enhanced IVIG, it is looking to add recombinant products using the technology to their product pipeline in the near future.⁶

There’s no denying that IVIG has enhanced and even saved the lives of many autoimmune disease patients. But, the cost of therapy, large dose size and limited raw materials are serious limitations to an otherwise efficacious, well-tolerated therapy. If Momenta succeeds in exploiting the sialic switch technology, those with autoimmune disease could benefit from improved therapies within just a few years.²

**ELISSA RITT**, MAS, is medical science liaison for NuFACTOR Specialty Pharmacy.

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**Figure 2. The Sialic Acid Sweet Spot**


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**References**

IN THE NEWS

Technology

IG Living App Now Available for iPhones and Androids

A wide range of IG Living’s information and resources can now be accessed from a tablet or smartphone. The new IG Living app allows individuals to view the current issue and all back issues of IG Living magazine. In addition, they can view, post and comment on IG Living’s Facebook page, as well as read and comment on IG Living blogs. To get instant access to all IG Living has to offer at any time or any place, simply download the IG Living app from any Apple app store or through Google Play on an Android device.

Manufacturer News

Second Octapharma Octagam 10% Manufacturing Site Approved by FDA

The U.S. Food and Drug Administration (FDA) has approved Octapharma’s manufacturing facility in Vienna, Austria, for the production of Octagam 10% (immune globulin intravenous [human] 10% [100 mg/mL] liquid preparation), which became available in the U.S. in October. This approval means that Octagam 10% can now be manufactured for the U.S. market at FDA-licensed facilities in both Stockholm and Vienna.

In July, FDA approved Octagam 10% for the treatment of adults with chronic immune thrombocytopenic purpura, a platelet disorder that can result in easy or excessive bruising and bleeding. Octapharma has been marketing Octagam 5% (immune globulin intravenous [human] 5% [50 mg/mL] liquid preparation) to treat primary humoral immunodeficiency since 2004. Octagam 5% is also manufactured at FDA-licensed manufacturing facilities in Stockham and Vienna.

“The FDA approval of Octapharma’s Vienna manufacturing site for Octagam 10% is great news for patients, as it will help facilitate product availability and enhance production flexibility,” said Flemming Nielsen, president of Octapharma USA. “Octapharma owns six manufacturing facilities internationally, which all utilize the latest technology and strict quality control processes.”

Product News

FDA Approves First Pathogen Reduction System to Treat Plasma

The Intercept Blood System for plasma, the first pathogen-reduction system for use by blood establishments in the preparation of plasma to reduce the risk of transfusion-transmitted infections, has been approved by the U.S. Food and Drug Administration (FDA). The system, marketed by Cerus Corp., can be used to reduce pathogens in plasma derived from whole blood and plasma obtained by apheresis, a collection process that separates red blood cells from plasma and then returns the red cells to the donor. Examples of some of the pathogens that could be reduced using the system include HIV, hepatitis B and C viruses, West Nile virus, prions and other unknowns. The inactivation of certain potential pathogens in plasma treated with the system is achieved through a photochemical process involving a controlled exposure to ultraviolet light and amotosalen, a chemical that facilitates the inactivation process. The plasma is then purified to remove the chemical and its byproducts. “The approval of devices like the Intercept Blood System allows blood establishments to prepare plasma that carries a lower risk of transmitting infectious pathogens through transfusion,” said Karen Midthun, MD, director of FDA’s Center for Biologics Evaluation and Research.

Plasma prepared using the Intercept Blood System was evaluated in eight clinical studies with 704 patients, and the data to support the use of plasma treated with the system were obtained from clinical trials conducted in various clinical settings, including acquired clotting disorders associated with liver disease and thrombotic thrombocytopenic purpura. Adverse events experienced by patients who received plasma prepared using the system were comparable to those experienced by participants who received plasma that had not been treated with it.
**Education**

**New Educational Blog Debuts for the Nursing Community**

NuFACTOR Specialty Pharmacy has launched a new blog titled IG Nursing Notes. The blog is written by NuFACTOR staff who have expertise in a variety of specialties, and is intended to provide timely information, nursing trends, helpful hints and much more to the nursing community. “IG Nursing Notes is committed to empowering nurses by providing encouragement and educational content that supports professional development,” said Michelle Greer, RN, MBA, senior vice president of sales for NuFACTOR, which is dedicated to solving the chronic problems of affordability, availability and safety in disease state management for chronically ill patients. To view the latest posts, go to nufactor.com/blog.

**Research**

**Meta-Analysis Identifies Predictors of Childhood Immune Thrombocytopenia**

In an analysis of 54 studies conducted between 1975 and 2013 of children ages 3 months to 18 years who were newly diagnosed with immune thrombocytopenia, researchers found that female gender, older age at presentation and the presence of antinuclear antibodies were among the characteristics that predicted the disease in children. Age older than 11 years was associated with chronic immune thrombocytopenia, and patients who developed chronic disease were a mean 2.68 years older than those with disease resolution. Platelet counts at presentation and the presence of antinuclear antibodies also increased risk for development of chronic disease.

Researchers also observed a considerable protective effect of intravenous immune globulin (IVIG) alone against the disease. Those who were treated with a combination of methylprednisolone and IVIG were more likely to develop chronic immune thrombocytopenia than were those treated with IVIG alone. “The protective effect of IVIG is remarkable and needs confirmation in prospective randomized trials, as well as future laboratory studies to elucidate the mechanism of this effect,” wrote Katja MJ Heitink-Pollé, MD, and colleagues of University Medical Center Utrecht and Wilhelmina Children’s Hospital in the Netherlands.

**Education**

**Grifols Partners with Schools to Teach About Plasma**

Grifols has partnered with Johnston County schools in North Carolina to teach eighth-graders about blood plasma and other life-saving therapies that plasma produces. Grifols worked with school administrators and Johnston Community College to create “Discover the Plasma,” an educational module that middle school science teachers have started using this spring. Johnston County schools and the community college helped Grifols draft the program’s content, which educators designed to meet curriculum requirements. And Grifols worked with an educational vendor in Spain, where the company is headquartered, on the graphics, website and demonstration videos. The five-part course, which will be administered at three pilot schools, uses an interactive website, games and lab activities.

**People & Places**

ADMA Biologics, a late-stage biopharmaceutical company that develops, manufactures and intends to market specialty plasma-based biologics for the treatment and prevention of certain infectious diseases, has appointed R. Michael Blaese, MD, Roy F. Chemaly, MD, MPH, John DeVincenzo, MD, Richard O’Reilly, MD, Jordan S. Orange, MD, PhD, and E. Richard Stiehm, MD, to its newly formed scientific advisory board.
**Research**

**Study Confirms Screening Newborns for SCID Improves Survival Rate**

Patients with severe combined immunodeficiency (SCID) demonstrated improved survival when they underwent hematopoietic cell transplantation (HSCT) within a few months of birth, before the onset of infection or after the infection resolved, according to results of a retrospective analysis. The study could put pressure on states that have resisted adding the condition to their newborn screening programs.

The analysis, conducted by Sung-Yung Pai, MD, a pediatric hematologist at Dana-Farber/Boston Children’s Cancer and Blood Disorder Center, and colleagues with the Primary Immune Deficiency Treatment Consortium, evaluated data from 240 infants with SCID who underwent HSCT between 2000 and 2009. They found that those who received the transplants at the age of 3-and-a-half months or younger had a 94 percent five-year survival rate. Ninety percent of those older than 3-and-a-half months who underwent HSCT before the onset of infection and whose infections resolved before transplantation survived five years. Donor type was significantly associated with survival, with 97 percent of patients with sibling donors surviving five years, whereas 79 percent of those with mismatched-related donors who received T-cell-depleted grafts without conditioning survived five years. The five-year survival rate was 66 percent among those who underwent conditioning with mismatched-related donors, 58 percent among cord blood recipients and 74 percent among recipients of other grafts.

“This confirms that transplants for SCID work well in very young children, but it also shows that any child with this disease can be treated with a high likelihood of a cure with a transplant from a parent or unrelated donor, not just a matched brother or sister,” said Richard J. O’Reilly, MD, chair of the pediatric department and pediatric bone marrow transplant service at Memorial Sloan Kettering Cancer Center. “Irrespective of the transplant approach used, if the child is transplanted early — without infection — you will have an extraordinarily good result.”

In 2010, the Department of Health and Human Services recommended that all states screen for SCID. But, states decide for themselves which conditions to test, and just more than half of them currently screen for SCID. The analysis was published in the *New England Journal of Medicine.*

**Medicines**

**FDA Approves New Dosing Option for Hizentra**

Hizentra is the first and only 20% subcutaneous immune globulin. It received FDA approval in March 2010 as a once-weekly IgG replacement therapy to help protect people with PI against infections and was approved for biweekly dosing in September 2013. The approval for flexible dosing is based on pharmacoepidemiology (modeling and simulation). Clinical trials using these alternative Hizentra dosing regimens were not conducted.

The U.S. Food and Drug Administration (FDA) has expanded the administration options for Hizentra (immune globulin subcutaneous [human] 20% liquid) to include the ability to individualize therapy with flexible dosing for people with primary immunodeficiency disease (PI). Flexible dosing means treatment at regular intervals from daily to once every two weeks. "Patient preferences on infusion frequency, time and volume can differ for many reasons, so having a treatment option like Hizentra that can be customized to fit individual lifestyles is important to both patients and physicians who treat them,” said Ralph S. Shapiro, MD, director of the Midwest Immunology Clinic. “Most important, flexible dosing options with Hizentra give PI patients the freedom to manage their condition based on their specific needs, while still providing a consistent level of protection against infections.”

Medicines

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**Research**

Study Confirms Screening Newborns for SCID Improves Survival Rate

Patients with severe combined immunodeficiency (SCID) demonstrated improved survival when they underwent hematopoietic cell transplantation (HSCT) within a few months of birth, before the onset of infection or after the infection resolved, according to results of a retrospective analysis. The study could put pressure on states that have resisted adding the condition to their newborn screening programs.

The analysis, conducted by Sung-Yung Pai, MD, a pediatric hematologist at Dana-Farber/Boston Children’s Cancer and Blood Disorder Center, and colleagues with the Primary Immune Deficiency Treatment Consortium, evaluated data from 240 infants with SCID who underwent HSCT between 2000 and 2009. They found that those who received the transplants at the age of 3-and-a-half months or younger had a 94 percent five-year survival rate. Ninety percent of those older than 3-and-a-half months who underwent HSCT before the onset of infection and whose infections resolved before transplantation survived five years. Donor type was significantly associated with survival, with 97 percent of patients with sibling donors surviving five years, whereas 79 percent of those with mismatched-related donors who received T-cell-depleted grafts without conditioning survived five years. The five-year survival rate was 66 percent among those who underwent conditioning with mismatched-related donors, 58 percent among cord blood recipients and 74 percent among recipients of other grafts.

“This confirms that transplants for SCID work well in very young children, but it also shows that any child with this disease can be treated with a high likelihood of a cure with a transplant from a parent or unrelated donor, not just a matched brother or sister,” said Richard J. O’Reilly, MD, chair of the pediatric department and pediatric bone marrow transplant service at Memorial Sloan Kettering Cancer Center. “Irrespective of the transplant approach used, if the child is transplanted early — without infection — you will have an extraordinarily good result.”

In 2010, the Department of Health and Human Services recommended that all states screen for SCID. But, states decide for themselves which conditions to test, and just more than half of them currently screen for SCID. The analysis was published in the *New England Journal of Medicine.*
Autoimmune Corner

Research

Stem Cell Treatment for MS Improves Immune System and Extends Remission

A new report released in December shows that stem cell transplants might soon offer multiple sclerosis (MS) patients an effective way to stave off relapses and improve their overall neurologic condition. The report was a follow-up of a study conducted in 2011 of 24 patients who received hematopoietic cell transplant (HCT) or another treatment known as high-dose immunosuppressive therapy (HDIT). After three years, progression-free survival had a rate of 90.9 percent, while clinical relapse-free survival was at 86.3 percent. "In the present study, HDIT/HCT induced remission of MS disease activity up to three years in most participants," the authors wrote. "It may therefore represent a potential therapeutic option for patients with MS in whom conventional immunotherapy fails, as well as for other severe immune-mediated diseases of the central nervous system."

