Chronic Lymphocytic Leukemia: A Rare Cancer

Diagnosing and Treating CIDP

Parenting: Preparing PIDD Kids for High School

Controlling IVIG Side Effects

PIDD Children
Understanding Kids’ Needs
GAMUNEX®-C
Immune Globulin Injection (Human) 10% Caprylate/Chromatography Purified

HIGHLIGHTS OF PRESCRIBING INFORMATION
These highlights do not include all the information needed to use GAMUNEX®-C safely and effectively. See full prescribing information for GAMUNEX-C.

GAMUNEX-C, [Immune Globulin Injection (Human) 10% Caprylate/Chromatography Purified]
Initial U.S. Approval: 2003

WARNING: ACUTE RENAL DYSFUNCTION and FAILURE
See full prescribing information for complete boxed warning.

- Renal dysfunction, acute renal failure, osmotic nephrosis, and death may occur with immune globulin intravenous (IGIV) products in predisposed patients.
- Renal dysfunction and acute renal failure occur more commonly in patients receiving IGIV products containing sucrose. GAMUNEX-C does not contain sucrose.
- For patients at risk of renal dysfunction or failure, administer GAMUNEX-C at the minimum concentration available and the minimum infusion rate practicable.

INDICATIONS AND USAGE
GAMUNEX-C is an immune globulin injection (human) 10% liquid indicated for treatment of:
- Primary Humoral Immunodeficiency (PI)
- Idiopathic Thrombocytopenic Purpura (ITP)
- Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

CONTRAINDICATIONS
Anaphylactic or severe systemic reactions to human immunoglobulin
- IgA deficient patients with antibodies against IgA and a history of hypersensitivity

WARNINGS AND PRECAUTIONS
- IgA deficient patients with antibodies against IgA are at greater risk of developing severe hypersensitivity and anaphylactic reactions. Have epinephrine available immediately to treat any acute severe hypersensitivity reactions.
- Monitor renal function, including blood urea nitrogen, serum creatinine, and urine output in patients at risk of developing acute renal failure.
- GAMUNEX-C is not approved for subcutaneous use in ITP patients. Due to a potential risk of hematoma formation, do not administer GAMUNEX-C subcutaneously in patients with ITP.
- Hyperproteinemia, with resultant changes in serum viscosity and electrolyte imbalances may occur in patients receiving IGIV therapy.

ADVERSE REACTIONS
- Thrombotic events have occurred in patients receiving IGIV therapy. Monitor patients with known risk factors for thrombotic events; consider baseline assessment of blood viscosity for those at risk of hyperviscosity.
- Aseptic Meningitis Syndrome (AMS) has been reported with GAMUNEX-C and other IGIV treatments, especially with high doses or rapid infusion.
- Hemolytic anemia can develop subsequent to IGIV therapy due to enhanced RBC sequestration. Monitor patients for hemolysis and hemolytic anemia.
- Monitor patients for pulmonary adverse reactions (transfusion-related acute lung injury [TRALI]).
- Volume overload
- GAMUNEX-C is made from human plasma and may contain infectious agents, e.g., viruses and, theoretically, the Creutzfeldt-Jakob disease agent.
- Passive transfer of antibodies may confound serologic testing.

DRUG INTERACTIONS
- The passive transfer of antibodies may transiently interfere with the response to live viral vaccines, such as measles, mumps and rubella. Passive transfer of antibodies may confound serologic testing.

USE IN SPECIFIC POPULATIONS
- Pregnancy: no human or animal data. Use only if clearly needed.
- Geriatric: In patients over 65 years of age do not exceed the recommended dose, and infuse GAMUNEX-C at the minimum infusion rate practicable.

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Important Safety Information for GAMUNEX-C

Gamunex-C, Immune Globulin Injection (Human), 10% Caprylate/Chromatography Purified, is indicated for the treatment of primary humoral immunodeficiency disease (PI), idiopathic thrombocytopenic purpura (ITP), and chronic inflammatory demyelinating polyneuropathy (CIDP).

Renal dysfunction, acute renal failure, osmotic nephrosis, and death may occur with immune globulin intravenous (IGIV) products in predisposed patients. Patients predisposed to renal dysfunction include those with any degree of pre-existing renal insufficiency, diabetes mellitus, age greater than 65, volume depletion, sepsis, paraproteinemia, or patients receiving known nephrotoxic drugs. Renal dysfunction and acute renal failure occur more commonly in patients receiving IGIV products containing sucrose. Gamunex-C does not contain sucrose. For patients at risk of renal dysfunction or failure, administer Gamunex-C at the minimum concentration available and the minimum infusion rate practicable.

Gamunex-C is contraindicated in individuals with acute severe hypersensitivity reactions to Immune Globulin (Human). It is contraindicated in IgA deficient patients with antibodies against IgA and history of hypersensitivity.

Gamunex-C is not approved for subcutaneous use in patients with ITP or CIDP. Due to the potential risk of hematoma formation, Gamunex-C should not be administered subcutaneously in patients with ITP.

Hyperproteinemia, increased serum viscosity, and hyponatremia may occur in patients receiving IGIV therapy.

Thrombotic events have been reported in association with IGIV. Patients at risk for thrombotic events may include those with a history of atherosclerosis, multiple cardiovascular risk factors, advanced age, impaired cardiac output, coagulation disorders, prolonged periods of immobilization and/or known or suspected hyperviscosity.

There have been reports of noncardiogenic pulmonary edema [Transfusion-Related Lung Injury (TRALI)], hemolytic anemia, and aseptic meningitis in patients administered with IGIV.

The high dose regimen (1g/kg x 1-2 days) is not recommended for individuals with expanded fluid volumes or where fluid volume may be a concern.

Gamunex-C is made from human plasma. Because this product is made from human plasma, it may carry a risk of transmitting infectious agents, e.g., viruses, and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.

After infusion of IgG, the transitory rise of the various passively transferred antibodies in the patient’s blood may yield positive serological testing results, with the potential for misleading interpretation.

In clinical studies, the most common adverse reactions with Gamunex-C were headache, fever, chills, hypertension, rash, nausea, and asthenia (in CIDP); headache, cough, injection site reaction, nausea, pharyngitis, and urticaria with intravenous use (in PI) and infusion site reactions, headache, fatigue, arthralgia and pyrexia with subcutaneous use (in PI); and headache, vomiting, fever, nausea, back pain, and rash (in ITP).

The most serious adverse reactions in clinical studies were pulmonary embolism (PE) in one subject with a history of PE (in CIDP), an exacerbation of autoimmune pure red cell aplasia in one subject (in PI), and myocarditis in one subject that occurred 50 days post-study drug infusion and was not considered drug related (in ITP).

*CIDP=Chronic inflammatory demyelinating polyneuropathy; PI=Primary immunodeficiency; ITP=Idiopathic thrombocytopenic purpura.


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.

Please see adjacent page for brief summary of GAMUNEX-C full Prescribing Information.
About IG Living
IG Living is the only magazine dedicated to bringing comprehensive healthcare information, immune globulin information, community and reimbursement news, and resources for successful living directly to immune globulin consumers and their healthcare providers.

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**MICHELLE GREER, RN**  
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**GIL WOLFE, MD**  
Professor and Chairman, Department of Neurology, University at Buffalo School of Medicine and Biomedical Sciences, SUNY

#### Diagnosing and Treating CIDP

“Although CIDP is rare and difficult to diagnose, once it is accurately diagnosed, there are treatment options.”

**ANNABEN KAZEMI**  
IG Living’s Patient Advocate

#### The Benefits of SCIG

“Many studies have documented the safety and effectiveness of SCIG therapy, and they validate that SCIG is a therapeutic equivalent to IVIG.”

**LESLEY TRIPP, RN, BSN**  
Registered Infusion Nurse

#### Controlling Side Effects of IVIG Infusions

“As an infusion nurse, I aim to provide a safe delivery of the prescribed therapy so my patients will receive the most benefits with the fewest side effects.”

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

#### Diagnosing Specific Antibody Deficiency: The Effects of the Conjugated-Pneumococcal Vaccine, Part 3

“The sticky question that arises from the definition of antibody deficiency is: Does one only consider the 10 non-conjugated for anti-polysaccharide antibody production or all 23?”

**AMY SCANLIN, MS**  
Freelance Writer

#### Treating PIDD Kids — Post-Diagnosis

“While PIDD was once thought to be a rare disease, because only the most severe forms were more easily recognizable, milder forms of PIDD are now recognized as being more common than previously thought, and also more readily treatable.”

**JIM TRAGESER**  
Freelance Writer

#### Understanding Chronic Lymphocytic Leukemia

“At present, medical researchers do not know what causes most cases of CLL.”

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### Connect with Other *IG Living* Readers through Monthly Teleforums!

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

- Email *IG Living* to be added to our email invitation list for the teleforums.
- *IG Living* will send you invitations to let you know when the monthly, hosted, toll-free teleforums will be held, as well as what topic relevant to the IG community will be discussed.
- The moderated, hour-long calls will be filled on a first-come, first-served basis and will be limited to 15 readers.

In addition to connecting with others, *IG Living*’s patient advocate can help you determine if there’s a patient organization support group in your area. Or, she can help you to start an IGL Readers Group of your own. To join a group or start one in your area, visit [www.IGLiving.com](http://www.IGLiving.com) and click on IGL Readers Groups.

Sign up for the Teleforums now by emailing akazemi@IGLiving.com or calling (800) 843-7477, ext. 1366.
Our mission is to support the IG community through education, communication and advocacy.

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Editorial

PIDDD Kids: The Challenge of Living a Normal Childhood

Historically, individuals diagnosed with a primary immune deficiency disease (PIDDD) died early in life. Today, those with PIDDD live mostly healthy, productive lives. Yet, despite how far medicine has come, enabling quicker diagnoses and improved treatments, living with PIDDD still has its challenges, especially for kids.

Karrie St. Clair knows this all too well. In our Transitions column interview, Karrie describes how her daughter, Katie, was sick since birth. Yet, it took until age 13 to get a diagnosis of common variable immune deficiency. Karrie’s quest to find a diagnosis by being a “persistent advocate” paid off, and today at age 22, Katie is living a fuller life and has just had her first child.

Katie’s story is not unusual. Our article Treating PIDDD Kids — Post-Diagnosis explores the ways in which physicians are diagnosing both mild and severe cases of PIDDD earlier based on antibody responses to protein and polysaccharide vaccines’ antigens. It also looks at treatment options to decrease the number of infections PIDDD kids experience.

But, even more importantly, this article explores the financial, environmental and emotional impacts of living with PIDDD — for both children and their families. As Dr. Terry Harville explains, PIDDD is an expensive disease to treat (costing $5,000 to $10,000 a month), which can place an extreme burden on families. But there are assistance programs available. Parents need to be especially cognizant of PIDDD children’s surroundings to ensure they are not exposed to elements that will cause a flare-up of their condition. And, they play a crucial role in helping their children maintain a positive and healthy outlook, despite their illness.

This means getting their children the psychological counseling they need, getting them involved in social support groups and taking a proactive part in their medical care.

PIDDD teens preparing to enter high school face even greater challenges. Our article Preparing Immune-Deficient Kids for High School by Mark Haggard, who is a high school coach and father of PIDDD kids, looks at the many different issues these teens face. He also provides some strategies that both teens and their parents can use to help mitigate those challenges. For teens, these strategies include familiarizing themselves with campus officials prior to the start of school, making friends, getting involved in activities and not letting their disease define them. Parents can help by communicating with teachers and the school nurses, being an advocate for their kids’ academic success and preparing their kids for the adult world.

Of course, this issue also includes articles on a host of other topics related to diseases treated by immune globulin (IG), including chronic inflammatory demyelinating polyneuropathy and chronic lymphocytic leukemia. It also explores topics related to IG treatment, including easing IG side effects and the benefits of subcutaneous IG infusion.

But, I hope the parents of PIDDD children and PIDDD kids themselves will find this issue especially helpful and will embrace some of the advice offered.

Ronale Tucker Rhodes, MS, Editor
Helping the Patient Community

I feel compelled to let you know how much I have thoroughly enjoyed IG Living over the years. It is an excellent, top-notch publication, and I cannot even begin to articulate how important it is for the patient community, or thank you enough for all of your efforts on behalf of patients. Excellence is never an accident; it is the result of high intention, sincere effort, intelligent direction, skillful execution and the vision to see obstacles as opportunities. No other distributor, manufacturer or stakeholder has anything that can compare. To stand apart from the competition, you must first stand together as a team, and I just wanted to applaud you for your efforts.

Oftentimes, when I get down or depressed during one of my monthly infusions, I will read IG Living and I recall that our lives are not determined by what happens to us, but by how we react to what happens; not by what life brings to us, but by the attitude we bring to life. A positive attitude causes a chain reaction of positive thoughts, events and outcomes. It is a catalyst — a spark that creates extraordinary results. Sometimes this is just the spark I need to continue my patient advocacy efforts.

Again, thanks for informing, educating and, most importantly, motivating me. I truly appreciate all each of you does for patients and the community!

— Dominick V. Spatafora, president Neuropathy Action Foundation

Great Facebook Post!

Working for a major healthcare provider, I see more than just a poor patient experience but [also] the costs associated with it. [Following] is a good example … of what … “noncompliance” [of medical treatment] has resulted in. It’s no wonder healthcare costs are out of control. Direct consequences and costs of noncompliance: Inadequate implementation of treatment has devastating consequences, such as resulting in 10 percent to 25 percent of hospital and nursing home admissions; 340 deaths per day; 20 percent of unintentional pregnancies in the U.S. at a cost of $2.6 billion; three times as many doctor visits; $2,000 per year in additional costs compared to patients who follow their treatment plan; and 33 percent to 69 percent of all medication-related hospital admissions in the U.S. at a cost of $100 billion.

It is especially revealing that estimates of the total annual healthcare costs in the U.S. resulting from patient noncompliance vary from $100 billion to $170 billion to $300 billion. First, this range (even after adjusting for the 11-year difference between the oldest and newest figures) points to the potential risk of false precision, the dramatic influence of assumptions and methodologies in such approximations, and the difficulty of computing cost-benefit ratios of efforts to enhance compliance. Second, even the most conservative figures delineate the tremendous fiscal impact of noncompliance, fully justifying the American Heart Association’s summation that “the cost of noncompliance in terms of human life and money is shocking.”

— Marty Schick

The Editor replies:

Thanks for some interesting statistics, Marty. Our readers should know that your email was in response to an IG Living Facebook post made by our staff on January 19, which read: “When a patient does not follow a doctor’s treatment plan or discontinues medication sooner than advised, doctors refer to this behavior as ‘noncompliance.’ Statistics show noncompliance can result in higher healthcare costs, increased hospital admissions and a breakdown of doctor/patient communication. Have you ever been noncompliant, and if so, why?” Clearly, as your statistics reveal, noncompliance is very costly!