According to National Institute of Allergy and Infectious Diseases Director Anthony Fauci, “These promising results support the need for future studies to further evaluate the benefits and risks of HDIT/HCT and directly compare this treatment strategy to current MS therapies. If the findings from this study are confirmed, HDIT/HCT may become a potential therapeutic option for people with this often-debilitating disease, particularly those who have not been helped by standard treatments.”

The report was published in the Dec. 29, 2014, issue of *JAMA Neurology*.

Research

Sibling Exposure Reduces MS Risk

A new study shows that higher exposure to infant siblings during the first six years of life is linked to a lower risk of multiple sclerosis (MS). In the study, a team at the Menzies Research Institute in Hobart, Australia, studied a group of 136 people with MS and compared them with a group of healthy controls, with respect to their exposure to siblings in early life. Increasing duration of contact with a younger sibling aged less than 2 years in the first six years of life led to decreased risk of MS. One to three years of contact reduced the risk by 43 percent, three to five years of contact reduced the risk by 60 percent and more than five years of contact reduced the risk by 88 percent. Tests also showed altered immune responses with exposure to infant siblings.

The research was conducted in response to the interest in the hygiene hypothesis of disease, which suggests that those who are not exposed to common infections in childhood are more likely to develop allergic or autoimmune disorders. Further work is now needed to determine how childhood exposure to infection and its effect on the immune system govern the risk of MS.

Research

Marijuana Chemicals Could Help Treat MS

Researchers have found that some chemical compounds found in marijuana (cannabis) can help treat multiple-sclerosis (MS)-like diseases in mice by preventing inflammation in the brain and spinal cord. The study sought to determine if the known anti-inflammatory properties of cannabidiol (CBD) and terahydrocannabinol (THC), compounds known as cannabinoids that are unique to cannabis and have interesting biological effects, could be applied to the treatment of inflammation associated with MS, and if so, how. They took immune cells isolated from paralyzed mice that specifically target and harm the brain and spinal cord, and treated them with either CBD or THC. In both cases, the immune cells produced fewer inflammatory molecules, particularly one called interleukin 17, or IL-17, which is strongly associated with MS and very harmful to nerve cells and their insulating covers. The researchers concluded that the presence of CBD or THC restrains the immune cells from triggering the production of inflammatory molecules and limits the molecules’ ability to reach and damage the brain and spinal cord.

Further research is needed to prove the effectiveness of cannabinoids in treating MS in humans, but there are reasons for hope, the researchers say. In many countries, CBD and THC are already prescribed for the treatment of MS symptoms, including pain and muscle stiffness. The study was published in the Journal of Neuroimmune Pharmacology.
Severe PIs:
Cutting-Edge Science Turns Tragedies to Cures

For decades, donor blood stem cell transplantation has been the only potential cure for severe primary immunodeficiency disorders, but it has been limited by failure and serious complication risks. Now a trifecta of scientific advances is transforming the prognosis for children once defenseless against life-threatening infections.

By Keith Berman, MPH, MBA

IF YOU ARE of a certain age, you may remember heart-wrenching images and a long-running — and ultimately tragic — story about a Texas boy whose 12 years of life inside a plastic isolation bubble introduced Americans to the plight of children born with severe combined immunodeficiency (SCID). David Vetter, dubbed the “bubble boy” in countless news features, lived in a succession of sterile chambers to avoid the fate of his older brother, who had succumbed in infancy from complications of overwhelming infection from the same disorder.

From his mother, David inherited a defective version of the IL-2 common chain receptor gene (IL2RG) that she carried on one of her two X chromosomes. David’s X-linked disorder, SCID-X1, affects only boys and accounts for 40 percent to 50 percent of all SCID cases. Any of at least 300 different mutations can disable the IL2RG gene, which encodes a protein critical for regulating growth and maturation of T and B lymphocytes and other immune cells responsible for killing bacteria, viruses, fungi and other invasive pathogens.

The next most common form, ADA-SCID, accounts for 15 percent to 20 percent of cases and equally affects boys and girls. In this instance, the genetic defect results in a non-functional enzyme called adenosine deaminase (ADA), which, like SCID-X1, leads to profoundly low numbers of T lymphocytes (T cells), B lymphocytes (B cells) and natural killer cells.
Altogether, more than 20 recognized forms of “classical SCID” are characterized by a very low T-cell count with near-absent responsiveness to immunogenic stimuli. These patients not only have profound deficiencies in cellular immunity, but also have very poor antibody response when they come in contact with bacteria, viruses and other pathogens that infants with normal immune function (for their age) can readily fight off. Without immune reconstitution or the kind of extreme measures used to protect David Vetter, nearly all of these children will die from overwhelming infection by the second year of life.

With an incidence estimated at just one in 30,000 to 60,000 live births, children with SCID are referred to major academic medical centers that have well-trained specialists who can manage infections and order prophylactic antibiotic and immune globulin (IG) therapy, and map out a definitive treatment plan. For most patients with classical forms of SCID, definitive treatment is to attempt to reconstitute the immune system by intravenously administering functional hematopoietic stem cells (HSCs) sourced from donor bone marrow, peripheral blood or umbilical cord blood. Ideally, these cells engraft in the bone marrow and restore cellular and antibody-mediated immunity.

The (Improving) Promise of Cure: Donor HSCT

In 1968, pediatricians at the University of Minnesota were the first to infuse HSCs from the bone marrow of an HLA-matched sibling to achieve immunological correction of an infant with X-linked SCID. Over the ensuing four and a half decades, specialists have turned to HSC transplants (HSCTs) as a potentially life-saving treatment for SCID with mixed results. Some infants have experienced partial or complete restoration of immune function, while others have suffered engraftment failure or serious complications, including graft versus host disease (GVHD) and toxicity from preparative immunosuppressive “conditioning.”

A number of factors (Table 1) appear to importantly influence the prospects for successful HSC engraftment and survival. Prominent among them are 1) the type of stem cell donor, 2) the “conditioning” regimen prior to transplantation that facilitates donor cell engraftment, 3) recipient age at transplantation and 4) recipient infection status at transplantation.

For many years, availability of limited numbers of transplant procedures together with combinations of these presumptive “risk factors” that vary from one SCID patient to the next largely frustrated efforts of clinicians to discern which of them significantly impacted long-term survival after HSCT. Finally, a collaborative network of 25 U.S. and Canadian institutions — the Primary Immune Deficiency Treatment Consortium (PIDTC) — tasked itself with retrospectively gathering and analyzing demographic, treatment and long-term survival data from 240 infants with classical SCID who had undergone allogeneic (human donor) HSCT over a 10-year period from Jan. 1, 2000, through Dec. 31, 2009. This large case series, published in 2014, yielded a trove of valuable information to aid HSCT treatment planning.

The PIDTC data convincingly show that infants with SCID should undergo an HSCT procedure within the first 3.5 months of life, ideally before a first severe infectious illness. Unfortunately, infants with SCID appear entirely healthy at birth. Usually nothing appears amiss directly up to the first hospitalization with severe and potentially life-threatening infection, when finally a battery of tests reveals the absence of a functioning immune system. Thus, diagnosis of SCID has historically been reactive — usually after a first or second severe opportunistic infection. The median age of the PIDTC SCID cohort, for example, was nearly 140 days at diagnosis; the median age at transplant was six months, by which time the health of most infants had already been harmed by opportunistic infection.

Newborn TREC Screening Jump-starts SCID Therapy

Seemingly on cue, a practical population-based newborn screening for SCID arrived to detect this occult disorder: the T-cell receptor excision circle (TREC) assay. First used in patients with HIV and hematological malignancy, the TREC assay was adapted to utilize the dried blood spots already universally obtained by heel-stick from infants in the first days of life to screen for metabolic diseases, cystic fibrosis, hypothyroidism and hemoglobin disorders. The TREC copy number is a biomarker for the output of T lymphocytes (lymphopoiesis) from the thymus. A very low
TREC value identifies infants with SCID, who have profoundly decreased circulating naïve T cells, SCID-like disorders including “leaky SCID” and Omenn syndrome, and other non-SCID conditions associated with low T-cell counts.

How reliable is the TREC assay for identifying the rare case of SCID among the many thousands of unaffected babies? In 2008, Wisconsin became the first state in the U.S. to screen all newborns. The results of infant screening over the first three years (Figure 1) essentially tell the story. The specificity of the TREC assay — the proportion of healthy individuals correctly identified as test-negative — was remarkable 99.98 percent. This is important as too many “false positives” would unnecessarily create parental worry and drive up costs for fruitless additional testing. Equally if not more important was its 100 percent sensitivity: TREC screening identified every case of SCID.3

Of the 207,696 infants tested, 0.035 percent had an abnormal TREC screen. On further testing, normal T-cell counts were found in 53 percent of the infants with abnormal screen, but the remainder had varying levels of T-cell lymphopenia (low T-cell count). Of the patients with severe T-cell lymphopenia identified by the TREC assay, further testing determined that 58 percent had matched sibling donors have the best overall prognosis, but a recent analysis reveals 1) substantially high survival rates for boys under age 5 years who receive an unrelated donor transplant and an HLA identical sibling transplant and 2) a 77 percent five-year survival rate for boys transplanted after age 5 years.

### Transplants for PI Disorders Other Than SCID

The uniform lethality of SCID in infancy demands that clinicians and families accept some risks in pursuit of a realistic potential cure. This same principle applies for a number of other rare primary immunodeficiency (PI) disorders. Prominent among those often treated with HSCT are the following:

- **Chronic granulomatous disease (CGD)** is an inherited disorder of neutrophil function caused by mutations of an enzyme critical for phagocytic killing of intracellular pathogens. Severe and prolonged infections of the lungs, lymph nodes and skin are frequently found at diagnosis. Life expectancy is short, with only about one-half of affected individuals still alive by age 30. First performed in the mid-1980s, HSCT remains the only curative therapeutic option.

- While selection of which CGD patients would benefit from HSCT is still debated, those with signs and symptoms suggesting a guarded prognosis may be the most appropriate candidates. After HSCT using well-matched donors, 18 of 20 children and young adults in a recent case series were alive at four to 117 months (median 61 months), with normal neutrophil function. Colitis affecting 10 of these patients resolved, and all seven others with growth failure experienced catch-up growth.

- **Wiscott-Aldrich syndrome (WAS)**, an X-linked recessive disorder that affects about four in every one million male births, is associated with a life expectancy of less than 20 years. Most patients die of infections, malignancy, autoimmune-related illness or bleeding complications.

- Five-year survival following HSCT, which is the only curative therapy for WAS, now stands at about 90 percent. Patients with matched sibling donors have the best overall prognosis, but a recent analysis reveals 1) comparably high survival rates for boys under age 5 years who receive an unrelated donor transplant or an HLA identical sibling transplant and 2) a 73 percent five-year survival rate for boys transplanted after age 5 years.
More than

10,000

patients and providers put their confidence in Hizentra.¹

“I choose daily.”

Hear perspectives from Melaine and other Voice2Voice® Advocates at: www.Hizentra.com/Perspectives. You can also sign up for Voice2Voice to connect with others who have walked in your shoes.

Melaine, Mom, Yoga/Cycle Instructor & Voice2Voice Advocate

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.

*Voice2Voice advocates are not healthcare professionals or medical experts. For medical questions, patients should contact their physicians. Voice2Voice advocates are compensated by CSL Behring LLC for their time and/or expenses.

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

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.

Hizentra®. Immune Globulin Subcutaneous (Human), 20% Liquid
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 at regular intervals from daily up to every two weeks (biweekly).

Dosage (2.2)

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
  
  Initial weekly dose = Previous IGIV dose (in grams) $\times 1.37$
  
  No. of weeks between IGIV doses

- **Biweekly**: Start Hizentra 1 or 2 weeks after the last IGIV infusion or 1 week after the last weekly Hizentra/IGSC infusion. Administer twice the calculated weekly dose.

- **Frequent dosing (2 to 7 times per week)**: Start Hizentra 1 week after the last IGIV or Hizentra/IGSC infusion. Divide the calculated weekly dose by the desired number of times per week.

- **Adjust the dose based on clinical response and serum IgG trough levels.**

**Administration**

- **Infusion sites** – 1 to 4 injection sites simultaneously, with at least 2 inches between sites.

<table>
<thead>
<tr>
<th>Infusion Parameters</th>
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<tr>
<td>Volume (mL/site)</td>
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<td>Rate (mL/hr/site)</td>
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*As tolerated

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**DOSE 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]).
- Hizentra is made from human plasma and may contain 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 $\geq 5\%$ of study subjects were local reactions (i.e., swelling, redness, heat, pain, and itching at the injection site), headache, 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](http://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 January 2015 revision
suggesting that the transplantation option may be worth considering for those with good clinical status. 10

- *X-linked lymphoproliferative disease (Duncan syndrome)* is a rare PI disorder affecting one to two boys per million. Mortality is high in part due to a unique susceptibility to Epstein-Barr virus, which can result in fulminant fatal disease. Once again, HSCT is the only curative treatment.

A multicenter study of 91 patients — 43 treated with HSCT and 48 not transplanted — documented an overall survival of 81 percent in the HSCT group and 62 percent in the non-transplanted group. Still under debate is whether a newly diagnosed child who is asymptomatic should receive HSCT therapy, as intravenous IG (IVIG) treatment can be instituted promptly; there is general consensus among experts that those with a matched sibling donor should be transplanted.8

Several other PI syndromes for which HSCT has been shown to dramatically improve the prospects for long-term survival include *hemophagocytic lymphohistiocytosis (HLH)*11 and *IPEX (immunodysregulation, polyendocrinopathy, enteropathy, X-linked syndrome)*.12,13

**Gene Therapy: The Promise of Cures Becomes Reality**

Donor HSCT is clearly curative for the majority of infants with SCID. But major problems persist. GVHD and other transplant-related complications commonly occur in the large share of infants for whom there is no fully matched sibling or family donor. Many children continue to have poor B-cell function and remain dependent on long-term IG therapy. Incomplete immune reconstitution following transplantation leaves many children at ongoing risk for serious opportunistic infections; residual immunodeficiency after partially HLA-incompatible HSCT is still responsible for an estimated 30 percent mortality rate at one year post-transplantation.14

For many years, immunologists have appreciated that — if achievable without introducing new health risks — the ideal curative therapy is to harvest bone marrow from the patient and “transduce” his or her own (autologous) HSCs by using special viral “vectors” to insert normal copies of the mutated gene. These normal genes can in turn produce the critical missing functional protein causing the immunodeficiency disorder.

Two small early trials, one in France15 and other in the United Kingdom,16 proved that autologous CD34-positive hematopoietic bone marrow stem cells transduced with a gammaretroviral vector delivering IL2RG and reinfused into patients with SCID-X1 resulted in a sustained restoration of both cellular and humoral immunity.

Unfortunately, less than three years after their triumphant findings were published in *Science* in 2000, the French investigative team reported two cases of leukemia. Eventually, five of 20 SCID-X1 patients developed leukemia, attributed to “insertional mutagenesis” that resulted when gammaretroviral vectors activated a known T lymphocyte oncogene. In January 2003, the U.S. halted more than two dozen gene therapy studies that utilized those vectors.

Several years later, researchers returned to the clinic with redesigned vectors to ferry functional genes, including novel “self-inactivating” gammaretroviral vectors and lentiviral vectors incorporating safety features intended to minimize the risk of inducing leukemia. A wave of early trial results strongly suggest that gene therapy is safe and curative in patients with SCID-X1 and ADA-SCID.
Gene therapy for SCID-X1. In a report on parallel trials in the U.S. and Europe, autologous bone marrow-derived HSCs transduced with a self-inactivating retroviral vector carrying a normal copy of the IL2RG gene were reinformed into nine infant boys with confirmed SCID-X1, including profound deficiency of autologous T cells. All were over age 3.5 months at the time of treatment; the median age was 8 months. All nine patients either lacked an HLA-identical related or unrelated donor or had an active treatment-resistant SCID-related infection. One patient with preexisting severe systemic adenosiviral disease died before he could be fully reconstituted with vector-modified CD34-positive HSCs. Another who received a graft with a low number of vector copies did not show evidence of vector DNA uptake and later received a mismatched umbilical cord blood transplant.

At a median follow-up of 29 months, all eight treated boys were still alive, and seven of the eight experienced T-cell proliferative capacity in the normal range, and associated functionality that led to resolution of their infections. No severe adverse events related to the gene-transfer vector or cell manipulations were reported in any of the children. At a median of 33 months of follow-up, there was no occurrence of leukemia. These children are scheduled to be followed and periodically tested over the next 15 years.

Gene therapy for ADA-SCID. In 2009, a multinational team reported that nine of 10 children receiving autologous HSCs transduced with a retrovirus carrying the functional ADA gene experienced immune reconstitution with increases in T-cell count and normalization of T-cell function. At a median follow-up of four years, all 10 patients were alive, with no reports of leukemia or other serious adverse outcomes. Eight no longer required ADA replacement therapy. The number of severe infections decreased from 0.93 per 10 person-months before gene therapy to 0.13 following gene therapy; the median number of hospitalizations days dropped from 45 to two.

A separate clinical study in the UK subsequently affirmed that, when it works, gene therapy for ADA-SCID resolves the profound T- and B-cell immunodeficiency and appears to all but eliminate the risk of severe opportunistic infections. In an astounding statement last November, Dr. David Kohn, the lead investigator of two trials of a gene therapy regimen developed at UCLA, reported that all 18 treated infants with ADA-SCID have been cured. The UCLA team plans to seek FDA approval for its gene therapy regimen.

What Comes Next

In this dawning era of TREC screening, clinicians can now identify infants with SCID at birth and employ HSCT to reconstitute a functional immune system during the critical first few months of life, with improved prospects for partial or complete cures. HSCT is already the gold standard treatment for SCID, but going forward we can expect to see fewer treatment failures and higher overall long-term survival statistics. This experience may additionally help clinicians make further refinements to HSCT therapy for other severe PI disorders, potentially leading to its use in more patients with better outcomes.

Remarkably, at the same time, new clinical research suggests that gene therapy is finally poised to realize its curative potential for infants diagnosed with SCID-X1, ADA-SCID and someday, hopefully, other genetically well-characterized PI conditions as well.

In the very near future, clinicians may find themselves in the position of making patient-by-patient decisions about when to perform HSCT and when to turn to gene therapy for SCID or certain other severe PI disorders. Given the stakes involved for these precious new beings and their parents, it is a future they can look forward to.

References


KEITH BERMAN, MPH, MBA is the founder of Health Research Associates, providing reimbursement consulting, business development and market research services to biopharmaceutical, blood product and medical device manufacturers and suppliers. Since 1989, he has also served as editor of International Blood/Plasma News, a blood products industry newsletter.
Specific antibody deficiency occurs in both children and adults, but its diagnosis and treatment continue to be controversial.

By Ronale Tucker Rhodes, MS

**FIRST REPORTED IN** a small group of patients in the early 1980s, specific antibody deficiency (SAD) is one of the most commonly identified immune abnormalities among patients presenting with recurrent and/or severe sinopulmonary infections.¹ SAD affects both males and females and all age groups, but it is more often diagnosed in adults. It is unknown what the prevalence of SAD is in the general population in the U.S. Some studies show that SAD is recognized in 5 percent to 20 percent of children older than 2 years who suffer from recurrent or severe infections, and one study determined the prevalence of SAD among adults with recurrent community-acquired pneumonia to be approximately 8 percent.²

Why, then, despite the prevalence of SAD, does controversy about its diagnosis and treatment exist, thus prolonging the diagnosis and treatment options for many?
What Is SAD?

According to Terry Harville, MD, PhD, medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, some immunologists disagree that SAD is an immunodeficiency. Yet, in 2014, the International Union of Immunological Societies (IUIS) Expert Committee for Primary Immunodeficiency lists SAD in its table of primary B lymphocyte/immunoglobulin deficiencies. According to IUIS, SAD is characterized by normal, and in some instances, somewhat low-normal B lymphocyte numbers with a deficiency to specific antigens.³

Sometimes termed “partial antibody deficiency” or “impaired polysaccharide responsiveness,” SAD describes a deficient-specific response to pneumococcal polysaccharide antigens in individuals who have normal responses to protein antigens, normal serum levels of immunoglobulins and normal IgG subclass concentrations.¹ It is a common antibody immunodeficiency defined as a poor antibody response to unconjugated pneumococcal polysaccharides present in the 23-valent pneumococcal vaccine.⁴ In essence, in SAD patients, the total quantity of antibodies that fight infection are normal, but the quality of those antibodies, or the ability for them to fight infection, is not.

“Typically, the immunoglobulin classes IgG, IgA and IgM are all in the normal ranges for age,” explains Dr. Harville. “Yet, the pneumococcal titers are low.” Immunoglobulins are antibodies produced by plasma cells that are used by the immune system to identify and neutralize foreign objects such as bacteria and viruses. But, there are also specific antibodies that the immune system produces in response to specific vaccines. The pneumococcal vaccine is used to test for the response to polysaccharide antigens that are found on the surfaces of most bacteria and other pathogens that cause infections. Measuring the pneumococcal titers reveals how well the body is able to make those specific antibodies to eradicate bacteria and infections.³
Symptoms of SAD

Most frequently, the symptoms of SAD are the same as other primary immunodeficiency disease symptoms. They can be as severe or in some cases moreso than some patients with a greater extent of antibody deficiency. These include recurrent ear infections, sinusitis, bronchitis and pneumonia. When symptoms begin differs. Some people present with infections with increased frequency during the first years of life, while the onset of infections for others may occur later in life.6

What Causes SAD?

The precise underlying mechanisms resulting in SAD remain unknown. Ongoing research is trying to elucidate the cause of SAD and how it may be inherited.6,7

Diagnosing SAD

Presentation with a history of recurrent infections is typically indicative that an antibody deficiency may be present. However, SAD can be a diagnostic challenge because total IgG levels are normal, whereas antibody deficiencies typically present with either absent or deficient total IgG levels.

Most frequently, the symptoms of SAD are the same as other primary immunodeficiency disease symptoms, and in some cases, more severe.

Diagnostic criteria for SAD include measuring the levels of the three main classes of immunoglobulin (IgG, IgA and IgM); using tetanus, pneumococcus and haemophilus influenza type b indicators to check whether specific immunoglobulins are present that can fight infection; testing the immune system’s response to vaccination if specific immunoglobulins are low; and counting the numbers of different immune cells in the blood (which should be normal in SAD).7

The problem, says Dr. Harville, is that the proper testing isn’t always performed. “A patient shows up to a clinic with recurrent infections, sinus problems, upper-respiratory problems, fatigue, headaches, and they just don’t feel good — a nonspecific set of symptoms that suggests a problem with infections. Because there is a lack of a cure with antibiotics, the decision is made to measure the serum IgG levels, and they’re normal,” he explains. And, frequently, that’s where the testing stops. But, what should be done, he says, is to have that patient tested for pneumococcal titers as well. If that person’s pneumococcal titers are low, then the diagnosis of SAD should be considered.8

The response to the pneumococcal vaccine, according to the Immune Deficiency Foundation (IDF), is why the criteria for the diagnosis of SAD have been somewhat controversial. Now, however, IDF says most immunologists agree that several patterns of responses after receiving the vaccine can be used to diagnose SAD. “Patients may fail to respond to any of the serotypes included in the vaccine and have a more severe form of SAD,” writes IDF. “Responses in which children respond to less than 50 percent of serotypes and adults respond to less than 70 percent have a moderate form of SAD with an increased risk of upper- and lower-respiratory infections that may warrant treatment. An additional subset of patients appears to respond normally initially [and] then lose protective levels within months.”6

Treating SAD

As mentioned previously, treatment for SAD can be controversial. Many patients’ infections can be controlled with the aggressive use of antibiotics, which has been the conventional treatment for SAD. But this is not true for many other patients who continue to have more frequent and severe infections. For these latter patients, many immunologists recommend intravenous immune globulin (IVIG) replacement therapy. If there is a clinical response (infections stop), it is still recommended that treatment be stopped after a period of time (typically in the spring) to determine if a deficiency still exists. This is especially true for children because many outgrow SAD as they get older.6

The patients’ immune response would then be re-evaluated at least five months after discontinuation of IVIG.7 If a deficiency still exists, IVIG would be restarted.