— Lianne Learnard Massachusetts

Your feedback, opinions, suggestions and anything else you’d like to share are important! Email us at editor@IGLiving.com or visit www.IGLiving.com and go to the Be Heard/Feedback page to send your comments.
THE IMMUNE SYSTEM is designed to specifically and directly recognize foreign proteins such as the diphtheria toxoid (DT) protein in the DT vaccine. Through this direct recognition process, signals can be generated that activate B lymphocytes, which are capable of making anti-DT antibodies. Polysaccharides (carbohydrate or sugar polymers) are commonly found on the surfaces of microorganisms, and are important for immune recognition and targeting of the microorganism. These become the target of antibodies, in part due to a bystander effect: The immune system is responding to a protein from the microorganism, and nearby B lymphocytes capable of making antibodies against a polysaccharide from the same microorganism get stimulated in the mix of lymphocytes in this close proximity. Due to the inherent inefficiency, it may take some time, occasionally years, before adequate anti-polysaccharide antibodies for protection from infection can be made. And, it is known that production of anti-polysaccharide antibodies is critical for the overall prevention and control of bacterial infections; this is one reason why infants and children are so susceptible to streptococcal ear infections.

Streptococcal organisms, also known as pneumococcus, cause many of the severe infections, especially in younger children and in those with antibody deficiencies. To help deal with this, vaccines were developed that attach (i.e., conjugate) the polysaccharide of importance to the DT protein. This results in the immune system undergoing the expected normal response to the DT protein, but it “tricks” the immune system into responding to the polysaccharide with formation of anti-polysaccharide antibodies. These so-called conjugated vaccines are Prevnar (pneumococcal 7-serotype conjugate vaccine to diphtheria CRM197 protein) and the newer Prevnar13 (pneumococcal 13-serotype conjugate vaccine to diphtheria CRM197 protein).

Here is where a dilemma occurs. If anti-polysaccharide antibodies can be detected after immunization with Prevnar13, is this the result of the inherent ability of the patient to respond to polysaccharide antigens, or is this solely an immunologic trickery effect from response with the DT protein?

To make the diagnosis of an antibody deficiency, it is common to perform a pre-/post-immunization study. Response to the non-conjugated pneumococcal vaccine (e.g., Pneumovax23) is thought to provide the most relevant information regarding making the diagnosis of an antibody deficiency. To perform the assay, blood is obtained (for pre-immunization antibody measurement), the vaccine is given, and after four weeks blood is again obtained (for post-immunization antibody measurement). The change in the pre- to post-immunization antibody levels indicates the responsiveness of the immune system. Currently, most immunologists believe that children up to approximately 6 years of age should have half of the anti-pneumococcal antibody levels in the protective range (>1.3 μg/mL) in the post-immunization specimen, and that the antibody levels should have increased more than three times, except for values that were already very high. For those older than 6 years of age, there should be a similar response in 70 percent of the antibodies tested.

Many available assays from commercial laboratories check for 12 to 14 of the 23 specific pneumococcal serotype antibodies from the non-conjugated vaccine. Before the introduction of the Prevnar with seven serotypes, this was fine, since there would be 12 to 14 non-conjugated serotypes to evaluate. After the 7-valent Prevnar, there could still be five to seven serotypes to evaluate. And, many immunologists divided the results in the 1) conjugated and 2) non-conjugated anti-polysaccharide antibody responses. Patients with normal antibody immunity would respond well in both categories. Younger children with otherwise normal immunity may have a somewhat better response in the conjugated category. Patients with an evolving antibody deficiency, that is, one that has not completely presented with all the disease features, may have a reasonable response to the conjugated polysaccharide antigens but poorer responses with the non-conjugated ones. And, patients with a more completely defined or severe antibody deficiency may have poor antibody responses to both conjugated and non-conjugated antigens. Therefore, the predicament is that younger children and those with evolving antibody deficiency may have relatively normal responses with conjugated vaccine.

Now, Prevnar13 is being used. This is wonderful for the
prevention of infections, but it results in more quandaries when considering the evaluation of patients suspected of having antibody deficiencies. If only 12 to 14 pneumococcal serotypes are evaluated, the person with evolving antibody deficiency who has previously received Prevnar13 may seem to have a reasonably good response, since most of the antigens present in Prevnar13 are present in the assay. Therefore, one is not truly assaying the non-conjugated responsiveness, yet the non-conjugated anti-polysaccharide antibody response is likely a better indicator of immune “normalcy” or, alternatively, antibody deficiency. The general solution to this dilemma is to assay for all 23 pneumococcal serotypes from the non-conjugated pneumococcal vaccine. There will then be the possibility of antibodies to 13 serotypes, which may have produced as a result of Prevnar13 vaccination, and antibodies to 10 non-conjugated serotypes. Then a better assessment of the situation can be made. The sticky question that arises from the definition of antibody deficiency is: Does one consider only the 10 non-conjugated for anti-polysaccharide antibody production or all 23? Since the goal is to determine the response to non-conjugated polysaccharide antigens, it makes the most sense to consider the 10 non-conjugated serotypes separately. Yet, if the 13 conjugated antigens produce poor responses, this is suggestive of a more severe antibody deficiency.

We will continue this discussion in the next issue.

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

Editor’s Note: This Immunology 101 column, introduced in the April-May 2010 issue of IG Living, is intended to be a basic course in immunology.

Imagine caring for your child with hemophilia with no factor, refrigerator, running water, electricity, or transportation to a clinic. This is the reality for thousands of families in developing countries. For just $20 a month, you can help an impoverished child with hemophilia. Become a sponsor today!

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Caring for people with hemophilia around the world—one at a time.
WHEN MY PATIENTS arrive for an intravenous immune globulin (IVIG) infusion, the first question I ask is: “How did you feel after your last infusion?” Several patients tell me they either didn’t experience any side effects or they felt a little tired for the next couple of days. These patients have the amazing benefits of IVIG without the sometimes debilitating side effects. Others, however, aren’t so lucky. As an infusion nurse, I aim to provide a safe delivery of the prescribed therapy so my patients will receive the most benefits with the fewest side effects.

Side Effects and Adverse Reactions

Immune globulin (IG) is derived from the human plasma of thousands of donors and includes essential antibodies. Some of the common side effects of IG include headache, migraine, fever, fatigue, itching, rash/hives, cough, chills/shaking chills, dizziness, nausea/vomiting, faster heart rate, upper abdominal pain and increased blood pressure. In comparison, the much more serious adverse reactions include renal failure, anaphylactic shock, blood clots, aseptic meningitis (especially with large doses or rapid infusion) and volume overload.

The most common side effects I observe in my patients are headache, flu-like symptoms, increased blood pressure and, occasionally, decreased blood pressure. Fortunately, many side effects can be controlled with careful monitoring of vital signs, as well as personalizing the infusion rate for each patient. For example, I have one patient who can tolerate the infusion at 200 ccs per hour and another who can tolerate only 30 ccs per hour.

For those patients who experience a post-infusion headache, I remind them to stay hydrated. Hydration is so important during IVIG infusions that I advise them to start drinking the day before and continue drinking during and after the infusion. Oftentimes, staying hydrated can make a significant difference in reducing the post-infusion headache. For other patients who experience severe headaches that require medication stronger than the ordered pre-meds, we discuss the importance of decreasing the rate and increasing titration times.

During the Infusion

Patients can expect to have their baseline blood pressure checked at the beginning of the infusion, before each titration and then hourly. They need to understand the importance of informing the nurse immediately if they experience any discomfort, including headaches. As soon as I know patients are beginning to get a headache, I assess the situation and determine the best course of action. Every patient is different. Some might
say the headache isn’t too troublesome and they will drink more water. Depending on the circumstances, I might decrease the rate of infusion while they hydrate and, once the headache begins to decrease and if the vital signs allow, I will increase the rate again.

Most doctors will order medications for patients to take prior to the infusion (pre-meds). Some examples of pre-meds are acetaminophen (Tylenol), diphenhydramine (Benadryl) and/or a steroid, to name a few. Patients should plan to take the oral pre-meds approximately 30 minutes prior to the infusion, and if a steroid is ordered, patients need to follow the instructions provided by the doctor. Sometimes an injectable steroid is given by the nurse prior to the infusion. Pre-meds are crucial as they help to avoid allergic reactions.

During their initial infusions, patients should plan on having someone drive them to and from the infusion center because Benadryl tends to make people sleepy and may make the patients too tired to drive home safely.

After the Infusion

Pre-meds often can be repeated after infusing, according to the physician order. I like to remind my patients who experience side effects such as headaches to continue hydrating, as well as to take acetaminophen and diphenhydramine again before going to bed the night of the infusion. Some patients are prescribed a steroid that can often decrease or eliminate post-infusion headaches that have been described to me as similar to a migraine. If patients continually suffer with severe headaches despite adjusting the infusion rate, hydration status and repeating pre-meds, I suggest they discuss this with their provider to explore other options.

Tips for Success

If patients fall asleep during the infusion (Benadryl often has that effect) and they are afraid the nurse will turn up the infusion rate, they should bring a friend or family member who will speak up for them. It’s important that patients know the rate they can tolerate. Over time, patients may have multiple nurses who may not be familiar with their case and who will infuse patients at the pharmacist’s recommendation, which may be too fast for some patients. If this happens, patients who know their rate can notify the nurse right away if they need the infusion to run slower than the recommended rate.

Patients also should try not to schedule anything after their infusion because it may run longer than usual due to side effects. I will not speed up the infusion because patients have somewhere to go afterward. Finally, be prepared for a long day. Even if the infusion usually takes only three hours, it may run six hours due to side effects, so patients should bring things to entertain themselves, as well as food and drink.

Be Proactive

To be proactive in their care, patients need to know their rights and their options, and they shouldn’t be afraid to speak up to their doctors or nurses. Remember: Patients are important members of the healthcare team. Still, many patients mention in blogs and conversations that the infusion nurse turns up their rate of infusion despite being told that it causes side effects. If that’s the case, they need to speak up! Call the infusion center, report the situation and request that nurse not be sent again. Asking to speak with a supervisor may be necessary to ensure the request is honored.

As an infusion nurse, I aim to provide a safe delivery of the prescribed therapy so my patients will receive the most benefits with the fewest side effects.

Clearly, being proactive is important because patient health, safety and comfort are at stake. When the infusion is done, the nurse has completed his or her duty, but patients might be stuck with uncomfortable side effects for the next couple of days or weeks. Therefore, communication is key. Patients need to voice their concerns so they will reap the benefits of IVIG without suffering the side effects.

LESLIE TRIPP, RN, BSN, is a registered infusion nurse in both home and clinic settings, and a mother and stepmother of five children.
Research

Privigen May Improve Function in CIDP Patients

New study results suggest that treatment with Privigen [immune globulin intravenous (Human), 10% Liquid], an intravenous immunoglobulin (IVIG), may lead to improvement in function in patients with chronic inflammatory demyelinating polyneuropathy (CIDP).

The Privigen Impact on Mobility and Autonomy (PRIMA) trial at the Peripheral Nerve Society Inflammatory Neuropathy Consortium Meeting in Rotterdam, Netherlands, a prospective, multicenter, open-label, single-arm study, investigated the efficacy and safety of Privigen in previously IVIG-treated and untreated patients with CIDP. The study achieved its primary efficacy endpoint, which was the percentage of patients responding — as measured by the Inflammatory Neuropathy Cause and Treatment (INCAT) scale — at study completion compared to baseline. The overall response rate was 60.7 percent. The 25-week treatment period permitted the observation that a response to IVIG can occur late (e.g., after more than six weeks of therapy).

The INCAT scale is used to measure a patient’s ability to perform tasks (i.e., walking, motor hand tasks, etc.). On this scale, patient scores rise with increasing weakness and disability, whereas improvement in basic motor functions is indicated by a reduction in the score. Results from this study showed that the mean overall INCAT score significantly improved from 3.7 at baseline to 2.3 at completion of treatment. Half of the responders achieved the clinically meaningful threshold by week four. This finding may encourage some treating physicians to continue IVIG therapy longer in their CIDP patients before assessing whether or not the therapy is working.

“CIDP is a rare, progressive disease that may cause permanent nerve damage, and studies show that current treatment options may not work for all patients,” said Jean-Marc Leger, MD, Hospital de la Salpetriere. “Finding new treatment options to slow the advancement of the disease is extremely important. The results from this study are promising as they suggest that Privigen may help decrease weakness and loss of motor function in people with CIDP.”

Technology

CSL Launches Hizentra Dosing Calculator App

CSL Behring UK Ltd. has launched the Hizentra Dosing Calculator App. Designed for use on iPhone and Android handsets, the app assists healthcare professionals with dosage calculation when administering Hizentra (human normal immunoglobulin, SCIG). It will calculate the correct volume of Hizentra to be infused based upon entry of only two key pieces of information: a patient’s weight and the weekly dosage required.

According to Eddie Owens, general manager, CSL Behring UK Ltd., “We see that healthcare professionals are clearly using mobile technology to access health information, and for us to engage with this growing target audience, we need to be present in this medium. The new Hizentra Dosing Calculator App aims to meet the demand that healthcare professionals have to access sound medical data from anywhere and at anytime.”

The app can be downloaded and installed for free directly from the phone, by going online to the iTunes store at http://itunes.apple.com/gb/app/hizentra-dosing-calculator/id483147404, or by visiting Google Play at https://play.google.com/store/apps/details?id=com.nitrogen.hizentra dose#?t=W251bGwsMSwxLDIxMiwY29tLm5pdHVlZ2VuLmhpemVudHJhZG9zZSJs.
**Medicines**

**Octapharma Initiative Expands Availability of Octagam 5%**

Octapharma USA has started an initiative to make Octagam (immune globulin intravenous [human] 5%), a therapy for primary immune deficiency, widely available to covered entities in the 340B Drug Pricing Program, which is managed by the Health Resources and Services Administration (HRSA) Office of Pharmacy Affairs (OPA). The 340B Drug Pricing Program is available to certain hospitals, clinics and outpatient treatment facilities that qualify as “covered entities” under Public Law 102-585, the Veterans Health Care Act of 1992, which is codified as Section 340B of the Public Health Service Act. More than 17,000 covered entity sites participate in the 340B Program, including six categories of hospitals that are generally considered safety net providers, and 11 categories of non-hospital covered entities, such as federally qualified health centers and specialized clinics and treatment centers.