IVIG, however, is not a U.S. Food and Drug Administration-approved therapy for SAD.9 Therefore, there is disagreement over whether someone diagnosed with SAD should receive IVIG therapy. “There are those who suspect that SAD is not a ‘true’ disease category, especially one deserving IVIG treatment,” explains Dr. Harville. Indeed, because of the doubt over the validity of the diagnosis, “there is resistance by third-party payers to cover IVIG treatment,” he says. One insurance company’s policy states that SAD “generally does not require
Jennifer Wang’s Story

For well over a decade, Jennifer Wang was sick with allergies, asthma, respiratory infections, sinus infections and the list goes on. Her symptoms started in her early teens. Even with back-to-back rounds of antibiotics and two sinus surgeries, she was always sick. “It’s disheartening to be sick and not know what’s wrong with you,” explains Jennifer. “You feel angry and confused, and it definitely has an impact on your psyche.” Jennifer had been seeing the same allergist/immunologist since 2004 when in mid-2013, “he finally figured it out.” Diagnosis: specific antibody deficiency (SAD). “That’s the problem with SAD; we have to get sick enough to finally figure it out,” says Jennifer.

Because Jennifer had been on different rounds of antibiotics for several months and she had had sinus surgery, neither of which stopped the recurrent infections, when she was diagnosed, she was prescribed intravenous immune globulin (IVIG). “After the insurance approved it, I was treated with IVIG,” says Jennifer. “I’ve never been denied, just delayed.” Jennifer was treated with IVIG for seven months, and she then switched to subcutaneous IG (SCIG) infusions. “I was doing SCIG weekly at first, but I was feeling down on days, like a roller coaster. So, my doctor switched me to every six days,” she explains. “That little tweak was exactly what I needed.” Jennifer is still on daily antibiotics, but she has switched from high dose to low dose, and she is hoping she eventually won’t need them.

The challenges Jennifer, now 28, has faced are many. When she was sick and didn’t know what was wrong with her, she said she always faked that she was fine. “You don’t tell people you’re sick. It’s this big thing looming over you that you hide,” explains Jennifer. But, since her diagnosis, she’s open to talking about it: “The shame and guilt sort of went away with the diagnosis.” Fortunately for Jennifer, she doesn’t have to work full time, and she has a flexible schedule. “It would be very hard to have to deal with an employer, especially with my every-six-day infusion schedule,” she says. “I’ve had the time to concentrate on my health.” But the one thing she’d really like to do she can’t: travel to China to see her husband’s family. “I went to China once and I was so sick. I thought maybe it was because they all smoke,” she explains. “I wish I could go again, but I don’t know if I am able with my disease.”

IG therapy has made all the difference for Jennifer. In college and graduate school, she was a professional belly dancer. Now, she does samba dancing. “I’m in a dance group called sambacolorado.com, and I dance 10 to 15 hours a week,” says Jennifer. “I wouldn’t be able to do that without IG therapy. I went through feeling like I only have the energy to do a load of laundry to being able to go to samba five days a week.”

“I know there’s no cure,” says Jennifer. “So, right now, the expectation is a lifelong treatment, which at first was a hard thing to wrap my mind around. Everyone wants to just be normal.” One of the things that helps Jennifer is keeping educated about her disease. “I’m still in discovery mode, and the more I find out, the clearer the picture becomes. I’d give up everything I have for a cure; I’m just glad with IG therapy, I can live a somewhat normal life.”

Jennifer’s passion now is spreading awareness about the disease. “I’m trying to figure out where my role is going to be in the advocate world now,” she says. “My heart feels like I need to do something, because it would almost be a crime to have this illness and not spread awareness.”

Jennifer’s advice: “If you feel like there’s something wrong with you beyond allergies and asthma, speak up. And, don’t feel ashamed of having the disease. You can still have a life. You may not be able to climb Mount Everest, or in my case travel the world, but you can still do a lot of things. I can dance for hours in high heels. Don’t let [SAD] suck the life out of you. Find what makes you happy. Life is not over.”
Coryn Barks’ Story

Laura Harland’s daughter, Coryn, was sick right after she was born. “She always had something. We were at the doctor every week,” explains Laura. “As soon as she got off an antibiotic, she was on another for something else. She had the typical childhood infections: ear infections, strep, sinus, pneumonia, colds, etc.”

But getting a diagnosis for Coryn wasn’t easy. “It was beyond frustrating,” says Laura. “I honestly don’t know how people who have gone longer without a diagnosis handled it. [Coryn] wasn’t growing or thriving because she was sick all the time. The ER doctors and nurses knew us because she was there all the time. I knew there was something wrong. It’s heartbreaking to see your child go through that and to be told she is normal when you know she is not.” Coryn was originally diagnosed with an IgG subclass deficiency at age 2. But, after additional testing for pneumococcal titers, she was diagnosed with SAD.

Coryn was treated with prophylactic antibiotics for about two years, which kept her from getting pneumonia. However, she did continue to have upper-respiratory and sinus infections. “So, at one of our immunology checkups, the doctor asked how she was doing. I was thrilled to tell him she hadn’t had pneumonia. He asked about other infections, since she had continued to have many, and that’s when he decided it was time to try immune globulin,” says Laura. Coryn was 4 years old when she started intravenous immune globulin (IVIG) therapy, and she was treated with it for 13 months, when she had a strep infection and the doctor decided to check her levels. “It turned out that her B cells were normal when they hadn’t been before, so he decided to take her off of IVIG to see if her immune system would start working,” explains Laura. “She did well for a couple of months, and then she started getting sinus infections again. She then had her adenoids removed, and she did well for a few month, but then she started getting sick again, and we couldn’t get her better. As soon as an infection was gone, a week later it was back. So, after 19 months, the infusions were started again.” Coryn is now 8 years old and is still being treated with IVIG every four weeks.

Trying to make others understand that Coryn’s infections represent a serious illness has been difficult. When Coryn gets sick, she doesn’t get fevers. “A fever for her would be 99.5,” says Laura. “It’s almost like her body doesn’t realize she’s trying to fight something — even when she has the flu. So when she goes to the nurse and says she doesn’t feel good and she doesn’t have a fever, she isn’t believed. That’s terrible for her self-esteem.” Coryn, then, tries to portray being fine because she doesn’t want people to know what’s going on. “Coryn has told me that she can act perfectly fine even when she’s miserable,” says Laura.

It’s tough for Laura, too. There are people she knows who don’t remember seeing Coryn when she was 2 or 4, when she was sick all the time. “People think strep throat or a cold isn’t that bad, but if it’s constant, it’s different, and people don’t understand,” explains Laura. “They don’t understand why I’m so passionate about vaccines and herd immunity because her body doesn’t accept vaccinations. I feel terrible as a parent for pulling her out of school for doctor appointments, treatments or illnesses and being looked at or judged when I have to pull her out of school because she’s too tired to stay.”

Still, Laura strives to make Coryn’s life as normal as possible. Coryn’s infusion days are made better because there is a little boy her age who also has an immune deficiency. “They sit in bed together and get their IV poles tangled up and go to the game room and play games,” says Laura. “Sometimes we go to the zoo after their infusions. It’s made a big difference for her to know that other kids have the same struggles as her and have to get poked as often as she does. She’s 8, and knowing someone else who is going through something similar makes it easier on her.”

Laura’s advice to other parents of kids with SAD: “Find someone who is going through the same thing. Meet another parent at an infusion clinic. Get on Facebook. No one truly is going to understand except someone who is going through it themselves. And having that person makes all the difference in the world.”
IVIG replacement for control of recurrent bacterial infections.” Yet, IVIG treatment is recommended for SAD by many of the major medical organizations, including the Primary Immunodeficiency Committee of the American Academy of Allergy, Asthma and Immunology, which states that IVIG is “probably beneficial” for the treatment of “primary immune defects with normogamma-globulinemia and impaired specific antibody production.”

The logistics and cost of IVIG therapy also lend themselves to controversy considering the high expense and potential shortages of this plasma product. “This is not an inconsequential issue,” says Dr. Harville. “Some people are trying to think of things in a global context of keeping the cost of medicine down.”

SAD Prognosis

The long-term prognosis for SAD can be different for children than for adults. For instance, it’s possible that some kids will simply outgrow it over time. But, it’s also possible that a diagnosis of SAD may progress into a more serious immune deficiency such as common variable immunodeficiency (CVID). The only way to monitor for progression is to periodically re-evaluate serum IgG, IgA and IgM levels, as well as specific antibody titer responses to the pneumococcal vaccine. IgA and IgM are not influenced by IG replacement and, if falling, can indicate the evolution toward CVID. When treated with IVIG, a fall in the serum level IgG trough values can also indicate a progression in disease. Unfortunately, when treated with subcutaneous IG, measurement of serum IgG levels may not be as useful, but a fall could be indicative of disease progression and/or lack of adequate replacement. If the serum levels of IgA or IgM are falling, there may not be a need to stop therapy for retesting. “Is it a transitional point toward normalcy or toward a full-blown CVID illness, or is it a prolonged state that doesn’t progress further?” asks Dr. Harville. “That’s a difficult question to answer. In most kids, it changes, and they will either outgrow it or it will turn into a full-blown CVID.”

According to Dr. Harville, the patients who are more likely to be evolving to CVID are those whose IgG levels become lower as they are retested over a period of seven to 10 years. On the other hand, it is more difficult to tell if patients are evolving to CVID if their IgG, IgA and IgM levels continue to remain normal at the same time that their pneumococcal responses are poor. “For an adult, the normal IgG level is 800 to 1,200 mg/dL,” explains Dr. Harville. “If someone has an IgA or IgG that is borderline and the pneumococcal titers are poor, that is an ominous sign that there is going to be more of an evolution of things occurring. Even though someone is in the ‘normal’ range, if they are below the mean, I think they may be on the cusp of having problems.”

There is also the possibility that patients if not treated early or properly will suffer recurrent or chronic infections of the ears, sinuses, bronchi and lungs, which can, possibly, result in permanent damage such as hearing loss or chronic lung disease and scarring. But with proper treatment to prevent infections and the development of impaired lung function, hearing loss or other organ systems, the outlook for SAD is good.6

IVIG is not a U.S. Food and Drug Administration-approved Therapy for SAD.

A Real Disease

SAD patients produce an adequate quantity of immunoglobulins, but those immunoglobulins are deficient in quality to fight infection. This, according to Dr. Harville, is where the controversy over this disease lies: “Some do not believe that if the serum immunoglobulin levels are normal that an immunodeficiency exists.” Yet, despite the controversy, it is a real disease. Without successful treatment with antibiotics or IG, these patients will continue to suffer with recurrent, serious infections that could lead to permanent disability. The good news is that there is greater awareness about SAD, and more patients are being properly diagnosed and treated.  

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

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Can They Boost the Immune System?

Primary immunodeficiency patients often turn to supplements to help boost their immune systems, but there is little proof that they have any beneficial effect, and they are not regulated by the U.S. Food and Drug Administration.

By Mindy Hermann, MBA, RDN

PRIMARY IMMUNODEFICIENCY (PI) patients are more susceptible to a wide range of infections and infectious symptoms. Some fall under the specter of contagious illness and include infections of the ear, sinus, lungs and skin. Others, such as bowel disorders, may or may not be infectious and often are accompanied by inflammation. Because finding effective medications to prevent and manage many of the problems experienced not just by PI patients but also by the general public can be challenging, manufacturers and marketers have stepped in with a wide array of supplements that claim to help treat, prevent and manage inflammation and boost immunity.

A visit to the supplement section of any of the dozens of online and brick-and-mortar drugstores can be overwhelming. A single search on one chain’s website for products with immune system benefits generated nearly 300 different nutrient and herbal supplements. For pneumonia alone, the site recommends alpha-linolenic acid, fish oil, flaxseed oil, green tea, iodine, lactobacillus, larch arabinogalactan, schisandra, vitamin A, vitamin C and zinc, while the list for bronchitis names choline, cowslip, English ivy, gaba (gamma-aminobutyric acid), ginseng, great plantain, n-acetyl cysteine, serrapeptase, sorrel, South African geranium, thyme and vitamin C.

Supplement labels and accompanying materials describe the product’s relationship to health, but most claims are neither scientifically supported nor approved by the U.S. Food and Drug Administration (FDA). Indeed, labels are required to state that their claims have not been evaluated by FDA and that products are not intended to diagnose, treat, cure or prevent any disease.

For the most part, supplements have not been shown to boost immunity. The immune system is quite complex, with many interdependent processes that are not yet fully understood. Not only are single supplements or supplement combinations unlikely
to positively impact the immune system, but some supplements taken in high doses have been associated with negative side effects, including increased rates of tumor growth.

The reality, however, is that people with chronic conditions often take supplements despite the lack of scientific proof. This overview summarizes research and other information on some of the most popular supplements, including vitamins and minerals, being marketed for boosting immunity.

Supplements Marketed to Boost Immunity

**Vitamin A** impacts T cells and B cells that play a role in the health of the mucosa, and vitamin A deficiency negatively affects the immune system. Supplement manufacturers recommend vitamin A for a wide range of skin and other disorders, including acne and eczema. But, it is unknown whether vitamin A supplements aid recovery from infections, enhance immunity or boost wound healing.

**Vitamin B2** claims regarding immune system enhancement are not supported by sufficient evidence in humans. The vitamin has been shown to increase resistance to certain infections in mice, however.

**Vitamin B6** deficiency is extremely rare. Like a deficiency of vitamin A, it suppresses certain immune responses involving T and B cells. Supplementation corrects the deficiency and relieves deficiency symptoms but does not enhance immunity or prevent infections in people who are not deficient.

**Vitamin C** is a popular remedy for preventing and treating the common cold. Some people also take vitamin C for bronchitis and bladder infections. Evidence suggests that vitamin C may help shorten the duration of the common cold but does not prevent it.

**Vitamin D** has been shown to stimulate the body’s response against the bacterium that causes tuberculosis. Vitamin D also may alleviate some of the skin symptoms associated with autoimmune diseases such as scleroderma and lupus, particularly when vitamin D supplements are taken to correct a disease-related deficiency of vitamin D in the body.

**Zinc** plays an important role in the health of the immune system, specifically related to proper functioning of T cells. While it is widely used to treat colds, prevent respiratory infections, boost
the immune system and treat certain skin conditions, evidence to support such uses is extremely limited. Zinc lozenges appear to shorten the length of a cold but do not prevent it. Additionally, zinc has not been shown to help treat the joint symptoms and inflammation of psoriatic arthritis or rheumatoid arthritis.

**Aloe vera** is more commonly used on the skin than as an oral supplement. Topical aloe vera appears to aid skin healing for minor conditions, but oral supplements have not been shown to have an effect on the immune system.

**Andrographis paniculata** is one of a group of Chinese herbs with suggested anti-inflammatory and anti-microbial properties. It has not been adequately studied in humans.

**Astragalus** is popular in Chinese medicine for supporting and boosting the immune system. Two of the more than 2,000 species, Astragalus membranaceus and Astragalus mongholicus, are most commonly used for such purposes as treating colds and upper-respiratory infections. High-quality human trials and scientific evidence for the benefits of astragalus are lacking.

**Black elderberry extract** made from the berries and flowers of the elder tree is a popular remedy for infections, colds and flu. A handful of studies support its ability to relieve symptoms of flu and, in combination with antibiotics, sinus infections, but additional evidence is still needed to confirm the benefits of this supplement.

**Echinacea purpurea** is one of nine known species of this popular plant that is native to the United States. It is heavily promoted as an immune stimulant that prevents and treats colds and upper-respiratory infections. Few studies have been well-designed, and results on the benefits of Echinacea against the common cold have been inconsistent. The National Institutes of Health (NIH) National Center for Complementary and Alternative Medicine (NCCAM), recently renamed the National Center for Complementary and Integrative Health (NCCIH), is studying the potential benefits of Echinacea in treating upper-respiratory infections and boosting immunity. Echinacea may cause allergic reactions in people who are allergic to ragweed.

**Garlic** enjoys a long history of use for boosting the immune system, fighting infections and preventing illness. While laboratory research supports the ability of garlic to suppress certain bacteria, viruses and fungi, these actions have not been proven in humans.

**Ginger root** is best known as a remedy for stomach upset and nausea. It also is taken to relieve joint and muscle pain, as well as symptoms of rheumatoid arthritis, but it has not yet been proven to be effective.

**Ginkgo biloba** is a tree whose leaf extract has been recommended for treating bronchitis and other respiratory illnesses. More evidence is needed on the effectiveness of the leaf extract and seeds, which can be toxic when consumed in large amounts.

**Ginseng** is generally categorized as American or Siberian/Chinese/Asian. American ginseng differs from the Asian varieties. It is promoted for immune stimulation and prevention and treatment of colds and flu. Evidence suggests that certain American ginseng products might reduce risk of getting the flu or colds and shorten their duration. Side effects can include diarrhea, nervousness and heart-related symptoms. Siberian ginseng supplements have been shown to improve cold symptoms when taken in a product that also contains the herb andrographis. NCCIH supports research to study ginseng and notes that additional large-scale studies are needed to prove its efficacy. Patients should discuss ginseng with their doctors before using it as a supplement since it may interact with certain prescription medications.

**Glycyrrhiza glabra** (licorice root) has been studied in combination with other herbs, and evidence is lacking on licorice root alone. A common folk remedy for bronchitis and sore throat, licorice root interacts with numerous medications and should be avoided or used only with extreme caution.

**Goldenseal**, and its active compound berberine, is a traditional folk medicine for treating colds and other respiratory tract infections, eye infections and stomach and intestinal disorders.
Goldenseal is sold as an extract, in capsules or combined with Echinacea for treating colds. Few studies support its effectiveness against any health conditions or its safety. Several of the potentially active compounds in goldenseal, including berberine, are poorly absorbed from the gastrointestinal tract when goldenseal is taken by mouth.

Goldenseal may interfere with the way that the liver processes a large number of drugs, speeding up or slowing the rate at which the liver breaks them down. This can change the effects and side effects of the particular medications. Medications that are activated in the liver include amitriptyline (Elavil), clarithromycin (Biaxin), clozapine (Clozaril), codeine, desipramine (Norpramin), donepezil (Aricept), fexofenadine (Allegra), fentanyl (Duragesic), flecainide (Tambocor), fluoxetine (Prozac), indinavir (Crixivan), irtraconazole (Sporanox), ketoconazole (Nizoral), lovastatin (Mevacor), meperidine (Demerol), methadone (Dolophine), metoprolol (Lopressor, Toprol XL), olanzapine (Zyprexa), ondansetron (Zofran), sildenafil (Viagra), tramadol (Ultram), trazodone (Desyrel) and others. PI patients who are considering taking goldenseal should talk first with their healthcare provider.

Mushrooms and fungi are thought to confer benefits to the immune system by interacting with bacteria and other microorganisms in the gut. In Asia, a parasitic fungus called cordyceps produces compounds that may stimulate the immune system and also prevent cancer. Further study is needed on the potential relationship between health status and mushrooms in general and certain fungi in particular.

Probiotics are beneficial bacteria cultures that establish a stronghold in the large intestine and benefit health by crowding out harmful bacteria, boosting immune function in the intestine and possibly strengthening the body’s immune system. In addition to common probiotic strains such as Lactobacillus and Bifidobacterium, probiotics supplements provide any number of lesser-known strains. Some supplements also contain inulin or other types of fiber that serve as food for the bacteria. These are called prebiotics. Research results on probiotics are promising but not yet conclusive.

Benefits of Supplements

Should PI patients take supplements? Supplements are not monitored or regulated by FDA, which means that the dose of the active compounds may be inaccurate or sometimes unknown. It also means that they may contain other substances not listed on the ingredients. In February, New York’s attorney general directed four major retailers — GNC, Target, Walmart and Walgreens — to stop selling some of their store-brand herbal supplements because they did not contain ingredients they claimed to and they contained ingredients not listed on their labels. Seventy-nine percent of the products tested had no DNA of the plants listed on the labels or were contaminated by other material, including rice, beans, pine and wheat. While instances like these are rare, there have been cases of people harmed by toxic or pharmacologic contamination of over-the-counter supplements. And, just like medication, supplements can cause hypersensitivity reactions. Furthermore, supplement-drug interactions are a very important issue. Lastly, there is tremendous financial cost often associated with these products, which have no proven benefit in nearly all cases.

Patients with PI should consult with their physician and check the excellent resources provided by NIH before adding supplements to their daily regimen. Nothing takes the place of a healthy lifestyle for supporting the immune system as best as possible. Positive actions include not smoking, eating a healthful and balanced diet, incorporating physical activity when possible, maintaining a healthy weight, getting enough sleep, managing other health conditions, practicing food and personal safety to avoid infections, and seeing a doctor regularly.

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Interviewing for a job isn’t always easy for candidates with a chronic illness. Here are some tips to help navigate the interview and keep the focus on what to bring to the position.

**RECENTLY, FAST COMPANY** asked Art Markman, professor of psychology and marketing at the University of Texas at Austin, if people with an invisible disability should disclose their illness before being hired for a job. His advice is that candidates should showcase their work and social skills and hold off on sharing information about their disability until after a job offer is made. “Anti-discrimination laws exist because there is a human tendency to discriminate in a number of ways. We often react negatively to people who are different from us and to things we don’t understand, often unconsciously,” he says. “Chronic illnesses can have both of these characteristics for potential employers, even those well-trained in what the law protects.”

But not everyone who has an invisible chronic illness chooses to withhold that information. In addition, chronic illness can take many forms, including both visible and invisible components. Those with rare diseases and autoimmune conditions treated
with immune globulin therapy illustrate this fact. These conditions are diverse — and growing more diverse all the time. Therefore, job interviewing strategies won’t necessarily be the same from person to person. Instead, they will depend on each person’s situation and comfort level. Other factors also need to be taken into consideration such as where candidates are interviewing and how open those companies are to recruiting and hiring people with chronic illnesses.

Following are nine tips that candidates can use to determine how to approach their own job search.

1. **Know one’s rights under the current disability legislation.**

   Susan Herrin, director of employment services at The Whole Person, a nonprofit organization offering a range of community-based services for people with disabilities, says the first thing candidates need to empower themselves is an understanding of what constitutes a disability. She points out that the Americans with Disabilities Act Amendments Act (ADAAA) of 2008 changed the landscape in this regard.

   “Under the ADA, disability was stated as interfering with daily activities of life, whereas under the ADAAA, the definition was broadened to include perceptions of others as being a person with a disability, as well as how the disability affects everyday life, so it’s really expanded on that definition quite a bit,” Herrin says. Also, having a history of a disability, even one that has been treated and no longer affects a person to the same degree every day, also constitutes a disability.

   If candidates do have a condition that meets the requirements for a disability, Herrin says the next step is to arm themselves with knowledge. They should learn their rights under the ADA, the ADAAA, and the Equal Employment Opportunity Commission (EEOC). They should go into interviews knowing what kinds of questions can and can’t be asked in relation to their condition. And, they should be aware that they don’t have to divulge anything about their condition if they don’t feel comfortable doing so.

2. **Learn how to approach potential employers about illness.**

   Herrin points out that attitude is critical when finessing the interview process. Many employers will have good intentions but won’t necessarily know how to accommodate those with visible or invisible disabilities. Rather than going into the situation feeling defensive or putting the onus entirely on the employer, candidates can help employers by having a good sense of what they need as an accommodation, both for the interview process itself and during employment. This might mean knowing not only what accommodation is needed, but also how the organization can put that support in place. Herrin says this might include educating the employer about resources available to them such as vocational rehabilitation services and tax credits designed to help companies support their employees with disabilities.

3. **Reframe illness as an asset.**

   Too often, as Markman mentions, candidates believe their disability will make them a victim of unconscious discrimination. But, that’s not necessarily the case. There are companies that actively pursue employees with disabilities, and there are even others that want to attract those candidates but aren’t quite sure how to go about doing so. Herrin advises candidates to find those employers by looking for signs that they want to be inclusive. For instance, these companies’ websites and marketing materials can be reviewed for language and images that speak to inclusion. Many of these companies have an EEOC or diversity specialist in their human resources departments. And, many of them take part in community job fairs that are inclusive of people who have disabilities. Organizations that serve people with disabilities can identify these organizations.

4. **Integrate illness into one’s professional marketing campaign.**

   Markman’s observation that candidates should focus on their work and social skills during the interview is accurate. This can be done whether or not a disability is divulged. The key is for candidates to know how to market themselves, and that can include marketing their illness. One way to do that is by focusing on soft skills, says Herrin. This includes the ability to get along with co-workers and customers. It can also mean being an ambassador for the employer as a person with the empathy needed to interact well with the diverse customer base the employer serves.