“Octapharma is committed to providing therapies to treat life-threatening conditions to all patients, including those who are treated in facilities that have historically faced challenges accessing IGIV,” said Octapharma USA President Flemming Nielsen. “We are pleased that the supply of Octagam 5% is now sufficient to adequately serve 340B covered entities that have in recent years experienced difficulties in accessing specialty drugs such as IGIV at 340B discount prices. Octapharma is committed to serving patient needs, regardless of where they receive treatment, and ensuring a steady supply of Octagam 5% to all our hospital customers.”

Octapharma USA intends to use FFF Enterprises of Temecula, Calif., and ASD Healthcare of Frisco, Texas, as the contact point for distribution. More distributors will be added later in the year. Octapharma USA, a subsidiary of Octapharma AG, one of the world’s largest human protein product manufacturers, has been marketing octagam 5% since 2004.

**Education**

**Immunoglobulin Nursing Society Holds First National Conference**

The Immunoglobulin Nursing Society (IgNS) held its first national conference in Orlando, Fla., August 3 through 5. The conference is a new educational initiative to develop and sustain the advancement of knowledge, education and practice of nursing in the field of immunoglobulin therapy. It featured in-depth educational sessions, hands-on practical workshops, industry-sponsored symposia and an exhibit hall featuring the top IG-related companies in the country.

Educational sessions included the latest developments in the field of IG therapy, including clinical indications, administration and adverse event prevention and management, updates on the IG clinical trial landscape and the role of nurse study coordinators, as well as an update on issues with IG reimbursement and patient advocacy. Several smaller group practical workshops were taught by leading IG nurse experts focusing on best practices in intravenous IG (IVIG) and subcutaneous IG (SCIG) administration.

“The IgNS National Conference represents a major shift in the current education and training of IG nurses who treat patients with chronic and life-threatening disorders,” said Jane Kirmse, RN, MSN, ARPN-BC, president of IgNS. “The National Conference will provide nurses with an exceptional educational program to expand their knowledge, advance their skills and experience professional growth.”
Research

**Slow Release of IG May Delay Alzheimer’s Disease**

Recent studies conducted by Dr. Giulio Maria Pasinetti, Saunders Family chair and professor in neurology and psychiatry at Mount Sinai School of Medicine in New York, suggests that the divergent outcomes in Alzheimer’s disease clinical studies of intravenous immune globulin (IVIG) may be due to differences in temporal administration and administered dosages.

Dr. Pasinetti and his team of investigators recently found that prolonged administration of human immunoglobulin in models of Alzheimer’s disease, using a dose of immunoglobulin approximately five- to 20-fold less than equivalent doses used in Alzheimer’s disease patients, is effective at attenuating Alzheimer’s disease-type cognitive dysfunction while promoting synaptic plasticity. “This experimental observation provides a rational basis for rectifying the inconsistency of study outcomes in Alzheimer’s disease clinical trials with IVIG,” said Dr. Pasinetti. “We now have the much-needed information supporting the potential application of slow release of immunoglobulins delivered subcutaneously to delay the onset of Alzheimer’s disease, even at pre-symptomatic stages of the disease.”

Dr. Pasinetti hypothesizes that the slow release of immunoglobulins into the circulation and eventually into the brain for a protracted period of time may delay Alzheimer’s disease dementia onset and eventually its progression through epigenetic changes in the downstream gene expression of C5a-mediated pCREB-C/EBP signaling components associated with modulation of synaptic plasticity and eventually learning and memory functions.

Research

**BRI and Novo Nordisk Collaborate on Autoimmune Disease Research**

Novo Nordisk and Benaroya Research Institute at Virginia Mason (BRI), Seattle, Wash., have entered into a three-year collaborative agreement to potentially speed up translational research of the diagnosis and treatment of rheumatoid arthritis, inflammatory bowel disease and lupus. The agreement establishes how Novo Nordisk and BRI research scientists and clinicians will collaboratively develop studies to better understand changes in the immune systems of patients living with these autoimmune diseases. The intent is to develop better therapies and improve how these treatments are used.

“Translational research” describes a research approach that seeks to move discoveries made in laboratory, clinical or population studies more quickly into clinical care. In this specific agreement, BRI scientists and Novo Nordisk researchers at the company’s Seattle research center will work together to study samples and data registered in BRI’s biobank of patients with these diseases, as well as people with no history of autoimmune disorders. The personal information of these patients will not be disclosed.

“Improving patient care through innovation is at the heart of our company culture, and this agreement represents one way that we can work together with the larger health care research community to achieve this objective,” said Per Falk, senior vice president, Biopharmaceuticals Research Unit, Novo Nordisk. “We’re pleased to be working closely with the Seattle scientific community, which is sharing its best and brightest with us in an effort to bring new medicines for patients.”

In the United States alone, as many as 1.5 million people suffer from rheumatoid arthritis or inflammatory bowel disease, and more than half a million people suffer from lupus.

People and Places

**Coronado Biosciences Inc.**, a biopharmaceutical company focused on the development of novel immunotherapy agents for the treatment of autoimmune diseases and cancer, has appointed Karin Hehenberger, MD, PhD, formerly senior vice president of scientific affairs, to executive vice president and chief medical officer.
Research

Plasmapheresis May Reduce LDL and Increase HDL Cholesterol

A recent study conducted by Grifols suggests that the plasmapheresis process may reduce levels of low-density lipoprotein (LDL), or “bad” cholesterol, as well as total cholesterol in individuals who have high baseline levels. The study also suggests that plasmapheresis could increase levels of high-density lipoprotein (HDL) or “good” cholesterol, among individuals with low baseline levels. Plasmapheresis is a technique used to separate plasma from the remaining blood components, which are then immediately injected back into the donor at the time of the donation. Plasma obtained during plasmapheresis is used to produce lifesaving medicines for patients who have rare, genetic and life-threatening diseases.

The multicenter longitudinal study was conducted in nine plasma donor centers in the U.S., with blood analyses performed prior to plasmapheresis procedures to measure initial levels of total cholesterol, HDL and LDL. Plasma was collected from first-time donors or from donors who had not donated plasma for at least six months. The researchers estimated from the study results that plasmapheresis could reduce the levels of LDL by more than 30 mg/dl among individuals with high levels (greater than 160 mg/dl) or higher than desirable levels (greater than 130 mg/dl) when plasmapheresis procedures are performed two to four days apart. This effect was more significant in women, in whom cholesterol could be reduced by up to 35 mg/dl. A similar reduction pattern is estimated to occur in individuals with high total cholesterol levels (greater than 240 mg/dl) or higher than desirable levels (greater than 200 mg/dl), with the reductions in these cases potentially reaching 45 mg/dl and 32 mg/dl, respectively.

However, the cholesterol-lowering effects of plasmapheresis appeared to last only as long as the procedure continued at regular intervals, with cholesterol levels gradually returning to baseline following long periods without plasmapheresis. The same pattern of reductions was seen, although to a lesser degree, when subsequent plasmapheresis procedures were performed more than 10 days apart. Among individuals with normal baseline cholesterol levels, the study results suggested that plasmapheresis would not cause significant changes.

Research

Grifols Launches New IVIG Alzheimer’s Trial

Grifols has launched a new study into methods of treatment for Alzheimer’s disease. Known as the AMBAR (Alzheimer management by amyloid removal) study, it will investigate combined treatment using albumin plasmapheresis and intravenous immunoglobulin (IVIG) at different doses. The researchers will attempt to find synergies between the two treatments in order to reduce the frequency and volume of plasmapheresis, ultimately making the treatment experience more pleasant for patients and easier for medical professionals to administer.

AMBAR is expected to last two years and will be directed by Dr. Merce Boada, clinical head of the neurology service at the Vall d’Hebron Hospital in Barcelona. According to Dr. Boada, the study “opens up new prospects and hopes in dealing with an illness where success involves maintaining the quality of life of these patients.”
Research

Shingles Vaccine May Be Safe for Autoimmune Disease Patients

A recent study conducted at the University of Alabama at Birmingham shows that the shingles vaccine appears to be safe and effective for those suffering from autoimmune diseases. In the study, data were collected on more than 460,000 Medicare patients who had one of several rheumatic or immune-mediated diseases. Of those, more than 18,600 patients with rheumatoid arthritis, psoriasis, psoriatic arthritis, ankylosing spondylitis (an inflammation of the spine) or inflammatory bowel disease received the shingles vaccine. In the 42 days following vaccination, there were no cases of shingles, including among the more than 600 who were taking so-called anti-tumor necrosis factor biologics to treat their other conditions. And, there was only one case of shingles seen among all the patients during that time. More than 42 days after being vaccinated, 138 patients did develop shingles, which is in the range of the effectiveness of the vaccine. After two years of follow-up, the investigators concluded that the vaccine reduces the risk of shingles in these patients. That conclusion also was based on accounting for the type of immune disease, treatment and the use of arthritis drugs and steroids.

Because the shingles vaccine is a live vaccine, the U.S. Food and Drug Administration and other organizations say the vaccine should not be used in patients taking immunosuppressive drugs including all biologic agents and some nonbiologics because these patients may develop shingles from the vaccine virus strain. "A live attenuated vaccine reduces [shingles] risk by 70 percent and 51 percent among immunocompetent individuals 50 to 59 years and 60 years and older in two randomized, blinded trials, respectively," the researchers wrote. And, “the risk of [shingles] is elevated by 1.5 to two times in patients with rheumatic and immune-mediated diseases such as rheumatoid arthritis and Crohn’s disease. This increase has been attributed to both the underlying disease process and treatments for these conditions.”

According to Dr. Bruce Hirsch, an attending physician in infectious diseases at North Shore University Hospital in Manhasset, N.Y., who was not involved in the study, “The findings are reassuring for a very specific group of patients.” However, the study does not address the vaccine in patients who have weakened immune systems related to other causes, Hirsch said. And, he cautions that the vaccine does have some risks and there is no long-term data on its effectiveness in these patients. “I don’t consider this study to be completely definitive,” Hirsch said. “The book isn’t closed, but I am cautiously optimistic. The vaccine seems to be safe and these kinds of patients are able to handle the vaccine and get a benefit from it.”

The study was published on July 4 in the Journal of the American Medical Association.

Diseases

JMF Opens Diagnostic Center for PIDDs

The Jeffrey Modell Foundation (JMF), in partnership with CSL Behring, has designated the Midwest Immunology Clinic in Plymouth, Minn., as a Jeffrey Modell Diagnostic Center for Primary Immunodeficiencies (PIDDs). The center is headed by Ralph Shapiro, MD, and is devoted to the diagnosis, treatment and care of people who have PIDDs and autoimmune disorders. “We know that as many as 500,000 cases of PI remain undiagnosed in the United States,” said Fred Modell, co-founder of the JMF. “Our goal is to give every child a chance to lead a healthy, normal life. We are thrilled to form this partnership with Midwest Immunology, Dr. Ralph Shapiro and CSL Behring.”
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Most children with PIDD can lead relatively normal lives, but it’s necessary for parents as caretakers to understand their child’s treatments and psychological needs, as well as the financial considerations and environmental concerns, to best manage their child’s disease.

By Amy Scanlin, MS

For parents, a child’s diagnosis of a primary immune deficiency disease (PIDD) is a scary and, often, confusing time. What were frustrating repeated occurrences of ear infections, sinus infections, bronchitis or pneumonia now is a diagnosis that sounds far more frightening than was ever thought possible. And while the diagnosis can be a relief and bring apparent closure to the question: “What is going on
with my child?” — especially for those kids who also have health problems of the digestive tract, anemia or even autoimmune diseases as a result of immune system dysfunction — there also is the recognition that a whole new world of issues is arising.

Kids can take heart that with a prompt diagnosis and proper treatment, they very often will lead normal lives of play, sports and, yes, even school. Parents can take heart that while PIDD was once thought to be a rare disease, because only the most severe forms were more easily recognizable, milder forms of PIDD are now recognized as being more common than previously thought, and also more readily treatable.

**PIDD Diagnosis and Treatment**

Typically symptoms of a severe PIDD are noticed when a child is still in infancy through his or her first few years of life. In some cases, though, diagnosis does not occur until a child is a teen.

To make a diagnosis, typically a protein vaccine (tetanus and diphtheria toxoids) and a polysaccharide vaccine (pneumococcal) are administered to determine a child’s ability to make specific antibodies in response to the vaccines’ antigens. The pre-immunization response (blood taken before the immunization) is tested and compared with the post-immunization response (blood typically obtained about four weeks after the vaccination). This response allows doctors to better understand the severity of the disease and how best to treat it. In a child 6 years of age and younger, more than half of the vaccine components tested should demonstrate good antibody responses, and for a child older than 6 years of age, 75 percent of the vaccine components tested should result in good antibody responses.

Based on the results, a diagnosis of some form of antibody deficiency may be made, and subsequent treatment therapies can be determined. “We need to stratify the extent of the problem and make a decision as to how aggressive therapy needs to be based on the history, physical examination and laboratory test results,” says Dr. Terry Harville, medical director at the Departments of Pathology and Laboratory Sciences and Pediatrics at the University of Arkansas for Medical Sciences.

If recurrent respiratory infections are the main problem and no specific antibody deficiency can be ascertained or only a few of the vaccine components fail to respond adequately, then treatment with antibiotics may be appropriate. If there is an inadequate antibody response in the testing, therapy with immunoglobulin (IG) replacement may be needed, and antibiotics may also be used. IG replacement therapy may also need to be started if there is a failure to achieve an antibody response, or if the doctor determines it necessary based on his or her expertise when considering the severity or types of infections. For an inadequate or nonexistent antibody response, there should not be a trial to see whether antibiotics alone can slow down or prevent illnesses.

As a group, antibody deficiencies represent the most common type of PIDD.

Sometimes, certain antibodies are missing, even though the majority of antibodies may be fine. This situation may be called a specific antibody deficiency. This is typically one of the most difficult to diagnose, since the testing may not demonstrate a severe deficiency in antibody production. And, it can be one of the most frustrating to treat because IG replacement therapy may be required, but a patient’s insurance carrier may not want to pay for the therapy since a more severe antibody deficiency cannot be demonstrated.