5. **Address health needs head on.**

   If candidates know at the outset that they will need to take a day off once a month for IG therapy, for example, Herrin says it’s best to be up front during the interview. “I would just be honest about it because it’s going to come up,” she says. Approaches include asking if there is any flexibility in the schedule or the potential to have coverage from another employee. Candidates don’t have to say why they need
the time off. Herrin points out that people need time off regularly during work hours for all sorts of reasons that have nothing to do with a disability. In an inclusive environment (one that accommodates all its employees — not just those with disabilities), she says, there will usually be a way to work out any scheduling issues.

6. Fill gaps in employment. When approaching an interview, a gap should never be left on a résumé. That time should be filled with something relevant. This might include volunteer work or some other form of community service. It could also mean stating that a personal situation created a need to be at home. Herrin has seen candidates be even more frank, stating: “I had an illness. I’m fully recovered now and ready to go to work.” This type of statement can be included in a cover letter or on a résumé. Leaving a gap leaves the recruiter wondering, which isn’t going to put candidates in a strong starting position.

7. Take advantage of vocational rehabilitation. Sometimes a chronic illness, especially one acquired in adulthood, can necessitate a career change because of new physical limitations, to preserve health or for other reasons. In this case, making use of vocational rehabilitation resources is the best route to take. “They can provide on-the-job training. They can help with college. They can help with job support, artificial limbs, transportation reimbursements, those kinds of things,” says Herrin. Vocational rehabilitation also works with other community organizations such as those that assist with job placement for people with disabilities to make sure people being served have the suite of services needed to not only enter the workforce but also to achieve success over the long term.

8. Consider part-time work. Sometimes people with chronic illness feel they can’t work because they can’t work full time. But, part-time work can be beneficial both for companies and people with chronic illness. Herrin urges people to consider part-time options, especially if they are entering the workplace for the first time or after an absence due to their condition.

9. Learn how to handle unexpected interview obstacles. Despite the best preparation, issues can still arise during the interview process. For example, the testing process might include elements that are hampered because of a disability. In cases like these, Herrin says candidates can explain that they didn’t realize a particular aspect of the interview process would come up, and they can ask for an accommodation at that time. Candidates can also follow up after the interview with the hiring supervisor and someone from the employer’s human resources team to address the issues that created a potential barrier to employment.

Just as there is no single chronic illness, there is no single way for candidates with a chronic illness to approach interviewing. Complicated, multifaceted illnesses require deep consideration with regard to how to approach job interviews. While the decision to reveal or withhold health information during the interview is personal, all candidates with chronic illness should enter the job search with an understanding of their rights and the tools needed to best negotiate each interview. This information, coupled with a positive attitude and a desire to support the employer’s efforts at supporting candidates, will lead to the most successful outcomes for everyone involved.

DANA MARTIN is a writer and editor in the Kansas City area who specializes in science, medicine and health.

Sources
2. In-person interview on Dec. 8, 2014, with Susan Herrin, director of employment services at The Whole Person, 3710 Main St., Kansas City, MO 64111.
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LET’S TALK

PROFILE:
Francesca Owens
By Trudie Mitschang

After years of illness and no diagnosis, Francesca Owens was finally diagnosed with myalgic encephalomyelitis and common variable immunodeficiency in 2007.

MANY PEOPLE FANTASIZE about moving to a remote island or village and “starting over.” For Francesca Owens, that fantasy became reality when she relocated to the tiny village of Spoleto, an Italian province in Umbria. The former stockbroker, athlete and internationally acclaimed artist had suffered years of declining health and misdiagnosis. Eventually, open heart surgery, followed by three strokes and a nervous breakdown, threatened to end her career and even her life. An eventual diagnosis of myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS) and common variable immunodeficiency (CVID) gave Francesca the knowledge and resolve to become her own healthcare advocate and get her life and career back. Today, she’s an advocate for the chronically ill with a mission to get effective yet out-of-reach drugs approved for use in the U.S.

Trudie: Tell us about your life before your diagnosis.

Francesca: Prior to becoming sick, I had my own investment consulting firm, I was serving in public office in Colorado, I worked as an environmental grant writer, I was an emerging international wildlife artist, and I was an athlete. I enjoyed backpacking around the world with my then-9-year-old daughter, Megan. But, at age 39, after my second pregnancy with my daughter Antonia, life as I knew it came to a screeching halt.

Trudie: When did you first suspect you were ill?

Francesca: After moving to Italy in 2007, my health worsened. I was convinced I was going crazy due to the noises, pinging and scratching sounds in my head. I finally flew to Colorado for a CereScan brain imaging scan and learned there was no flat line depression or frontal lobe damage as I’d been told previously, but there was a lot of inflammation. The scan led to my diagnosis of ME/CFS.

Trudie: What led to your first diagnosis?

Francesca: After moving to Italy in 2007, my health worsened. I was convinced I was going crazy due to the noises, pinging and scratching sounds in my head. I finally flew to Colorado for a CereScan brain imaging scan and learned there was no flat line depression or frontal lobe damage as I’d been told previously, but there was a lot of inflammation. The scan led to my diagnosis of ME/CFS.

Trudie: What was your initial prognosis?

Francesca: I contacted the internationally renowned ME/CFS expert Dr. Nancy Klimas at Nova University in Florida. Her consultation led to numerous other diagnoses of myofascial pain syndrome, postural orthostatic tachycardia and natural killer cell dysfunction, to name a few. As ME/CFS
is incurable without any treatment options, the future looked dim.

Trudie: How did things turn around?

Francesca: Research has shown that intravenous immune globulin (IVIG) is a promising treatment for ME/CFS, but it is not approved by the U.S. Food and Drug Administration (FDA) for that use. My health continued to deteriorate, and I eventually consulted Dr. Isaac Melamed of IMMUNOe Health Centers in Colorado, who diagnosed me with CVID. I was then prescribed IVIG treatment in Colorado, although it took longer to get the treatment approved in Italy. I receive IVIG every 21 days, and I take low-dose naltrexone and imunovir, B12 injections, prescription vitamin D and two cardio aspirin. In the ME/CFS world, we call this a drug cocktail mix because two of my prescriptions are not FDA-approved.

Trudie: What have you learned from this journey?

Francesca: Chronically ill patients need to play an active role in their treatment plan; being passive is not a good thing. My doctors in Florida and Colorado wrote letters pleading with the local hospital of Spoleto to continue my IVIG treatment there. After two years, my local village came through for me. I have been having regular IVIG treatments since last July, and I have witnessed the medical protocols in two different countries. I have to say that both do a great job.

Trudie: Tell us about your art.

Francesca: During the last seven years, I continued to make art without showing it. Year after year, I just painted, sculptured, collaged and kept going. The idea of leaving a legacy became my focus.

Francesca: How did you begin showing again?

Francesca: To my surprise, the community of Spoleto offered me a free space to show my work, and volunteers collected more than 55 pieces from my studio and hung them for me. My 13-year-old daughter Antonia walked visitors up and down the corridor explaining the significance of each painting and how it related to each medical crisis I suffered. It’s funny; I was a globally recognized wild tiger advocate using my paintbrush to support the World Wildlife Foundation. I could never have predicted that I would become an international patient advocate for ME/CFS and CVID.

Trudie: You incorporated IVIG into a sculpture. Tell us about that piece.

Francesca: I purchased a used IV pole and sneak out my used immune globulin bottles and some of my IV hoses to create it. Since I love prosecco and can only have it in small quantities, I re-created a pastel pink rose garden with ivy vines and my double-sized empty prosecco bottle for this sculpture. The prosecco bottle hangs upside down with all used IV tubes in the cork and flows down to fluted champagne glasses with Umbrian salami and cheeses draped over the IV pole, too. The Italians laugh at my sense of humor, and so do I.

Trudie: You seem very ambitious. What keeps you going?

Francesca: Besides making art and running an ME/CFS safe exercise group, I am writing two books. I fight for my health every day, and when I am not fighting for my health, I do the things I love every minute. My life with chronic illness is far more beautiful than 95 percent of all the other people I know. My goal is to travel the exhibit and raise chronic illness awareness. I’ve also started the online Chronic Illness Art Museum.

Trudie: What is your favorite quote and why?

Francesca: “Art is the funnel, as it were, through which spirit is poured into life.” I plan on making art into my late 70s, but should my time end sooner than expected, I will have left my mark. Until then, I won’t go down without a fight.

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.
**When You Make Plans, the Universe Laughs**

By Ever Fecske Mazza

JUST SAYING “WHEN you make plans, the universe laughs” puts a smirk on my face. Why? It’s true! Although I have a feeling I’m not laughing in the same way the universe is. Looking at where my life is today, I wonder if I will ever learn to stop making plans. I plan everything! I think about every little detail before it happens. I think about the different ways a situation could unfold and what will happen next. I find comfort in making plans; it’s something I look forward to — a goal to reach. Plans add a little certainty to an uncertain existence. Yet, why am I always so surprised when nothing ever goes as planned? Insanity? Very possibly — especially with all the drugs I’m on. Haha.

Making plans of any sort is almost laughable with a chronic illness. Long-term life plans, plans to take a trip, even lunch plans are difficult. So how can we manage life without plans? Do we just do the best we can to keep them? The inability to make plans takes living in the moment to a new extreme. In fact, the “moment” is all we really can be certain about. Here and now is all we really know we have. The problem is that when life is never on schedule, it becomes challenging to see the good that is right in front of you.

That’s why I’ve been practicing appreciating every moment — especially those moments when I feel good. Why is it that we only notice when we feel terrible? Yet, when there is a day without pain or congestion, we don’t think about it. I take those good days for granted, and it’s only when I am feeling miserable that I reminisce about the day I felt good. It isn’t easy, but the more I force myself to see the good in things, the easier it becomes and the less I think about my plans or my pain.

I have a tendency to get anxious and bogged down by the daunting task of managing my health. It’s overwhelming because as time goes on, I just seem to be collecting diagnoses. Things are inevitably getting more complicated, and sometimes it feels like my illness overpowers any plans I could ever make. That’s when I stop myself. I take a moment and look around. Sometimes I lay down on the grass to feel the ground beneath me and see the oak tree above. I look at the complexity of the branches and how they tangle together perfectly, and the blue sky and sun shining through; then, my mind is calm. In that moment, I have no plans but to appreciate the moment. It’s the simplest thing; the oak tree in my front yard has been there growing for hundreds of years, and just looking at it brings me back in touch with what is good.

What are some things that calm your mind? Think about some of the simple things you love, and then stop and appreciate them. I love a really good orange; I love how the peel makes my hands smell. After I finish eating the orange, I get to enjoy it all over again. I love how my hair looks and feels at the beach. It springs up to my ears from the salty sea air so I have the tightest curls. I love a cozy pair of oversized sweat pants, a soft blanket, vanilla candles, lemon trees, a red front door, watching it rain, the first bite of a glazed doughnut, baby kisses and a campfire. I could go on forever thinking about the simple things that bring me joy in a moment. It’s in these moments that we are laughing with the universe.

**EVER FECSEKE MAZZA** was diagnosed with CVID and interstitial lung disease in 2004. She is a new mom of a sweet little boy named Boston, and loves every minute of it! She lives in Los Angeles, Calif., with her husband, and when she isn’t changing diapers and playing with her son, she enjoys wedding planning, baking, flower arranging, cooking, shopping and anything that sparkles!
Modern Infusions?
By Ilana Jacqueline

A NEW VIRTUAL reality experience is coming to life for pediatric patients at the Children’s Hospital of California. Noise-silencing headphones and oversized screens are displaying Xbox games, action movies and even life-size Skype sessions with friends. It’s all part of the new “Infusionarium” created for children who must come to the hospital for long periods of infusion treatments. Great idea, right? While the trend of adventurous infusions hasn’t reached as far as my hometown in South Florida, I’ve found that many hospitals are incorporating modern facilities for patients who come in for their IV treatments.

I recently switched from getting my infusions at my doctor’s small office to my local hospital’s infusion center. I was nervous about the transition because I’d always had my infusions done in private, and frankly, when I’m in the middle of a treatment, I’m just not that talkative. In fact, I usually like to take that time to either work on my laptop or kick back and sleep.

But, then, I’d never actually been in an infusion center, so I had no idea how they were set up. I’ve had infusions on hospital gurneys and uncomfortable exam tables, even on the occasional loveseat. But an actual center dedicated to IV treatments? That was a new experience. Would I be in a private room inside of a larger hall? Would I be stuck in an uncomfortable chair for hours? Would they even have Internet access?