As a group, antibody deficiencies represent the most common type of PIDD. Millions of antibodies, called immunoglobulins (IgGs), circulate in the blood, helping the immune system to fight all kinds of infections. By definition, antibody deficiencies require replacement of the missing antibodies with IG infusions, either by the intravenous route (IVIG) or the subcutaneous route (SCIG), in order to prevent the complications associated with recurrent infections and, most importantly, to prevent death. IgGs are obtained by “batching” them from the plasma of thousands of donors. The plasma is processed to isolate the antibodies and to remove viruses or other infectious agents. When infused regularly (every three to four weeks for IVIG therapy, or up to several times per week for SCIG therapy), the infusions can help to bring IgG levels to normal or near-normal levels for a child’s age to prevent infections. It is now recognized that maintaining the serum IgG levels well into the normal range helps to prevent many complications associated with infections.
Whether treatment is administered in a hospital or at home, follow-up doctor visits help a child to be re-evaluated, and they allow the doctor to learn of any infections or illnesses a child may have had since his or her last visit. They also are a good opportunity for the doctor to determine whether a child’s treatment dosage is still correct for his or her growing body. The frequency of follow-up visits is determined as part of the initial treatment plan, and it is continually adjusted as needed. A child also may need to be seen for other issues in addition to his or her PIDD. Asthma, allergies, arthritis, celiac disease or other things keeping him or her ill may need to be identified, evaluated and treated.

If breakthrough infections occur, antibiotic treatment also may be required. Frequently, the dose of antibiotics may need to be higher and the duration longer. For example, a five-day course would constitute undertreatment. Further, a 10-day course may need to be expanded to 14 days, and if sinusitis is present, one to three months of continuous antibiotics may be required. Despite being on adequate dosing of replacement IG, if frequent breakthrough infections are occurring or chronic sinus or chronic lung infections are present, then continuous dosing with antibiotics may be necessary. Although in the past, low-dose antibiotics may have been used to act as “prophylaxis” against infections, it is now recognized that patients do better with full treatment dosing on an ongoing basis.

There are situations in which IG replacement therapy may not be sufficient. For instance, if there is a problem with the function of neutrophils, such as with chronic granulomatous disease, it may be necessary to use daily antibiotics, typically trimethoprim/sulfamethoxazole to help reduce or prevent bacterial infections, itraconazole or voriconazole to help reduce or prevent fungal infections, and gamma interferon injections to maintain appropriate health. When the issue is a more severe problem with T lymphocyte production or function that also affects the ability of B lymphocytes to function appropriately for making normal antibodies (which would typically be diagnosed in infancy), a child may require hematopoietic stem cell transplantation (HSCT). That child will initially receive any antibiotics needed and be started on IG replacement therapy as soon as possible. “We also have to formulate a plan to protect these kids from viruses and other infections,” says Harville. A child may need isolation, especially from other children who may be infected with viruses, until the immune system has been reconstituted via HSCT.

HSCT is used for reconstitution of immunity in the most severe immunodeficiencies, as well as for blood disorders and cancer therapy. Hematopoietic stem cells, when working properly, produce billions of new blood and immune cells daily. Bone marrow is historically the most common source for HSCTs. However, today, frequently the source of stem cells for transplantation is from the peripheral blood. Donors no longer are required to undergo anesthesia and have needles poked through their bone to obtain bone marrow. Now, donors sit for a few hours, typically reading, watching movies or sleeping, while their blood exits through a catheter from a vein into a machine that removes the stem cells, and then is returned to their bodies. This allows for minimal discomfort and maximal yields of stem cells, since testing can be performed to determine how much of the donors’ blood needs to be processed in order to achieve the needed amount of stem cells.
**Financial Considerations**

“This is not a cheap disease,” says Dr. Harville. IG replacement therapy can cost up to $5,000 to $10,000 a month, and still other costs can be considerable depending on the other issues affecting a patient’s health and well-being. For example, missed work to take an ill child to be evaluated can place a strain on employment, and a child may be missing school frequently.

A financial assessment arranged through a social worker at the clinic or hospital can assist the family in paying for gas, accommodations and meal vouchers if the family must travel a long distance for treatment. Some clinics and hospitals have a “Medicaid van” that will pick up a patient who lives a distance from the clinic and return him or her at the end of the day, if reliable transportation is not otherwise available for that patient. There are many plans in place to help a family in need, and getting a social worker involved early is crucial.

For many, once the stress of the financial implications have been thought through and planned for, a family is better able to focus positively on the medical treatments ahead.

**Environmental Concerns**

Even if treatment for a PIDD has begun, a child’s environment cannot be overlooked. “Parents must be careful of environmental issues that may produce adverse outcomes,” says Dr. Harville. “All the therapy provided won’t help a child if an underlying issue exacerbating the illness is still present in their environment.”

Smoking and mold are two very common irritants that can promote more severe respiratory disease, despite IG replacement therapy and antibiotic usage. Mold behind walls, under sinks, in an attic or crawl space, places that may be otherwise unseen, can cause major problems for a child, even if others in the home have fewer respiratory symptoms. In addition, even being around a smoker who may not be smoking at the time can have an adverse impact. The residual smoke on the smoker’s clothing and in his or her hair can be sufficient to create problems, especially for a baby being held by the smoker.

Parents should take notice of when a child experiences symptoms and flare-ups to help find clues as to what environmental agents may be causing them. As an example, if a child gets sick every time he or she is in the home or car of a smoker, the answer is that child should no longer be in those places. It’s not good enough that the person isn’t smoking at the time.

**Psychological Healing**

A positive relationship is widely recognized between mental health counseling and the presentation of chronic illness symptoms in adults, including improvements in immune function, as well as a decrease in the effects of stress symptoms. It is suggested that the same can be true for a child, particularly in the areas of relaxation and hypnosis. These interventions are already used successfully in situations such as pediatric anxiety.

In Dr. Harville’s experience, often a main stressor for a PIDD child is the parent under stress and the resultant child’s perception of the parent’s stress. “A child who picks up on the parent’s worry doesn’t do as well,” he says. On their own, “kids don’t tend to worry as much, because their situation may be normal for them.” A child may get upset that he or she can’t do all the things his or her friends can do on the playground, but for a child, that state of health is his or her “normalcy.”

If a child starts sympathizing with the parent’s worry, it should be determined whether he or she has acquiesced to the issue rather than coped with it. Some children are more intuitive than others and may, in turn, have more psychological problems, but more typically, a child will worry more about whether he or she is going to get that new computer game or the latest MP3 player than whether his or her lungs are deteriorating.

Joining a local support group and organizations that provide learning and support resources such as the Immune Deficiency Foundation can be a critical part of the emotional journey toward healing. According to Dr. Harville: “All the assurance I can give during a visit isn’t the same as another parent sharing their story.” In the process of a support group, each parent also finds comfort in helping others. Volunteers are often able to sit with families while they wait, provide emotional support, a hand to hold and guidance.
Parents’ Roles as Helper, Healer, Caretaker

When the diagnosis of a PIDD is made, parents immediately take on the roles of coordinator, observer and secretary, in addition to caregiver. When a child has a more complicated disease or other complications, the parents’ task of having to potentially coordinate visits to multiple specialists — with each determining what he or she believes is the right course of treatment — while carefully observing how a child is responding to the treatments can be more than a full-time job. Yet, parents also must be careful note-takers and communicators to ensure that the multiple specialists are each informed of each other’s care approaches to treatment, and how each of their parts in the overall care is impacting a child. This, then, can become essentially a second full-time job for many mass complications are present.

Play often helps parents to see what is on a child’s mind.

Parents are encouraged to take notes, including times of day, location and symptoms, which can help doctors to know how a child is doing and what they can do better to help a child in his or her healing. As an added benefit, this also helps parents feel they play a productive and proactive part in a child’s medical care.

As a child ages, understanding of his or her condition typically changes. A young child may view the many doctor visits as punishment because he or she doesn’t understand the disease and the need for treatment. That child may only see that he or she is missing out on what other children can do. It is important for parents to never use the threat of “the doctor will give you a shot” as a means to try to control a child’s behavior. This creates anxiety in a child who frequently has previously undergone “shots” and blood draws, and who will likely require many more in the future. While it is easy to say that parents must help a child to understand that these visits and sometimes treatments are not punishment, the question becomes how to do this when a child has such limited understanding of the situation and the parents cannot reward each doctor’s visit.

Play often helps parents to see what is on a child’s mind. Whether children are drawing or playing, parents are often able to gently guide them to talk about what they are thinking and feeling. It is a child’s great ability for imagination that makes him or her such a good candidate for relaxation and hypnosis interventions for stress management. Helping a child channel this imagination in a positive direction with the help of his or her doctor can result in less anxiety and better well-being.

“I tell kids and their parents that you can’t pick your genetics,” says Dr. Harville about the guilt parents feel over their child’s condition. “It’s not their fault; it is just the way things are, and together we’ll figure out how to best manage the situation. There’s no one at fault, and no one is the culprit.”

Once a child becomes older, there is greater understanding about his or her specific health needs. Educational materials can help to explain how treatments are beneficial, and they can enable an older child’s curiosity and, in turn, ease his or her anxiety.

The diagnosis of a PIDD is a scary and often frustrating time for parents. However, in most cases, management of the disease can be straightforward, and a child can lead a relatively normal childhood. In addition to medical treatments, environmental impacts may have a significant effect on the health of a child with PIDD. Making changes to a routine to severely limit or completely curtail exposure to irritants will pay off. Even when things appear down, keeping a positive attitude is important. It helps parents to better cope and can help prevent a child from “feeling” the parents’ anxieties that in turn create anxieties in a child. Kids watch parents’ cues to learn how they should feel. A child’s team of medical experts is there to help and has many resources to make this potentially arduous journey more comfortable for all.

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

References

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Diagnosing and Treating Chronic Inflammatory Demyelinating Polyneuropathy

This autoimmune disease is difficult to diagnose due to its rarity and many variants, but once it is diagnosed, there are treatment options.

By Michelle Greer, RN, and Gil I. Wolfe, MD, FAAN
The primary function of the immune system is to differentiate between self and non-self, to keep self healthy and to destroy or neutralize non-self. When the immune system malfunctions and attacks itself, it is known as an autoimmune disease. Chronic inflammatory demyelinating polyneuropathy (CIDP) is considered an autoimmune disease. CIDP occurs when the myelin sheath that covers the nerves and assists with impulse transmission is attacked. This is known as demyelination. Due to the nature of the immune attack, there is usually inflammation. The result is an interruption in nerve signals between the peripheral nerves and the muscles they control.

CIDP presents slowly, usually over several months, unlike the acute form of demyelinating neuropathy, which is known as Guillain-Barré syndrome (GBS). GBS presents rapidly, usually over days, but sometimes even more quickly, and frequently, it occurs following some sort of infection or illness. Unlike GBS, CIDP is usually a chronically progressive neuropathy, and it is rarely associated with antecedent illnesses or respiratory failure.1

**Diagnosing CIDP**

Usually, CIDP presents as a motor predominant neuropathy with prominent proximal weakness, meaning the muscles responsible for movement closest to the torso are affected first. The weakness is typically symmetrical, affecting both sides of the body equally. Occasionally, CIDP can present in the pattern of a mononeuropathy multiplex, large-fiber neuropathy with sensory ataxia, pure motor neuropathy or small-fiber neuropathy.1

It’s not uncommon for CIDP to go undiagnosed for a while due to many factors. The symptoms may be vague and brushed off until they become more profound and/or interfere with everyday functioning. And once an individual does go to a physician, a definitive diagnosis still may not follow.

Neuropathy has many causes, and CIDP has several variants (see Table 1).2 Therefore, it is important that a

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<td>A. Symmetric proximal and distal motor predominant CIDP</td>
</tr>
<tr>
<td>B. Lewis-Sumner syndrome (LSS) (or multifocal acquired demyelinating sensory and motor neuropathy)</td>
</tr>
<tr>
<td>C. Demyelinating neuropathy with IgG or IgA paraprotein</td>
</tr>
<tr>
<td>D. Sensory predominant demyelinating neuropathy</td>
</tr>
<tr>
<td>E. CIDP neuropathy with central nervous system (CNS) demyelination</td>
</tr>
<tr>
<td>F. Demyelinating neuropathy associated with systemic disorders:</td>
</tr>
<tr>
<td>1. Hepatitis B or C</td>
</tr>
<tr>
<td>2. HIV</td>
</tr>
<tr>
<td>3. Lymphoma</td>
</tr>
<tr>
<td>4. Diabetes mellitus</td>
</tr>
<tr>
<td>5. Systemic lupus erythematosus or other collagen vascular disorders</td>
</tr>
<tr>
<td>6. Thyrotoxicosis</td>
</tr>
<tr>
<td>7. Organ or bone marrow transplants</td>
</tr>
<tr>
<td>8. Nephrotic syndrome</td>
</tr>
<tr>
<td>9. Inflammatory bowel disease</td>
</tr>
<tr>
<td>G. CIDP in patients who have inherited neuropathy</td>
</tr>
</tbody>
</table>


**Table 2. Diagnostic Tests**

<table>
<thead>
<tr>
<th>Test</th>
<th>What is it?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Electromyography (EMG)*</td>
<td>A procedure to measure and record muscle activity to show which muscles and nerves are affected.</td>
</tr>
<tr>
<td>Nerve Conduction Study (NCS)*</td>
<td>A procedure to measure the speed and efficiency of electrical signals of the nerves.</td>
</tr>
<tr>
<td>Lumbar Puncture</td>
<td>A spinal tap to look at the cerebral spinal fluid for abnormalities. Protein in the CSF is usually indicative of an immune response and can be present in CIDP.</td>
</tr>
<tr>
<td>Nerve Biopsy</td>
<td>A section of the nerve is taken and examined to look for cause of damage. Only done if diagnosis is unclear.</td>
</tr>
</tbody>
</table>

*An EMG and NCS are almost always both conducted in order to appropriately diagnose CIDP.
thorough health history and physical and neurological examination be performed to determine the cause of the neuropathy. CIDP is rare, but its incidence ranges greatly due to the potential of over- or underdiagnosis. An individual may be thought to have CIDP when it is actually another form of neuropathy, and the reverse can happen as well. Many physicians and patient groups have worked on a standard way to identify CIDP more quickly and accurately, but appropriate diagnosis remains a challenge.

Symptoms are first noticed as numbness, tingling, pain and weakness, which are vague and can be the initial symptoms of many conditions. This usually occurs first in the toes and feet, eventually resulting in foot drop or drag and increased difficulty in walking. The weakness and numbness are typically symmetrical — equal on both sides of the body — and sensory loss is often in a stocking and glove distribution.

A diagnosis of CIDP is based on an electrophysiologic pattern of multifocal demyelination identified through an EMG/nerve conduction study, elevated CSF (cerebral spinal

### Table 3. Standard Immunotherapy for Immune-Mediated Neuropathies

<table>
<thead>
<tr>
<th>Therapy</th>
<th>Neuropathy Types</th>
<th>Route</th>
<th>Starting Doses</th>
<th>Maintenance Doses</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prednisone*</td>
<td>CIDP, VN</td>
<td>Oral</td>
<td>60-100 mg/day for 4 weeks</td>
<td>60-100 mg every other day, reducing dose by 10 mg every 2-4 weeks. In diabetics, consider daily dosing to simplify glucose control</td>
</tr>
<tr>
<td>Methylprednisolone*</td>
<td>CIDP, VN</td>
<td>IV</td>
<td>1 gram daily or every other day for a total of 3-5 doses</td>
<td>Reduce dose by 10 mg</td>
</tr>
<tr>
<td>Azathioprine (Imuran)*</td>
<td>CIDP</td>
<td>Oral</td>
<td>50 mg/day</td>
<td>Increase by 50-mg increments every 2-4 weeks to 2-3 mg/kg/day</td>
</tr>
<tr>
<td>Cyclophosphamide (Cytoxan)*</td>
<td>CIDP, VN, MMN</td>
<td>Oral</td>
<td>50 mg/day</td>
<td>Increase to 1.5-2 mg/kg/day</td>
</tr>
<tr>
<td>Cyclophosphamide</td>
<td>CIDP, VN, MMN</td>
<td>IV</td>
<td>0.5-3 gm/m2</td>
<td>Repeat dose monthly for 6 months</td>
</tr>
<tr>
<td>Cyclosporine (Neoral, Sandimmune)*</td>
<td>CIDP</td>
<td>Oral</td>
<td>100 mg twice daily</td>
<td>Increase by 100-mg increments to 3-6 mg/kg/day on a twice daily schedule</td>
</tr>
<tr>
<td>IVIG</td>
<td>GBS*, CIDP, MMN</td>
<td>IV</td>
<td>2 gm/kg divided over 2-5 days</td>
<td>0.4-1 gm/kg as a single dose every 3-8 weeks as needed</td>
</tr>
<tr>
<td>Plasmapheresis</td>
<td>GBS, CIDP, MMN</td>
<td>IV</td>
<td>Exchange total of 250 mL/kg plasma</td>
<td>Total exchanges of 50-250 mL/kg may be repeated as needed over 7-14 days</td>
</tr>
<tr>
<td>Rituximab (Rituxan)*</td>
<td>MMN, IgM-associated neuropathy</td>
<td>IV</td>
<td>375 mg/m2 every week x 4 weeks</td>
<td>Repeat 375 mg/m2 as needed 9-15 months later</td>
</tr>
</tbody>
</table>

Abbreviations: CIDP=chronic inflammatory demyelinating polyneuropathy; GBS=Guillain-Barré syndrome; IgM=immunoglobulin M; IVIG=intravenous immunoglobulin; MMN=multifocal motor neuropathy; VN=vasculitic neuropathy.

* Not FDA approved for this indication.

fluid) protein and, when necessary, nerve biopsy. These tests, combined with a thorough health history and neurological exam, will help guide the physician to a correct diagnosis (see Table 2).

**Treating CIDP**

Once CIDP is diagnosed, treatment options are considered and discussed. The treatment of CIDP is based on immunomodulating therapies that are summarized in Table 3. Immunomodulation refers to suppression or alteration of the immune response so that attack on the self subsides and symptoms improve. CIDP does respond to corticosteroids; however, long-term use of high-dose steroids comes with its own set of issues. Side effects can be severe and affect multiple organ systems. Plasmapheresis is generally reserved for refractory patients — those who have tried all the standard therapies and the condition is still not controlled.² The only treatment that has received U.S. Food and Drug Administration (FDA) approval for the management of CIDP is intravenous immunoglobulin (IVIG).

**Rare, but There Is Hope**

Although CIDP is rare and difficult to diagnose, once it is accurately diagnosed, there are treatment options. CIDP can be treated with a variety of immunomodulatory therapies, including FDA-approved IVIG. Fortunately, CIDP can be managed to help patients live relatively normal and healthy lives, and there are many patient-to-patient support groups that include oversight by experts in neuromuscular disorders.

MICHELLE GREER, RN, is the vice president of sales at NuFACTOR Specialty Pharmacy. GIL I. WOLFE, MD, FAAN, is a professor and chair of the Department of Neurology at University at Buffalo School of Medicine and Biomedical Sciences, SUNY, where he holds the Irvin and Rosemary Smith Endowed Chair.

**References**

Understanding Chronic Lymphocytic Leukemia

CLL, the second-most common form of blood cancer, is being tested for more often and diagnosed earlier, and with new treatments, the quality of life for patients is vastly improved.

By Jim Trageser

Chronic lymphocytic leukemia (CLL) is one of the least publicized forms of cancer in the popular media, despite being the second-most common form of leukemia, which is referred to as cancer of the blood. Those who develop CLL usually do not have symptoms in the early stages. Patients diagnosed with this disease are usually in their middle years or older. However, patients as young as in their 20s and 30s may develop this type of illness. The most common age bracket for developing CLL are people in their 60s and 70s. It is thought that this disease may be more prominent in older people because the bone marrow (where blood cells are made) may not be as effective as it was in earlier years.

The National Institutes of Health reports that people of Eastern European or Jewish descent are more likely to develop CLL than are others. And, while it represents about one-third of all cases of leukemia, CLL is still relatively rare: Only about one in 200 people in the United States will develop CLL in their lifetime, with about 16,000 new cases diagnosed each year.

What Is CLL?

CLL tends to progress more slowly than other forms of leukemia, hence the word “chronic” in its name. In fact, CLL patients can be affected unknowingly with this illness for several years due to its slow-growing nature. CLL affects the lymphocyte cell line. This cell line belongs to the white blood cell group, which plays an important role in fighting infection. CLL develops from the B cell origin of lymphocytes, as opposed to the T cell.
lymphocytes. B cells help fight infection; T cells are “killer” cells. Without the proper function of B cells, one’s body cannot properly fight infections. Consequently, someone with CLL has an impaired immune system and can be at risk for more infections.

As with all forms of leukemia, CLL originates in the bone marrow, the spongy interior of bones where all blood cells are created. The blood cells start out as hematopoietic stem cells (immature cells that have not yet determined their cell line maturation), and further develop into mature blood cell lines such as platelets, red blood cells and the group of white blood cells. CLL affects the stem cell line that has been designated to form into mature B cells. But, due to the development of this disease, the lymphocytes grow out of control and accumulate in the bone marrow and blood, where they crowd out healthy blood cells.

Although CLL is a slow-growing cancer, it is best known for overcrowding the bone marrow and entering the peripheral blood system. Since blood cell production in the bone marrow needs to remain in constant balance, the overproduction of white blood cells causes a disturbance in other cell lines. Patients may exhibit forms of anemia, as well as high lymphocyte counts that can be detected in a complete blood count (CBC) test that detects the overabundant CLL cells proliferating out of balance.

Since CLL is a cancer of the B cell lymphocytes, it also can affect the lymphatic system and infiltrate lymph nodes, the spleen and other organs. The lymphatic system is, along with the bloodstream, part of the circulatory system that carries lymphatic fluid, which is a key part of the body’s immune system. When infiltration occurs, patients are diagnosed with “bulky disease,” meaning that they have palpable lymph nodes and other organs that have been infiltrated with the disease. CLL that is confined to the bone marrow is not considered “bulky disease” and is usually diagnosed at earlier stages.

Symptoms of CLL
As mentioned, CLL develops slowly and with today’s advanced medical diagnostic tools, it is often diagnosed before any symptoms appear. Those patients who do exhibit symptoms may experience any of the following: enlarged lymph nodes, swollen abdomen, fatigue, excessive sweating, changes in appetite, recurring infections, sudden weight loss, fever and excessive bruising (generally late in the disease’s development). Fatigue is one of the most common, chronic symptoms patients with CLL will battle daily. Patients may have frequent infections (colds, bronchitis) diagnosed. Many physicians today routinely check blood work on their patients, especially in older ages. A CBC with differential can help lead to the diagnosis of CLL.

Causes of CLL
At present, medical researchers do not know what causes most cases of CLL. It does not appear to be the type of cancer that is caused by inherited DNA mutations, and so it is likely the product of DNA damage acquired after birth. Yet, close relatives of those with CLL are more likely to develop it than are others.

In addition, environmental factors may be a risk factor for developing this disease. For example, people who were exposed to the herbicide Agent Orange, used in the Vietnam War to clear jungle, tend to have a higher risk of developing CLL.

There also are some suggestions that long-term exposure to other pesticides and herbicides may raise the risk, particularly among farmers, ranchers and agricultural workers, but this is still being studied. Interestingly, radiation exposure, smoking and other common risk factors known to be associated with other cancers do not seem to be tied to CLL.

To summarize, the major risk factors for CLL are advanced age (most cases develop in patients over 70 years of age) and ethnicity (Eastern European and Jewish people are most likely to develop the disease). It is extremely rare in Asia. Men also are slightly more likely to develop CLL than women, but the reason for this disparity is not understood.

Diagnosing CLL
Until the last 20 years, most CLL cases were not diagnosed until noticeable symptoms had arisen, which indicates progression of disease. Today, with many geriatricians and
general practitioners ordering blood work for their elderly patients, CLL can be diagnosed in its earliest stages.

People with CLL may display many different symptoms. Some may be anemic due to overcrowding of the bone marrow that doesn’t allow the red blood cell line to develop and mature. Patients may have infiltrated lymph nodes, therefore presenting with palpable lymph nodes. Patients may have a swollen abdomen due to spleen or liver involvement. But the most common complaint is fatigue; this is due to the many effects CLL has on the body and blood cells.

If a physician suspects a patient may have CLL or other blood-related cancers, a variety of tests can be ordered to determine the diagnosis. The first test to be performed is a CBC. In this test, the number and types of blood cells are counted and compared with what a healthy person should have. If patients have too many lymphocytes in their blood work, then CLL may be suspected. Usually, a decrease in red blood cells and/or platelets also will be reported. The physician may add another blood test called LDH, which can indicate the presence of CLL. The physician will then order a bone marrow biopsy to confirm the diagnosis. After taking a small piece of bone and marrow (where blood cells grow), the specimens are stained to indicate normal and abnormal cells. Then they are sent to a pathologist who will look at the cells under a microscope to confirm if the bone marrow has abnormal cells, such as abnormal lymphocytes — identifying a diagnosis of CLL.

Treatment for CLL

Once patients are diagnosed with CLL, the next step is to determine if it has spread. This is known as “staging,” and those who have had a friend or relative with cancer are familiar with the terms “stage 1 cancer” or “stage 4 cancer” (stage 1 being the earliest form, and stage 4 indicating the cancer has spread). As additional tests are ordered, samples may be taken from the lymph nodes, bone marrow or spinal fluid to see if and how far the cancer has spread. The doctor also may order a CT scan to look for infiltration into lymph nodes in the abdomen and spleen.

Early-stage CLL is typically treated with observation. If the CLL has progressed, or if there are complications (repeated infections or indications that a rare but aggressive form of CLL is present), chemotherapy may be prescribed. Radiation is generally not the first line of therapy for eradicating CLL, but it may be used to help fight infiltrated lymph nodes or other tissue where CLL cells have implanted.

Fludarabine is a chemotherapy drug that interferes with DNA's ability to reproduce itself. (DNA is the molecule that contains all of a person’s genetic information; when a cell divides, it must first make an identical copy of its DNA so each of the two new cells will have all the genetic information of the original cell.) Since cancerous cells reproduce so much more rapidly than healthy cells, the drug has more of an impact on the cancerous cells than healthy ones. The drug also tends to stay in the bloodstream, so it does not affect other tissues very much. This is a very well-tolerated drug and is usually used in the first line of therapy.

Early-stage CLL is typically treated with observation.

Another drug called Rituxan, which is a monoclonal antibody, is widely used to fight CLL. Rituxan is able to attack cells that are CD20+. These cells include CLL cells. Since Rituxan can “find” CD20+ cells, it is a well-tolerated drug because it only affects those cells. Rituxan is commonly used in conjunction with Fludarabine. The combination of these two drugs has shown to be extremely effective in treating CLL.

A chemotherapy drug called Leukeran (chlorambucil) also may be given, although it has mostly been replaced by Fludarabine. Leukeran is an alkylating agent, which attaches an extra molecule to a cell’s DNA, so the cell can no longer divide and reproduce. Again, this affects cancerous cells more than healthy ones.

Cytoxan is also an alkylating agent, working similarly to Leukeran. Cytoxan is generally used in combination with Fludarabine and Rituxan in patients with advanced bulky disease. These drugs together have been shown to be one of the most effective treatments for CLL.

All of these drugs also may be prescribed in combination, along with blood transfusions, to provide healthy white blood cells to increase patients’ immunological defenses.

Alemtuzumab, known as Campath, also targets CLL cells. This monoclonal antibody targets cells that exhibit CD23+ antigens. Through advances in science, we have learned that CLL cells specifically can display this antigen, therefore only CLL cells are targeted and irradiated. Campath is usually used as a second line of therapy, although studies are being conducted to determine...
whether Campath in combination with other drugs can achieve a first remission.

Bendamustine is another chemotherapy agent that may be prescribed. It works by creating cross-strands between DNA strands so that cells cannot reproduce. 14

The compromised immune system caused by CLL can be assisted in fighting infection with vaccines or regular immunoglobulin (IG) therapy. These treatments help the body restore its ability to fight infection. And, patients undergoing chemotherapy will have the typical side effects that accompany use of the various drugs. Since B cells express IG (the most abundant form of globulin found in the blood that helps fight infections) and CLL affects B cells, intravenous IG replacement therapy is usually necessary to boost the immune system to help keep the patient from acquiring multiple infections.

The only currently available treatment considered to provide a cure (which is no evidence of disease for five years or more) is a bone marrow transplant or stem cell transplant. Transplants may be attempted if a remission has been achieved. 1

Managing the Disease

Since the only cure for CLL is a bone marrow transplant or stem cell transplant, and patient selection for this therapy can be limited due to the potential side effects, most people with CLL will live with the disease for the rest of their lives. Patients who still have evidence of disease but no symptoms can be considered in “remission” but not cured.

About half of all patients diagnosed in early stages of CLL will live more than 12 years. 1 Given the typical age at which CLL appears, many of those patients will die of a cause other than CLL. Many patients with early-stage CLL will live years with the disease with no treatment other than regular follow-up visits with their doctor, and with few changes in their lifestyles.