My anxieties were alleviated soon after I arrived for my first treatment. A friendly nurse set me up in one of the 10 large recliners in the room, each divided by a curtain.

“OK, here’s your heat button and your massage button,” she said.

“My what and my what?” I asked.

Turns out this infusion center had heated recliners with a massage (well, more like a weird vibrate) function. It was unexpected and so useful. How many times had I practically had to keep my teeth from chattering from cold IV fluid?

The nurse moved behind my chair and dropped down a small television in front of me.

“We don’t have all the channels, but most of your basic cable,” she explained.

So, basically, this was like modern treatment heaven. The hospital’s Internet was also lightning fast. I was grateful to be able to continue working from my laptop and found that it was much easier to concentrate when I wasn’t frozen, distracted by being in an uncomfortable position or struggling to constantly adjust myself to get comfortable.

When I got home later that day, I gushed to my friends: “I just had the best infusion! It was like going to a spa!”

Well, it wasn’t exactly their vision of a spa day. But for me it turned an otherwise monotonous and uncomfortable chore into a relaxing and cozy experience. My only regret was that I hadn’t looked into an infusion center earlier. For many patients with immune deficiencies, their treatments (when not done at home) may have to be done in a doctor’s office or hospital room. I’d urge patients to check out local infusion centers to see if their insurance will cover a move to a center more fitting to their needs.

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.
When I find myself engaged in conversation about immune disease and how it’s affected me, I ebb toward the relevant and the relatable. Lately, I’ve been sticking to a favorite: our family’s medical costs. I need a therapist and a financial advisor every time I go to the mailbox; explanations of benefits from my insurance company remind me constantly of the many dollars accruing due to our immune diseases. With two teenagers on subcutaneous immune globulin (SCIG) replacement and my tumor necrosis factor infusions, we are quite efficient in collecting collection companies.

“I don’t know how you guys do it!” is the comment I field most. It takes a great amount of self-control not to tell people the truth, the whole truth and nothing but the truth: Living with chronic illness isn’t for sissies. But how does one make light of something as serious as being on the cusp of medical bankruptcy? Let me share with you a few situations.

Expensive Spit

Our kids have often been described as the “healthiest looking sick people.” Though they appear normal on the outside, their broken immune systems on the inside are either clueless or AWOL (or both!) when foreign cells like bacteria come to town. A lesson in Immunology 101 explains that when an immune deficient patient receives IG replacement therapy, they receive a new immune system that knows how to fight bad germs so the patient doesn’t get as sick. But, this wonderful science comes at a hefty price, and our kids aren’t afraid to tell you or demonstrate how helpful their immune systems can be.

When Caleb was in kindergarten, the time he spent outdoors was the best part of his day. Despite needing to be wary of infection risks due to cuts and scrapes, Caleb chose to play fighter jet pilot and race car driver because they were the ones who received the most heroic road rash. The more gnarly and playground sand-infested your wound, the more respect you earned. One fateful day caught Caleb off guard. He was challenged with the choice of duty or good deed. He chose the latter, and the phone conversation went a little something like this:

“I don’t want you to be alarmed, Cheryl, but Caleb was involved in another accident today,” Mrs. Chydoll, Caleb’s kindergarten teacher, said.

“What did he do now?” I asked.

“He’s OK, we think. We just need for you to know he was trying to help a fellow classmate when she scraped her elbow on the stucco wall …” Mrs. Chydoll explained. And, then I thought I heard her say: “… with his tongue.”

“Could you repeat that? I didn’t quite catch what you just said,” I replied.

“Caleb was trying to help the girl out, something about her in his lane and trying to push her up against the rails. Some NASCAR thing. I don’t know. But he felt bad and wanted to make it all better so it doesn’t get infected. He licked her elbow and spit a couple of times on her scrape, saying something about having the most expensive and healthiest spit around,” said Mrs. Chydoll.

I was doing everything I could not to laugh, but it was impossible. Even Mrs. Chydoll broke after I explained to her that because of Caleb’s infusions, we’ve oftentimes teased him about having the most expensive and healthiest spit around.

“Maybe the next time I get a paper cut, I’ll ask Caleb to meet me at the water fountain,” Mrs. Chydoll joked.

Of course, we told Caleb to stop licking his classmates when he got home from
school that day, but praised him for doing what we thought was the gentlemanly thing by trying to clean the girl’s elbow.

“Yeah, but now I’m gonna have to really work hard getting my place back,” Caleb grumbled.

“Because you helped someone?” I asked.

“Not just because I helped ‘someone,’” he exclaimed. “I helped a ‘she-monster’! They’re the enemy!”

Our Refrigerator Is More Expensive Than Your Refrigerator

Molly and Caleb were just babies when they started to infuse IG in a hospital setting. As they grew older, we were able to switch to homecare.

Every four weeks, I have the privilege of ordering infusion supplies, including IG. When the supplies come to our door, we pack perishables in the fridge and other goods in their respective bins just like a good home pharmacy should. One particular delivery day, our firstborn, Calvin, bopped through the front door with a box of supplies in his arms.

“Mom! Supplies are here! And I have to do a silly math homework. Can you help me? What’s for snack?” Calvin bellowed with one breath.

Once we got supplies and snack covered, we jumped to silly math.

“We have to sort the food in our refrigerator into food groups, then estimate what costs the least and what costs the most,” Calvin explained. “Isn’t that silly?”

“Not unless you want a good grade!” I joked.

We got busy immediately so I could get on with making dinner, folding laundry, getting Caleb and Molly’s infusions ready and coming up with a plan for world peace.

“I’ve got everything sorted, but I’m having a little trouble with this new food group, Mom,” Calvin announced.

Sure enough, Calvin had pulled the IG out of the butter compartment and had lined the bottles up in a neat row. Being a supportive big brother, Calvin knew how precious the bottles of “liquid gold” were to Caleb and Molly. He was also quite resolved that IG was going to be included in his assignment because “if it’s in the refrigerator, it must be important in keeping us alive.” I attached a note to his teacher explaining why human plasma was 1) in my butter compartment and 2) why it is a “food” group.

When Calvin got home from school, he was beaming with excitement. “Mom! MOM!”

“What’s up?” I asked, fearing the worst. “We WON!”

“We won what?”

“We won the refrigerator separator silly assignment!” Calvin jumped.

“Whaaaa?”

Come to find out, the student who estimated the closest total cost of the items in his or her refrigerator earned (won in a fifth-grader’s eyes) the choice of sticker, Jolly Rancher and pencil.

“And, because mine was too much money to believe, we looked everything up on the Internet, and sure enough, I was right!” Calvin celebrated, performing his happy dance.

I looked at his assignment sheet and read his teacher’s note: “15 Grand! That’s some spendy, lifesaving butter!”

You Is Expensive

“You is kind. You is smart. You is important.” That’s my favorite line from the movie The Help. Words like these remind me to embrace something precious like a cheerful caress, celebrated memories, my kids’ IG.

The other day, I was feeling down in the dumps, achy and frustrated from news about a recent blood draw and my immune disease. I felt someone edging closer behind me. It was my Molly. She turned my shoulders so I was forced to meet her eye to eye (well, eye to chin since she is 2 inches taller than I am). Molly embraced me as gently as she could, enveloping me in her being. She caressed my hair as I took her sweetness in as deep as I could, and then she whispered: “You is kind. You is smart. You is important.”

I did everything I could to breathe in her words.

Then, I said with all I had in me: “You is expensive. You is worth it.”

CHERYL L. HAGGARD is a stay-at-home mom and has three children with PI, two of whom have CVID.
Choosing the Right Summer Camp for a Chronically Ill Child

By Jessica Leigh Johnson

SPRING IS HERE, the school year is winding down and summer is right around the corner. As usual, I’m more excited than my kids about summer vacation. As a stay-at-home mom, I look forward to three glorious months of spending all day every day with my four children. What could be better?

But if previous summer breaks are any indication of how this one will go, by the third day, I’ll have heard the words “I’m bored” one too many times from one too many children, and I’ll be ready to send them back to school — but I won’t be able to. Not until September.

Thank goodness for baseball, swimming lessons and 4-H. Anything to keep them busy. Of course, there’s also summer camp. Six days and five nights of nonstop, kid-oriented amusement — provided by someone else. Where do I sign them up?

Because three of my kids have a primary immunodeficiency, sending them off to summer camp for a week isn’t a minor decision. Many factors come into play. But there are ways to ensure parents like me choose the right camp that will meet their child’s physical, educational and entertainment needs, while providing Mom and Dad with the confidence that their child is safe and healthy while away from home.

Here are some things to consider when researching summer camps for a child with a chronic health condition:

Whether to attend a traditional camp or special-needs camp. What is the difference, and which type of camp would best meet the child’s needs? Traditional camps offer a wide variety of activities, opportunities for campers to experience new things and exposure to other campers and staff. But, there are some camps that offer limited activities geared toward special-needs campers’ abilities with trained and knowledgeable staff with expertise to understand these children’s varying challenges, and a supportive and fun atmosphere to share with others. One drawback is that special-needs camps do not exist in every state. A list of camps geared toward children with a number of health conditions and/or disabilities can be viewed at the Federation for Children with Special Needs website at www.fcsn.org/camps or in the article “Summer Camp Checklists” in the April-May 2014 edition of IG Living at www.IGLiving.com/magazine/archive.html.

Distance from home. How far away is too far for campers with chronic health issues? While there may be more varied experiences and opportunities...
farther from home (like swimming in an ocean or skiing in the mountains), staying somewhat local has its advantages, especially if a child becomes ill. The closer the camp is to home, the faster a child can get to an emergency room in a medical emergency and then follow up with the child’s regular doctor. Attending a local camp also makes it easier to check out the camp facilities ahead of time and meet with the staff and counselors.\textsuperscript{7}

\textit{Are staff members trained and experienced?} While most camp counselors are fun and enthusiastic teenagers working a summer job, well-trained adults might be more equipped and level-headed when dealing with a health crisis.\textsuperscript{7} When selecting a camp, be sure the staff members are able to make needed accommodations for a child with special health concerns and that they are willing to work with parents so that they feel comfortable with the arrangement. Determining the level of training and experience of camp staff can put parents’ minds at ease when leaving their child in the care of strangers.

\textit{What is the ratio of staff members to kids?} A child will receive more individual attention in a program with one adult for every five campers than in a program with one adult for every 15.\textsuperscript{5} For a child with a chronic illness, a closer adult-to-camper ratio will ensure changes in health don’t go unnoticed.

\textit{Are there medical professionals on-site?} For a child with a chronic illness, attending a camp with a nurse on-staff is a must. Parents should call the camp several weeks ahead of time and ask to talk to the nurse. And, they should find out how and when medications are dispensed. For example, if a child needs to take medication before bed, will the nurse still be there? If not, other arrangements must be made. If a child’s medication needs to be refrigerated, there should be a designated, safe storage area for it.\textsuperscript{5} Parents should be sure that several of the camp staff — not just the nurse — are trained to respond to medical emergencies such as seizures or severe allergic reactions to food or insect bites. Calling a child’s doctor several weeks ahead of time is also recommended to find out what therapies or procedures could be skipped or postponed for the length of camp, or what alternatives could be used (for example, using an acapella several times a day in place of compression vest treatments for those with chronic lung conditions).

\textit{How will parents communicate with staff members and their child during camp?} If a child will be attending a daytime-only program, parents may have opportunities to speak with counselors at drop-off and pick-up times. For overnight camp, parents need to ask if staff members will be readily available by phone or email.\textsuperscript{3} Also, it’s important to find out how often they will be able to speak with their child. Some camps have strict guidelines when it comes to campers contacting their parents. While this policy helps campers stay focused on their activities, it can be scary for parents and a child with chronic illness. It is important to figure out ahead of time how to get information about a child’s status. It’s OK to request that the camp director, nurse or a child’s counselor call with updates. Exceptions to the rules can usually be made for chronically ill children, provided there is a plan in place ahead of time.

\textit{What are the sleeping and eating arrangements?} If overnight camp is being considered and sharing a room with multiple campers poses a threat to a child’s well-being, ask if arrangements could be made to have a separate and possibly adjoining room so that the child can feel included, while not sharing sleeping quarters with a lot of other children. If a child requires a special diet, make sure the camp can provide it. If not, ask if any special foods/meals can be brought to camp with that child and prepared for him or her by the kitchen staff.

Depending upon the specific health needs of a particular child, an overnight camp may be out of the question. In a situation like this, parents can look to their local school district, YMCA or parks and recreation department, which are likely to offer many daytime-only programs for kids according to their specific interests and abilities. Sports camps, art camps and outdoor nature discovery camps are among the many programs offered to children in many communities across the country.

No matter what camp parents choose, the opportunity to spend a few days away from home with other children is an experience that a chronically ill child need not miss out on. Camp is a great opportunity to take a break from doctor appointments and therapy sessions, and a time to enjoy just being a kid. More likely than not, the week will go by without incident. If something should go wrong, having a plan in place for such an occurrence will eliminate confusion and anxiety for parents, staff and the child. The good news for parents is, if all goes well at summer camp, most camps offer winter programs as well! \textsuperscript{3}

\textbf{JESSICA JOHNSON} is a stay-at-home mom and mother of four kids, three of whom have X-linked agammaglobulinemia. She is a member of American Christian Fiction Writers and has written one book about the loss of her son to a primary immunodeficiency.

\textbf{References}

Selecting SCIG Pumps and Needle Sets

By Trudie Mitschang

Whether you are a primary immunodeficiency patient new to subcutaneous immune globulin (SCIG) infusions or have been managing your own treatment for years, the choice of products and supplies is an important aspect of your long-term treatment comfort and success.

Let’s Talk Needles

Any discussion of needles can be somewhat uncomfortable, but here are some basics to keep in mind. The gauge of a needle refers to its thickness or diameter. Ironically, the larger the number, the slimmer the bore of the needle. Numerous SCIG infusion sets are available, with needle sizes ranging from 24 gauge to 29 gauge, and 6-, 8-, 9-, 12-, 14- and 16-mm lengths. Because IG is a very viscous fluid, it is believed that a larger bore may decrease resistance and potentially decrease the infusion time.

Clarity on Catheters

Most needles used for SCIG are metal that are inserted and left in the subcutaneous space during the infusion. Another option is a soft Teflon catheter. The catheter is inserted using a metal needle; however, the metal needle is removed, leaving the soft catheter in place. Although these types of catheters are more commonly used for longer-term continuous infusions, they also can be used for SCIG therapy. When used for SCIG, the needles are available with an inserter, which allows the patient to insert the needle with the push of a button instead of manually. These, however, are available in a single line infusion set (not multifurcated).

Tubing and Pump Talk

Infusion sets come in a variety of options of furcation (number of sites). The type of tubing set selected will depend on the number of sites and the infusion pump that is used. Infusion sets are available with single, bifurcated, trifurcated, quadfurcated and five- and six-leg branches attached to a single trunk, which is connected to the syringe or other medication container. The infusion set tubing is connected to the syringe at the Luer lock end and is primed prior to insertion of the needle.

Different styles of infusion sets also vary in tubing diameter, which is important to consider when selecting the infusion pump. For instance, a mechanical pump may require larger bore tubing to decrease...
resistance of immune globulin, which is a high-viscosity drug, whereas a digital pump can use smaller bore tubing without changing the rate or resistance.

Some infusion pumps use an IV bag or “cassette,” which is filled with the SCIG product and connected to the pump. A number of different infusion pumps have been used for administration of SCIG. Most are syringe pumps that will accept a 50 mL or 60 mL syringe, but there is also a smaller version that accepts a 10 mL or 20 mL syringe. Some can also be programmed for different infusion rates.1

Dealing with Discomfort

Once the needle is placed during an infusion, the process should not be painful. If there is pain during the infusion, one reason may be that the catheter needs to be replaced. If the infusion sites are painful during SCIG, patients may need to check with their nurse or pharmacist regarding how to adjust the needle length or placement.

When in Doubt, Ask

While many factors go into choosing needles, ease of use and comfort during infusion are what will determine how successful your treatment will be. Ask your healthcare provider, infusion pharmacist or nurse how to select the right SCIG infusion set for your healthcare needs.  

Reference

SCIG Pumps

RMS FREEDOM60, Reusable Pump
The FREEDOM60 Syringe Infusion System from RMS Medical Products is a completely portable syringe infusion system, without the need for electricity or batteries, that operates at a constant, safe pressure. It is available with RMS’ Precision Flow Rate Tubing (that can also be used with other pumps), which features F-numbers that provide a different level of flow restriction for precise delivery of the medication, an emergency slide clamp on every set, a Leur disc connector to act as a barrier to contamination and an extension set available for longer length. The FREEDOM60 pump can be used for almost any subcutaneous or intravenous administration in a standard 60 mL syringe. (800) 624-9600, rmsmedicalproducts.com

Crono S-PID 50 Infusion Pump
The Crono S-PID 50 from IntraPump is the newest high-volume ambulatory infusion pump for controlled subcutaneous administration of prescribed medications. Crono S-PID 50 combines high technology with innovative design. It is small and lightweight, accurate and has the flexibility to change flow rates during an infusion with ease, which makes it ideal for home therapy and provides flexibility for the patient to administer medication any time of day without interruption to daily life or leisure activities. Crono S-PID 50 is used with a dedicated 50 mL syringe. There is also a Crono Super PID pump designed for pediatrics with dedicated 10 mL and 20 mL syringes. (866) 211-7867, intrapump.com

SCIG Needles

Soft-Glide Single- and Multi-Needle Sets
EMED Technologies offers its Soft-Glide single- and multi-needle Sub Q infusion sets in 24 and 27 gauges and a wide selection of needle lengths, including 4mm, 6mm, 9mm, 12mm, 14mm and 16mm, to accommodate all age groups and skin types. The sets feature soft translucent wings to facilitate placement and patient comfort, and they are available with Safety Wings to encapsulate the needle upon removal. The sets’ coating technology minimizes needle discomfort and anxiety, and its optimized needle contour maximizes flow. (888) 550-6500, www.emedtc.com

HlgH-Flo Needle Sets
RMS Medical Products’ HlgH-Flo Subcutaneous Safety Needle Sets feature a safety closure that covers the needle after use to reduce risk of needle stick injuries, 20-inch tubing for fewer tangles and lower residual volume, a backcut needle tip design for less tearing of the skin, and a flexible “flying-hinge” that bends to fit the body for less irritation and greater site comfort. They come in 24, 26 and 27 gauges in 4mm, 6mm, 9mm, 12mm and 14mm lengths. A Y-Connector is available for combining sets for up to eight sites or to enable infusing with two different needle lengths. (800) 624-9600, www.rmsmedicalproducts.com
Imagine no Tegaderm™

“ The Crono pump is easier and more life friendly than most other pumps. Since I am in college it is convenient to be able to put it in my pocket if have to be at class. Also, shorter infusion time and smaller needles.”

“Now I only use two sites as opposed to 4 sites and my infusions are down to one hour (they were two hours until recently)”

“I now have hope for a much more active life with my family.”

The Infusion Set

The neria™multi infusion set is designed for the subcutaneous delivery of immune globulin.

The combined benefits of a low profile needle with a pre-attached adhesive make for simple insertion technique and eliminates the need to apply Tegaderm™, providing a high level of user comfort. The small bore double layer tubing helps minimize drug wastage and prevents kinking. The 36-inch tubing length means less tubing tangles.

neria™multi has a standard luer lock connection and can work with any infusion pump using a standard luer lock reservoir.

neria™multi is available in 27G and 8mm, 10mm and 12mm needle lengths.

The Pump

The Crono pump is the smallest, most accurate pump on the market for the subcutaneous delivery of immune globulin.

The Crono SPID50 and the Crono Super PID pumps work with a proprietary syringe, the design intended to ensure the overall size and weight is small and discreet for the user to wear. “You can put it in the pocket of your blue jeans.”

The Crono pump administers in tiny pulses of medication for better absorption at the site. The pump is flexible, and has the ability to alter the flow rate, during the infusion without stopping or changing any tubing.

Crono syringes have a standard luer lock connection and can fit any subcutaneous infusion that has a standard luer lock.
Clearview-MS Multi-Site Needle Set
The Clearview-MS needle set from Norfolk Medical is a multiple site subcutaneous delivery system used specifically for SCIG infusion. They are available in 2-, 3-, 4- and 5-leg sets that allow for multiple subcutaneous infusion sites. Needles come in 24 gauge and 27 gauge in a variety of lengths, ranging from 4mm, 6mm, 9mm and 12mm.
(847) 674-7075, norfolkmedical.com

Neria Multi Infusion Needle Sets
Intrapump offers a variety of styles of neria and neria multi infusion sets, which are steel needle infusion sets with a variety of furcation options. Also offered are neria soft and neria soft90, both of which are soft cannula infusion sets. All are designed for subcutaneous drug delivery. The sets come in 27 and 29 guages in 6mm, 8mm and 10mm needle lengths, and they come with a pre-attached adhesive and sit flat against the skin, so there is no need for extra Tegaderm when using them.
(866) 211-7867, intrapump.com

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All needle sets are not created equal
Say Goodbye to Needle Pain - Ask Your Provider for Soft-Glide® Needle Sets!

EMED Soft-Glide® SCiG Safety Needle Infusion Sets

Safe
Proprietary and patented wing design requires only a simple squeeze on the wings after removal to ensure that the needle is enclosed completely and securely

Easy to Use
Soft-Glide® needle sets are easy to insert and remove, having soft, flexible wing designs that provide a comfortable grip for reliable insertion and removal

Comfortable
The Soft-Glide® proprietary and patented needle design and coating reduces penetration force and skin friction, resulting in a virtually painless insertion

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El Dorado Hills, CA 95762
www.emedtc.com
For a more comprehensive list of resources, visit the Resources page at IGLiving.com.

**Ataxia Telangiectasia (A-T)**
- WEBSITES
  - A-T Children’s Project: www.atcp.org

**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)**
- WEBSITES
  - GBSCIDP 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?id=11120be6FWE0
  - Kawasaki Disease Foundation: www.kdfoundation.org
  - KidsHealth: kidshealth.org/parent/medical/heart/kawasaki.html

**Mitochondrial Disease**
- WEBSITES
  - United Mitochondrial Disease Foundation: www.umdif.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.multi-sclerosis.org/index.html
  - Multiple Sclerosis Association of America: www.msaa.com
  - 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: www.myositis.org
  - International Myositis Assessment and Clinical Studies Group: www.niehs.nih.gov/research/resources/collab/imacs/main.cfm
- ONLINE PEER SUPPORT
  - The Cure JM Foundation: www.curejm.com
  - Michigan Immunodeficiency Foundation: www.facebook.com/groups/108404062584350
  - Myositis Association Community Forum: tmacomunityforum ning.com
  - Myositis Support Group: www.myositisupportgroup.org
  - Myositis Support Group – UK: www.myositis.org.uk

**Pedicatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)**
- WEBSITES
  - P.A.N.D.A.S. Network: pandasnetwork.org
  - Midwest PANS/PANDAS Support Group: www.giveforkids.com

**Peripheral Neuropathy (PN)**
- WEBSITES
  - Neuropathy Action Foundation: www.neuropathyaction.org
  - The Neuropathy Association: www.neuropathy.org
  - Texas Chapter of the Neuropathy Association: www.handsfeethert.org

**Primary Immune Deficiency Disease (PI)**
- WEBSITES
  - Immune Deficiency Foundation: www.primaryimmune.org
  - Jeffrey Modell Foundation: www.info4pi.org
  - American Academy of Allergy, Asthma & Immunology: www.aaaai.org
  - International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: www.ioppi.org
  - New England Primary Immunodeficiency Network: www.nepin.org
  - Rainbow Allergy-Immunology: www.uhospitals.org/immunology/allergy-immunology
- ONLINE PEER SUPPORT
  - IDF Common Ground: www.idfcommonground.org
  - IDF Discussion Forum: idffriends.org/forum
  - IDF Friends: idffriends.org
  - Jeffrey Modell Foundation Message Board: www.info4pi.org
  - Michigan Immunodeficiency Foundation: www.facebook.com/groups/108048062584350
  - Immune Deficiency Foundation: www.immune-df.org

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

**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
  - 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.livingwithsp.com
  - Stiff Person Syndrome: www.stiffpersonssyndrome.net

**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
  - 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.livingwithsp.com
  - Stiff Person Syndrome: www.stiffpersonssyndrome.net
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Connect with other patients and caregivers living with PI. Your Patient Advocate will also be your guide to all things MyIgSource.

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Access educational books, magazines, emails, and more about managing your PI journey.
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