All patients with CLL, even those in remission, will want to maintain frequent, regular contact with their doctors. 15

Looking Ahead

There are numerous new approaches in treatment now in clinical trials or earlier research that offer hope of improved treatment and even nonsurgical cures for CLL. From programming the body’s own T cells (one type of infection-fighting white blood cell) to attack and destroy cancerous cells, to using genetic therapies to cause the body to reject cancerous cells, progress is being made by researchers on several fronts. 16 Other possibilities include targeting proteins that are unique to cancerous cells. All of these treatments offer the promise of more effective means of slowing the development and spread of CLL, and some may lead to an actual cure.

Currently, CLL patients can live a normal life and have minimal, if any, side effects from the newest treatments available. As treatments improve their efficiency in fighting CLL, and supporting therapies like IG treatment strengthen patients’ ability to fight infections, the quality of life for patients diagnosed with CLL continues to improve.

JIM TRAGESER is a newspaper editor and has contributed to two reference books.

References

Let’s Talk!

By Trudie Mitschang

Constance McNamara Romanowski lives with multiple chronic illnesses, most notably the rare and debilitating Evans syndrome (ES). Her journey has been a difficult one, but today Constance is the executive director of the Evans Syndrome Community Network, whose mission it is to provide a safe networking haven for individuals whose lives have been impacted by ES. Her story inspired us.

Trudie: What was it like growing up with chronic illness?

Constance: At the age of 2, I had a violent reaction to penicillin. This was at the very beginning of my personal experiences with weakened immunity and autoimmunity problems. From that point on, I was chronically anemic. When I started menstruating at age 11, my flow would be so heavy that I would pass out. Every time my blood work came back showing that I was anemic, the doctors would just explain it away, saying I “wasn’t eating well.” Later in life, the doctors said it was because I was in my childbearing years, and later they said it was because I was in perimenopause. Unfortunately, no tests were ever performed to determine the underlying cause.

Trudie: How were you diagnosed with ES?

Constance: In April of 2005, I got very sick. Years before, I was diagnosed with irritable bowel syndrome, and this seemed like a really acute attack. I ended up at the urgent care clinic, where I was diagnosed with stomach flu and prescribed an antibiotic. A week later when I returned to work, I noticed very tiny red dots appearing on the insides of my wrists. They didn’t itch, weren’t raised and were all uniform in size. I saw my doctor and she sent me to the emergency room for a CBC and chest X-ray. As I waited to be seen, the red dots spread to my feet and up my legs.

Trudie: What happened next?

Constance: It turned out the red dots were called petechiae, which are actually tiny hemorrhages just under the skin, the result of a low platelet count. A normal count would be 100,000 or higher; mine had fallen to about 4,000. And my hemoglobin, which should be at least 11.5, was 8. They did a battery of tests, including a bone marrow biopsy, ruling out all other diseases before they told me that I had Evans syndrome.

Trudie: What is ES?

Constance: ES is a combination of idiopathic autoimmune hemolytic anemia and idiopathic thrombocytopenic purpura. Those two things together are ES. In layman’s terms, this means that the immune system has turned against itself, and the wrong cells are being marked for destruction. The immune system is in overdrive, marking healthy red blood cells and platelets for destruction by the spleen.

Trudie: What are the symptoms?

Constance: Symptoms vary, but things to look for include dark urine; an enlarged or painful spleen; fatigue; pale skin; a rapid heartbeat; shortness of breath; yellowing of the skin and/or whites of the eyes; abnormally low blood pressure; excessive or easy bruising; clusters of tiny red or purple dots on the skin; prolonged bleeding from skin injury; bleeding from the gums and nose; blood in the urine and/or stool; and unusually heavy menstruation. The crazy thing is that a person can have ES and have absolutely no symptoms at all.
Trudie: How is ES treated?
Constance: Prednisone tops the list. Sometimes anti-rejection drugs are used to treat the problem like a rejected organ. Sometimes doctors turn to an IV treatment called rituximab, which is a chimeric monoclonal antibody that was first used in the treatment of lymphoma. Once all other medical interventions are exhausted, a final option may be a bone marrow transplant.

Trudie: What was your experience with intravenous immune globulin (IVIG)?
Constance: For me, IVIG was the stop-gap that saved my life while waiting for IV steroids to kick in. I don’t think most people realize that when they give blood or plasma, there are many things that can be done with that donation. IVIG is one of those things. It’s mind-boggling to think how many people had to donate blood in order for me to get just one bottle of IVIG — it’s likely thousands, and I’m very thankful.

Trudie: Are you still on IVIG?
Constance: No, but there may come a time when I need it.

Trudie: What is the Evans Syndrome Community Network and what motivated you to start it?
Constance: When I was first diagnosed, I went straight to the Internet in search of information. The problem was there was not much available. But, I realized that I wasn’t in this alone; ES strikes roughly one in one million people, so there had to be others out there. I needed to find a way to get from this desperate, hand-wringer stage to a place where I felt more empowered.

My husband and I started a website and began gathering information that we thought might be helpful. We called it the Evans Syndrome Community Network. Our hope was that we could reach others all over the world who shared this diagnosis.

Trudie: How did you get the word out?
Constance: Through a fluke. I joined Facebook, and I started a group there as well to benefit those impacted by ES. The group grew slowly at first, and then it started to pick up. Now, we have almost 400 members from all over the world, and they’re quite talkative. We’re going through the process of creating a full-fledged nonprofit foundation for our group. Our website is shut down and going through a refit, but our Facebook page is up and running. We also are working with the National Organization of Rare Disorders (NORD) in their effort called RareConnect, a new social network where rare disease patients can connect with others globally.

Our hope was that we could reach others all over the world who shared this diagnosis.

For more information about Evans Syndrome, visit: www.rareconnect.org/community/evans-syndrome and www.evanssyndrome.net.

Trudie: What are your goals and dreams?
Constance: My ultimate goal in life is just to make a positive difference in the lives of those around me. This helped guide us when we came up with the motto for the Evans Syndrome Community Network: “Together, we will make a difference, standing shoulder to shoulder.”

Trudie: What advice would you offer others?
Constance: Be persistent if you think something is wrong. Don’t let anybody tell you it’s in your head or you’re being silly. Be your own best advocate. Try to live each day to the fullest. Every moment is a blessing. And, remember: You can make a difference. Just try it and see.

TRUDIE MITSCANG is a staff writer for IG Living magazine.

If your life depends on immune globulin and you have a unique experience to share, we want to feature you in this column! Email us at editor@IGLiving.com.
Reader: My son is a teenager with a primary immune deficiency disease and, as a result, needs immune globulin (IG) therapy. All of his friends are leaving home for college. My son would like to do the same thing, but it scares me to have him far away. Do you have a checklist of things I can do to help him prepare?

Annaben: While it is scary to let your son go off to college and trust that he will be compliant with his therapy, it can be done successfully. It is important to remember that even though your son’s disease affects the whole family, it is still his disease and, ultimately, the treatments are his responsibility.

The transition to independence should be started long before your son leaves home, while he is still under your care. I would suggest you and your son set a goal that he become independent with his healthcare for at least six months prior to leaving home.

Nancy Creadon, RN, an IG Living advisory board member with several years of experience in specialty infusions, also has some advice: “At the university orientation, you and your child will be introduced to personnel at the Student Health Center, and you will have the opportunity to complete documents that notate his insurance and private health information regarding past medical history. Your son should complete these documents and turn them in. Then, once he arrives at school, the nurses and physicians will already have complete information, easing the stress of trying to articulate the past medical history in the event he may need more immediate care. As the mother of four college students, I have been very pleased and thankful for the outstanding care these medical professionals provide. They understand that their mission is to educate and help the kids achieve independence in managing their own care.”

Here is a list of things your son should be able to do while at college:

1. Make his own appointments.
2. Create a list of questions to ask healthcare providers before his appointments.
3. Know the name of his disease and be able to explain what it is to new healthcare providers.
4. Create a list of current medications, allergies, diagnoses, emergency contacts and providers with contact information. (It is a good idea to keep a copy on a thumb drive or smartphone.)
5. Have a copy of his insurance card with the subscriber, group number and subscriber number, and understand the terms.
6. Access insurance information online and know how to navigate it so that he can find in-network providers.
7. Understand the importance of a HIPAA-compliant release form. (Remember, once your son turns 18, you will need his permission to access his health records and speak with his healthcare providers.)
8. Understand co-pays for office visits and the pharmacy, and know the expected amount.
9. Call in a refill for prescriptions
10. If applicable for home infusion, take over ordering infusion supplies and keep them organized.
11. Keep an infusion log and health diary.

Also, have your son fill out the patient forms at his current doctor’s office, and check them to make sure the information is correct and complete. And, consider having your son visit one of his current physician’s partners whom he has not seen before. This will allow him to practice introducing himself and his disease to a new doctor. It also will help him learn what to expect when visiting a new doctor in a new area without you, but with the comfort of knowing his current doctor can check on his progress toward independence.

ANNABEN KAZEMI is the patient advocate for IG Living magazine.
If you are a Hizentra patient or caregiver

Sometimes talking to someone who "gets it" is KEY.

Voice2Voice

Your key to explore Voice2Voice online

Punch out this Web key and plug into your computer's USB port to learn all about Voice2Voice.

Voice2Voice is a peer-to-peer support program from CSL Behring, the maker of Hizentra.

Voice2Voice connects Hizentra patients and caregivers with advocates* who have direct experience with Hizentra and know what it's like to live with primary immunodeficiency disease (PIDD).

Go online and view stories from patients like Jacob and his mother, Janet, a Voice2Voice advocate, to see what it's all about!

Sign up for Voice2Voice.
You can enroll online at Hizentra.com/V2v or call 1-877-355-IGIQ (4447) for assistance.

Janet
Jacob

*Voice2Voice advocates are not healthcare professionals or medical experts. For medical questions, please contact your physician. Individuals appearing in the Voice2Voice videos are compensated by CSL Behring LLC for their time and/or expenses.

Important Safety Information
Hizentra treats various forms of primary immunodeficiency (PI) in patients age 2 and over.
Hizentra should not be used if you have had serious negative reactions to immune globulin (Ig) preparations or a deficiency of an Ig known as IgA. Because Hizentra contains the amino acid proline as stabilizer, patients with hyperprolinemia (too much proline in the blood) should not take Hizentra.

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.

Please see additional Important Safety Information on next page.
Please see brief summary of full prescribing information for Hizentra on adjacent pages.
For people with PIDD

Hizentra is the Ig therapy that's deliberately designed for SubQ use

Backed by the expertise of CSL Behring, Hizentra 20% is currently being used by more than 10,000 patients and providers,¹ a number that's growing every day

- Hizentra helps keep IgG levels stable with low-volume self-infusions
  - The first and only 20% Ig concentration delivers a consistent level of protection against infection
  - Individualized dosing means you can have confidence that you are getting the dose that's right for you

**Important Safety Information** (continued)

Tell your doctor about any side effects that concern you. Your doctor will monitor for potentially serious reactions that have been seen with Ig treatment, including thrombotic events (blood clotting); aseptic meningitis syndrome (brain swelling); osmotic nephropathy (a kidney condition); hemolysis (a blood problem) and transfusion-related acute lung injury.

The most common drug-related adverse reactions in the clinical trial for Hizentra were injection-site reactions (swelling, pain, redness, heat, or itching); 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.

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

**Please see brief summary of full prescribing information for Hizentra on adjacent pages.**

You are encouraged to report negative effects of prescription drugs to the FDA. Visit [www.fda.gov/medwatch](http://www.fda.gov/medwatch), or call 1-800-FDA-1088.

Reference: ¹ Data on File. Available from CSL Behring as DOF HIZ-103.
CSL Behring

BRIEF SUMMARY OF PRESCRIBING INFORMATION

Hizentra®,
Immune Globulin Subcutaneous (Human), 20% Liquid

Before prescribing, please consult full prescribing information, a brief summary of which follows. Some text and references refer to full prescribing information.

1  INDICATIONS AND USAGE

Hizentra is an Immune Globulin Subcutaneous (Human) (IGSC), 20% Liquid indicated as replacement therapy for primary humoral immunodeficiency (PI) in adults and pediatric patients 2 years of age and older. This includes, but is not limited to, the humoral immune defect in congenital agammaglobulinemia, common variable immunodeficiency, X-linked agammaglobulinemia, Wiskott-Aldrich syndrome, and severe combined immunodeficiencies.

2  CONTRAINDICATIONS

Hizentra is contraindicated in patients who have had an anaphylactic or severe systemic reaction to the administration of human immune globulin or to components of Hizentra, such as polysorbate 80.

Hizentra is contraindicated in patients with hyperprolinaemia because it contains the stabilizer L-proline (see Description (11)).

Hizentra is contraindicated in IgA-deficient patients with antibodies against IgA and a history of hypersensitivity (see Description (11)).

3  WARNINGS AND PRECAUTIONS

5.1 Hypersensitivity

Severe hypersensitivity reactions may occur to human immune globulin or components of Hizentra, such as polysorbate 80. In case of hypersensitivity, discontinue the Hizentra infusion immediately and institute appropriate treatment.

Individuals with IgA deficiency can develop anti-IgA antibodies and anaphylactic reactions (including anaphylaxis and shock) after administration of blood components containing IgA. Patients with known antibodies to IgA may have a greater risk of developing potentially severe hypersensitivity and anaphylactic reactions with administration of Hizentra. Hizentra contains ≤50 mcg/mL IgA (see Description (11)).

5.2 Thrombotic Events

Thrombotic events have been reported with the use of immune globulin products1-2, including Hizentra. Patients at increased risk may include those with a history of atherosclerosis, multiple cardiovascular risk factors, advanced age, impaired cardiac output, hypercoagulable disorders, prolonged periods of immobilization, Factor V Leiden, known or suspected hyperviscosity, and/or those who use estrogen-containing products.

Because of the potentially increased risk of thrombosis, consider baseline assessment of blood viscosity in patients at risk for hyperviscosity, including those with cryoglobulins, fasting chylomicronemia, markedly high triglycerides (triglycerides), or monoclonal gammapathies. For patients judged to be at risk of developing thrombotic events, administer Hizentra at the minimum rate practicable.

5.3 Aseptic Meningitis Syndrome (AMS)

AMS has been reported with use of IGIV3 or IGSC. The syndrome usually begins within several hours to 2 days following immune globulin treatment. AMS is characterized by the following signs and symptoms: severe headache, nuchal rigidity, drowsiness, fever, photophobia, painful eye movements, nausea, and vomiting. Cerebrospinal fluid (CSF) studies frequently show pleocytosis up to several thousand cells per cubic millimeter, predominantly from the granulocytic series, and elevated protein levels up to several hundred mg/dL. AMS may occur more frequently in association with high doses (≥2 g/kg) and/or rapid infusion of immune globulin product.

Patients exhibiting such signs and symptoms should receive a thorough neurological examination, including CSF studies, to rule out other causes of meningitis. Discontinuation of immune globulin treatment has resulted in remission of AMS within several days without sequelae.

5.4 Reactions Reported to Occur With IGIV Treatment

The following reactions have been reported to occur with IGIV treatment and may occur with IGSC treatment.

Renal Dysfunction/Failure

Renal dysfunction/failure, osmotic nephropathy, and death may occur with use of human immune globulin products. Ensure that patients are not volume depleted and assess renal function, including measurement of blood urea nitrogen (BUN) and serum creatinine, before the initial infusion of Hizentra and at appropriate intervals thereafter. Periodic monitoring of renal function and urine output is particularly important in patients judged to have a potential increased risk of developing acute renal failure. If renal function deteriorates, consider discontinuing Hizentra. For patients judged to be at risk of developing renal dysfunction because of pre-existing renal insufficiency or predisposition to acute renal failure (such as those with diabetes mellitus or hypovolemia, those who are overweight or use concomitant nephrotoxic medicinal products, or those who are over 65 years of age), administer Hizentra at the minimum rate practicable.

Hemolysis

Hizentra can contain blood group antibodies that may act as hemolysins and induce in vivo coating of red blood cells (RBCs) with immunoglobulin, causing a positive direct antiglobulin (Coombs') test result and hemolysis.4-6 Delayed hemolytic anemia can develop subsequent to the administration of globulin therapy due to enhanced RBC sequestration, and acute hemolysis, consistent with intravascular hemolysis, has been reported.9 Monitor recipients of Hizentra for clinical signs and symptoms of hemolysis. If these are present after a Hizentra infusion, perform appropriate confirmatory laboratory testing. If transfusion is indicated for patients who develop hemolysis with clinically compromising anemia after receiving Hizentra, perform adequate cross-match testing to avoid exacerbating on-going hemolysis.

Transfusion-Related Acute Lung Injury (TRALI)

Noncardiogenic pulmonary edema may occur in patients administered human immune globulin products.1-3 TRALI is characterized by severe respiratory distress, pulmonary edema, hypoxemia, normal left ventricular function, and fever. Typically, it occurs within 1 to 6 hours following transfusion. Patients with TRALI may be managed using oxygen therapy with adequate ventilatory support.

Monitor Hizentra recipients for pulmonary adverse reactions. If TRALI is suspected, perform appropriate tests for the presence of anti-neutrophil antibodies in both the product and patient's serum.

5.5 Transmissible Infectious Agents

Because Hizentra is made from human plasma, it may carry a risk of transmitting infectious agents (e.g., viruses, and theoretically, the Creutzfeldt-Jakob disease [CJD] agent). The risk of infectious agent transmission has been reduced by screening plasma donors for prior exposure to certain viruses, testing for the presence of certain current virus infections, and including virus inactivation/neutralization steps in the manufacturing process for Hizentra.

Report all infections thought to be possibly transmitted by Hizentra to CSL Behring Pharmacovigilance at 1-866-915-6958.

5.6 Laboratory Tests

Various passively transferred antibodies in immunoglobulin preparations may lead to misinterpretation of the results of serological testing.

6  ADVERSE REACTIONS

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

6.1 Clinical Trials Experience

Because clinical studies are conducted under widely varying conditions, AR rates observed in clinical studies of a product cannot be directly compared to rates in the clinical studies of another product and may not reflect the rates observed in clinical practice.

US Study

The safety of Hizentra was evaluated in a clinical study in the US for 15 months (3-month wash-in/wash-out period followed by a 12-month efficacy period) in subjects with PI who had been treated previously with IGIV every 3 or 4 weeks. The safety analyses included 49 subjects in the intention-to-treat (ITT) population. The ITT population consisted of all subjects who received at least one dose of Hizentra (see Clinical Studies (14)). Subjects were treated with Hizentra at weekly median doses ranging from 66 to 331 mg/kg body weight (mean: 181.4 mg/kg) during the wash-in/wash-out period and from 72 to 379 mg/kg (mean: 213.2 mg/kg) during the efficacy period. The 49 subjects received a total of 2264 weekly infusions of Hizentra.

Table 2 summarizes the most frequent adverse reactions (ARs) (experienced by at least 2 subjects) occurring during or within 72 hours after the end of an infusion. Local reactions were assessed by the investigators 15 to 45 minutes post-infusion and by the subjects 24 hours post-infusion. The investigators then evaluated the ARs arising from the subject assessments. Local reactions were the most frequent ARs observed, with injection-site reactions (e.g., swelling, redness, heat, pain, and itching at the site of injection) comprising 98% of local reactions.
Subjects were treated with Hizentra at weekly median doses ranging from 59 to 267 mg/kg.

In a clinical study conducted in Europe, the safety of Hizentra was evaluated for 10 months including local reactions, to all infusions was 1303 to 2264 (57.6%).

The proportion of subjects reporting local reactions decreased over time from approximately 20% following the first infusion to <5% by the end of the study.

Three subjects withdrew from the study due to ARs of mild to moderate intensity. One subject experienced injection-site pain and injection-site pruritus; the second subject experienced injection-site reaction, fatigue, and feeling cold; and the third subject experienced injection-site reaction and hypersensitivity. All reactions were judged by the investigator to be "at least possibly related" to the administration of Hizentra.

6.2 Postmarketing Experience

Because postmarketing reporting of adverse reactions is voluntary and from a population of uncertain size, it is not always possible to reliably estimate the frequency of these reactions or establish a causal relationship to product exposure.

Hizentra

The following adverse reactions have been identified during postmarketing use of Hizentra.

This list does not include reactions already reported in clinical studies with Hizentra (see Adverse Reactions [6.1]).

- **Infusion reactions:** Allergic-anaphylactic reactions (including swollen face or tongue and pharyngeal edema), pyrexia, chills, dizziness, hypertension/changes in blood pressure, malaise.
- **Cardiovascular:** Thromboembolic events, chest discomfort (including chest pain)
- **Respiratory:** Dyspnea

General

The following adverse reactions have been reported during postmarketing use of immune globulin products:

- **Infusion reactions:** Hypersensitivity (e.g., anaphylaxis), headache, diarrhea, tachycardia, fever, fatigue, dizziness, malaise, chills, flushing, urticaria or other skin reactions, wheezing or other chest discomfort, nausea, vomiting, rigors, back pain, myalgia, arthralgia, and changes in blood pressure
- **Renal:** Acute renal dysfunction/failure, osmotic nephropathy
- **Respiratory:** Apnea, Acute Respiratory Distress Syndrome (ARDS), TRALI, cyanosis, hypoxemia, pulmonary edema, dyspnea, bronchospasms
- **Cardiovascular:** Cardiac arrest, thromboembolism, vascular collapse, hypotension
- **Neurological:** Coma, loss of consciousness, seizures, tremor, aseptic meningitis syndrome
- **Integumentary:** Stevens-Johnson syndrome, epidermolysis, erythema multiforme, dermatitis (e.g., bullous dermatitis)
- **Hematologic:** Pancytopenia, leukopenia, hemolysis, positive direct antiglobulin (Coombs') test
- **Gastrointestinal:** Hepatic dysfunction, abdominal pain
- **General/Body as a Whole:** Pyrexia, rigors

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.

Table 2: Incidence of Subjects With Adverse Reactions (ARs)* (Experienced by 2 or More Subjects) and Rate per Infusion (ITT Population), US Study

<table>
<thead>
<tr>
<th>Injection-Site Reaction</th>
<th>Number* (Rate †) of Reactions (n=683 Infusions)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Edema/induration</td>
<td>467 (0.68)</td>
</tr>
<tr>
<td>Erythema</td>
<td>346 (0.51)</td>
</tr>
<tr>
<td>Local heat</td>
<td>108 (0.16)</td>
</tr>
<tr>
<td>Local pain</td>
<td>88 (0.13)</td>
</tr>
<tr>
<td>Itching</td>
<td>64 (0.09)</td>
</tr>
</tbody>
</table>

* Excluding infections.
† Rate of ARs per infusion.
‡ Includes injection-site reactions as well as bruising, scabbing, pain, irritation, cysts, eczema, and nodules at the injection site.
§ Number of infusions administered during regularly scheduled visits.

Most local reactions were either mild (93.4%) or moderate (6.3%) in intensity. No deaths or serious ARs occurred during the study. Two subjects withdrew from the study due to ARs. One subject experienced a severe injection-site reaction one day after the third weekly infusion, and the other subject experienced moderate myositis. Both reactions were judged to be "at least possibly related" to the administration of Hizentra.

European Study

In a clinical study conducted in Europe, the safety of Hizentra was evaluated for 10 months (3-month wash-in/wash-out period followed by a 7-month efficacy period) in 51 subjects with PI who had been treated previously with IVIG every 3 or 4 weeks or with IGSC weekly.

Subjects were treated with Hizentra at weekly median doses ranging from 59 to 267 mg/kg body weight (mean: 118.8 mg/kg) during the wash-in/wash-out period and from 59 to 243 mg/kg (mean: 120.1 mg/kg) during the efficacy period. The 51 subjects received a total of 1831 infusions of Hizentra.

Table 4 summarizing the most frequent ARs (experienced by at least 2 subjects) occurring during or within 24 hours after the end of an infusion. Local reactions were assessed by the subjects between 24 and 72 hours post-infusion. The investigators then evaluated the ARs arising from the subject assessments.
SOMETIMES I WISH I were a many-handed god like Vishnu. I feel like this when I’m fatigued, my hands aren’t working well, and “stuff” needs to get done. It’s as though I’m spinning plates in the air on sticks and trying to make sure they don’t fall to the ground: the house needs to be cleaned, I have laundry to do, the dog needs to be walked. It can all be so overwhelming, and I wonder at these times how I’m going to get everything done before the plates come crashing to the ground.

It’s taken me five years to learn a simple lesson, and I’ll share my secret with you: Stop trying to do it all yourself, and ask for help. You know what? People are very happy and willing to help.

I can’t believe I’m confessing this to all of you. I am a very proud person, and I have a hard time asking others for help. In the past, people have been unreliable and disappointing, so I learned to count on numero uno to get it all done. But now that I have an “invisible illness” — one that isn’t readily seen — my life has changed not only physically, but also emotionally. I was embarrassed to ask for assistance when it appeared I could do the task myself.

One day while taking a stroll with one of my dearest friends, I realized the shopping bag I was carrying was getting too heavy. I just couldn’t hold it anymore. I apologetically asked her to please carry my shopping bag for me. She looked at me with a smiling face and kind eyes and said: “Many hands make light work.” She then promptly took my bag and we continued walking. I had never heard the saying before. Zing! It hit me to the core. This simple proverb (by writer John Heywood) helped me overcome my resistance to ask for assistance. I have no problem at work getting the help I need for projects, so why was I not applying this attitude to my personal life?

Now, when my husband leaves before I do and I haven’t put on my jewelry, I sometimes have colleagues at work do it for me. Once, I happened to be picking up a prescription and asked the pharmacist to put on my necklace and bracelets. Turns out we had a delightful chat about accessories. I’ve learned to speak up and ask people to walk more slowly with me, wait while I write something down because it takes longer, or even open a bottle of water because the cap is on too tightly. Turns out they are only too happy to be helpful, and I’ve noticed a deeper connection between us during these brief interactions. I hear Blanche DuBois’ voice saying, “I’ve always relied on the kindness of strangers,” when a kind gentleman at the sandwich shop carries my beverage to the table because I can’t carry both my plate and a drink without dropping them.

I am deeply humbled and in continuous awe of the generous spirit of others’ kind deeds. My chronic illness has taught me many lessons over the years: how to be more patient, how to value every day more and, now, how to accept help. A spinning plate will drop and crash sometimes. That’s OK; it’s a reminder that either I’m doing too much or I need to reach out for those extra helping hands. I’m glad I’m not a many-handed god. I’d miss out on the special moments shared with others along this chronic illness journey.

STACY OLIVER was diagnosed in 2008 with multifocal motor neuropathy (MMN). She is the assistant director of the Center for the Writing Arts at Northwestern University, a jewelry and accessory artist (www.dancingstonesjewelry.com), and she is working on her secret super hero identity as Neuropathy Girl.
“DID YOU PACK HER BLANKIE?”
I hollered in hopes my husband, Mark, heard me. I was elbow deep in the fridge, frantically packing for our bimonthly trip to the hospital for another lifesaving intravenous immune globulin (IVIG) infusion.

“Yes, dear,” Mark sing-songed sarcastically while he swapped out our son Caleb’s car seat with Molly’s (it was Caleb’s turn the week before).

“Is it the blankie with holes big enough for her right thumb to fit through? You know she’ll be impossible for Nurse Annie if Molly can’t suck her thumb while they’re trying to start her IV,” I warned and nagged in the same breath.

“Yes, dear,” Mark repeated, and then, before I had a chance to translate thought to word, Mark jumped in: “And I packed the soft pink blankie made by Aunt Judy, not the gray one made by Aunt Sophie. The one by Aunt Sophie feels like sandpaper and isn’t ‘holey’ enough.”

“And don’t forget her special bear. You know, the one with the cute little bow tie and silky white vest,” I added.

One hour later, we were off to the Children’s Hospital (again), packed
to the gills with a few of our primary immune deficiency disease (PIDD) kid’s favorite things.

“My Favorite Things” to most is the title of a classic holiday song. For the PIDD community, favorite things are absolutely necessary items that comfort, calm, soothe and sustain during all-too-frequent illnesses, infusions and IV sticks.

When our PIDD kids were still in diapers, IV placements were agonizing for all involved. We can finally laugh about it now (10 years later), but by the time the IV was secured, we’d be covered from head to toe in EMLA cream, misguided saline and miles of extra tubing.

Around the fifth go-around with “IV wars,” our wonderful nurse asked: “Do you guys have any pets?”

“Not at this time, why?” I asked.

“I was just thinking that when our patients get to interact with the service dogs that visit the hospital,” she explained, with a sense of calm in her voice, “it’s amazing how frightening procedures become much easier on all of us.”

Adding another favorite thing to our stockpile didn’t make much sense, especially because a dog would be one more thing I had to feed, clean up after and take to the doctor. But because little about the kids’ immune disease made much sense, it made perfect sense to adopt George, a 120-pound chocolate Labrador retriever (a.k.a. the four-legged stomach).

About three weeks into life with George, he started not feeling so well. He was mopey and downhearted. When he stopped eating, drinking and playing fetch, we really knew something was wrong. So not only was I a complete wreck over the new four-legged family member, but also at a breaking point with Molly because her favorite thing — the dapper-dressed teddy bear — had disappeared, and it was her turn for IVIG.

“Mark, that bear better magically appear outta thin air, or you might as well put me in the funny farm,” I announced sternly from under Molly’s toddler bed.

“Um, not to be an uninformed and insensitive hack, but what are you doing, um, Honey?” Mark stumbled, befuddled as to why I was straining over something under Molly’s bed.

“Looking for Elvis, what else?”

“Hun, Elvis has been dead for, uh…”

“Not the Elvis!” I bellowed from below. “Molly’s little stuffed bear! She named it Elvis!”

“I guess our daughter thinks he’s a hunk of burning love!” Mark joked, badly.

“One thing’s for sure, if we don’t find this bear by IV time tomorrow, the only thing burning will be our ears from Molly’s inconsolable screaming!”

We were pushing midnight with no Elvis sightings. I was beginning to lose hope when a strange object on the backyard grass caught Mark’s eye.

“That’s strange,” he said walking cautiously toward the unknown object. As he sneaked closer and closer, it became clearer and clearer what had invaded our backyard.

“Cheryl, come here!” Mark whispered, bent at the hip and motioning for me with his left hand. “You gotta see this!”

Mark pointed toward the grass with a grin on his face, trying hard not to start laughing. With every bit of self-restraint he could muster, Mark announced, “Elvis has left the retriever.”

Sure enough, lying in the soft green grass among the other, um, favorite things he had ingested, George had deposited Elvis perfectly intact: bow tie, vest and the same silly grin that made him so adorable. Despite the adventure he’d just been on, including a desperate need for Spray ‘N’ Wash and bleach, Elvis was going to be OK. As for George, the minute he relieved himself of Elvis, he was like a new dog ready to resume his duties as keeper of the PIDD kids.

I don’t recall laughing as hard as I did that night and, yes, Elvis still managed a successful appearance for Molly’s infusion the following day. On the drive home from the hospital, relieved beyond words that the last few days were behind me, I looked at Mark and said, “I sure could use a tall, icy-cold slug of my favorite drink.”

Mark met my eyes and responded in his best Okie, “Why don’t I stop at a gas station and pick us up a little sumpin’ sumpin’?”

We couldn’t stop laughing, which sure felt better than all the fretting we’d been through the past week. Then Mark, in his best Julie Andrews, began singing, “Raindrops on roses and whiskers on kittens.”


CHERYL L. HAGGARD is a stay-at-home mom and has three children, two of whom have CVID. She and her husband, Mark, also operate Under the Hood Ministries at www.underthehoodministries.org.
Searching for an elusive diagnosis can be a long, daunting and frustrating journey for a chronically ill patient, especially when that patient is your child. Karrie St. Clair began her quest when her baby girl, Katie, first became sick just three days after she was born in 1989. Hospitalized with a high fever, she tested negative for spinal meningitis and was released. But, three weeks later, she was hospitalized again.

Rocky Beginning

As the years went by with Katie constantly sick, Karrie found it extremely difficult to hold down a job. One time when she was a toddler, Katie once again ran a high fever. Because state law where they lived at the time mandates that a child with a fever higher than 100 degrees cannot be left at a licensed family homecare center, Karrie had to call in sick for three days. On day four, with Katie’s temperature lower, Karrie dropped her off at daycare and left for work. Later that morning, the daycare facility called to report Katie’s fever had returned and she needed to be picked up. At this point, Karrie’s exasperated boss gave her an ultimatum: “What is more important to you, Karrie, your job or your daughter?” Stunned, Karrie replied, “You’re a grandmother, I shouldn’t have to answer that.” Without hesitation, she left to pick up her daughter and didn’t return to her job.

About this time, Karrie’s home life came apart. Karrie and her husband, George, decided to go their separate ways, leaving Karrie a single mom on welfare caring for Katie. Going on welfare “wasn’t a choice,” explains Karrie. “It was survival. I didn’t like it, and it was very difficult trying to pay the rent, have enough food and buy clothing for a growing toddler. But you do it. You find the resources.” Despite these challenges, Karrie was grateful for the time she and Katie enjoyed together. Katie “was singing the ABC song with me when she was 3 years old ... and she got sick less when she stayed home with me.”

Turning Point

The turning point finally came when Katie was 13 years old and living with her father. Karrie received a call from George that Katie was very sick and had been admitted to a local hospital. She would be tested for mononucleosis, tuberculosis and every kind of well-known disease the doctors could think of, but still they were unable to reach a diagnosis. After a week of waiting for answers, Karrie had an aha! moment: “A little light bulb went off in my head. ... My daughter has been sick almost all of her life. What if she has an immune deficiency?”

After a week of waiting for answers,
light bulb went off in my head. ... My daughter has been sick almost all of her life. What if she has an immune deficiency?”

With this epiphany, Karrie went home, and despite having only minimal Internet skills at the time, decided to research the term “immune deficiency.” “I didn’t have to look and search long,” says Karrie. “My answer came up immediately with the Immune Deficiency Foundation (IDF) website. I browsed through the various disease types and kept going back to common variable immune deficiency (CVID), because it was exactly what Katie had experienced all of her life.” Armed with her newfound knowledge and half a ream of paper that explained CVID, Karrie gave the information to Katie’s father and her doctors. Katie was tested and confirmed to have CVID. When the doctors came back with the news, Karrie “felt blessed because now we had something to work with. Finally, we could hopefully make my daughter more comfortable, and not sick all of the time.” At 13 years old, Katie had a conclusive answer to her poor health and could begin treatments with intravenous immune globulin (IVIG).

Nonetheless, Karrie felt both relief and fear. She knew that CVID could be treated, but she also realized that Katie’s condition was very serious and the disease made her extremely fragile.

Advocate for Your Child

While working with Katie’s doctors wasn’t always easy, Karrie always remained a persistent advocate. Oftentimes, they told her: “Kids get sick; no big deal.” It was difficult for them to comprehend how a child could have a medical chart the size of a 50-year-old’s. Undaunted, Karrie continued to advocate for her daughter’s care. For example, when Katie was admitted to the hospital right before she was diagnosed, Karrie joined a conversation between George and a disease specialist as they discussed the possibility of giving Katie growth hormones because she was petite for her age. Karrie put an end to that very quickly by pointing out that she was a very slow developer herself, making it likely that Katie was following the same growth pattern. Learning this, the doctor completely agreed with her. Karrie notes: “It is very important for doctors to know both sides of the child’s medical history.”

Now 22 years old, Katie has been receiving IVIG infusions for nine years, with noticeably improved health. Since receiving therapy, she’s “capable of living a much fuller life, without being sick all of the time,” explains Karrie. “We have scares here and there, and sometimes she ends up in the hospital. But it’s not as often as it used to be, and now the doctors know what they are dealing with.” And to add icing to the cake, Katie gave birth to a sweet little girl in April, making Karrie a brand-new grandmother.

Karrie has some words of wisdom for parents who are going through similar situations with their children: “Hang in there! It’s not easy. … Know that you aren’t alone. Be your child’s advocate with your pediatrician or family doctor and bring awareness about PIDD. No child should have to wait 13 years before being diagnosed.

CARLA SCHICK is a staff writer for IG Living magazine.

If you have a life-transitioning story brought about with the help of IG, we want to share it with our readers. Email us at editor@IGLiving.com.
**Parenting:**

**Preparing Immune-Deficient Kids for High School**

PIDD kids face unique challenges in high school, but there are many strategies that they and their parents can use to ensure a successful high school experience.

By Mark T. Haggard

*ENTERING HIGH SCHOOL* can be the most exciting time in a child’s life. There are Friday night football games, pep rallies, dances, driver training — in short, acting like an adult but not having any of the responsibilities. Ah, to be 15 again. But primary immune deficiency disease (PIDD) kids carry a special burden with them because they must deal with unique issues that other students do not. These issues may leave them isolated in a community where the most active are the most well-received. But, there are strategies, for both students and parents, to help PIDD kids more quickly become part of the school community.

**Students: Be Knowledgeable**

Before freshman orientation, students should familiarize themselves with the school by perusing the high school’s website. There they will find pictures of the people on campus who will be important for the next four years of their lives. For instance, if there is a problem and going to parents does not seem to be an option, there are trained guidance counselors on campus. If the problem is in class, teachers are there to help them. If the issue is bullying, most high schools have an on-campus police officer, and school administrators are taking a much stronger line against bullying now than they have in the past.

When school starts, they should keep an organizer or a calendar to note due dates, meetings and school activities. For PIDD kids who miss school frequently, this will make things much easier. There are few things as impressive (and intimidating) for teachers as watching a student making notes in a day planner.

**Students: Know Your Group**

One of the challenges of growing up is finding out “who you are.” The sooner students can see themselves in the larger societal picture, the sooner they can begin to work toward success. No matter how much administrators try to break down barriers on campus, students are ultimately going to gravitate toward those with whom they share common interests. Not everyone
is a Ferris Bueller, a “righteous dude” who is “liked by everyone: motorheads, geeks, jocks, nerds, wasteroids, dweebies.” In fact, most students fall in with one group. And having a strong group of friends, a support system, will go a long way to helping students navigate the high school community. On the other hand, by trying to go it alone, they may find themselves at the mercy of some bad elements.

Students: Be Active!

Freshmen should get involved in activities on campus. This will start the process of building the friendships needed to get through high school. It also will afford PIDD kids something other than a disease to occupy their minds and bodies.

According to Lori Riggins, the mother of two children with adenosine deaminase severe combined immunodeficiency (ADA-SCID), involvement in school has enriched her kids’ lives. Both of Lori’s sons were active in high school, Rhett as part of his high school’s chapter of Future Farmers of America, and Zack playing drums for the high school band.

Sports are great outlets too. Zack ran cross-country in high school. Tyler Yates, diagnosed with common variable immune deficiency (CVID), was a soccer standout at his high school and parlayed that into college. Jordan Leventhal, another CVID patient, played three different sports while attending the American Hebrew Academy in Greensboro, N.C.

Unfortunately for PIDD kids, the benefit of being part of an athletic team is countered by the potential illness from even the slightest injury. Therefore, sports that minimize the repeated wounding of the body, no matter how slight, are better than those that do not. But, students must be allowed to do what they enjoy most. For instance, Tyler kept a first aid kit on the sideline of the soccer field at all times, and he wore elbow pads and knee pads to cushion any potentially injurious collision. Soccer was Tyler’s “sanctuary,” where he could forget about the troubles created by PIDD.

Another drawback for PIDD kids is that being active in the high school community can be a double-edged sword: healthy for their spirits but a drain on their energy. Practice after school, coupled with meetings before school or at every lunch period, can quickly run their bodies down. That’s why it’s important for these students to educate their team coaches and club advisors about their conditions and the fact that they may be sick occasionally. And, it is equally important for these students not to pack their schedule so tightly that they miss the rest that their bodies need to stay healthy. For instance, Arianna Kazemi, who has CVID, is just entering her freshman year of high school this year and her passion is dance, but she also has her sights set on college. According to Arianna, “My goal is to get As and high Bs.” To reach that goal and also dance five days a week after school, she is taking time off from cheerleading.

The most important class PIDD students can take in their high school careers is “Living with Immune Deficiency.”

Students: Don’t Be Defined by Your Immune Deficiency

Students should define themselves by something other than their immune deficiency. According to Mark Leventhal, Jordan’s father, “If you let it, CVID can become all-encompassing,” and it will become “emotionally debilitating.” Our children happen to have an immune deficiency, but that should not be their identity. They should be “talented,” be “awesome,” be a “baller” or be a “scholar.” Immune deficient should be way down on the list of things that describe them.

Parents: Communicate with School

Continual communication between parents and teachers and administrators is essential. As Lori Riggins explains: “We live in a small community and have gotten to know their teachers, administrators and health service personnel. We found everyone very willing to go the extra mile to keep us informed.” She became good friends with every school nurse and developed a system in which the nurse would call if there was an outbreak of any communicable diseases at school. “I was the expert regarding the boys’ disease,” she added, “and advocated many, many times, but we always had a very good response from all involved in the boys’ life.”

Annaben Kazemi, Arianna’s mother, agrees. She met with district leaders
last spring and planned to meet with Arianna’s teachers before the year started again this fall. During the first week of school, she intended to “pop in” to see how things were going.

Despite their knowledge and experience, educators still must be educated when it comes to PIDD kids. Arianna shares a notebook about CVID with her teachers. At the very least, PIDD kids should have an individualized health plan (per IDEA code 504) for the days that they will miss. In extreme cases, they have the right to an individualized education plan (IEP), a legal contract that allows a student academic accommodations from the school. (For more on IEPs see www.wrightslaw.com.)

Parents: Prepare Adolescents for Life

When asked about advice for parents preparing to let go of their children with PIDD, Mark Leventhal said that young adults need to be self-sufficient. The high school years are ideal for teaching children to prepare for the adult world. And, the way to prepare them is to not coddle them until the last minute. Instead, during what is likely the last four years with them, parents need to be training them to deal with the intricacies of their disorder. Adolescents need to learn to “know their bodies” — to watch for the signs of becoming sick so that they can immediately get treatment before a small sickness gets out of hand. Teens also should take charge for themselves, researching what is occurring inside their bodies, scheduling infusions and making their own medical appointments.

As with all challenges in life, parents should encourage their PIDD kids to be proactive rather than reactive so they can become resilient, independent adults. It doesn’t show up on their school schedule, but the most important class these students can take in their high school careers is “Living with Immune Deficiency.” Passing that class will prepare them for a long and productive life.

MARK T. HAGGARD is a high school teacher and football coach, and has three children, two of whom have CVID. He and his wife, Cheryl, also operate Under the Hood Ministries at www.underthehoodministries.org.
The Benefits of SCIG

By Annaben Kazemi

IT’S NOT UNUSUAL for patients to feel anxious when they are considering switching from intravenous immune globulin (IVIG) therapy to subcutaneous immune globulin (SCIG) therapy. However, for those patients who experience adverse events to IVIG or who develop venous access problems, SCIG is a viable option.

SCIG vs. IVIG

SCIG offers patients an alternative to IV infusion. It is self-administered, and patients infuse more often in smaller amounts, thereby stabilizing the highs and lows that patients feel between IVIG infusions. SCIG also is a convenient method because patients can set their own schedules for infusions at home; they are not affected by the time constraints of a healthcare provider’s schedule or limited by a clinic’s infusion hours. Because SCIG is administered under the skin using several sites and doesn’t require an IV, it eliminates IV placement problems, as well as lowers side effects. And, while patients often experience mild to moderate side effects of redness and soreness at the sites of infusion with SCIG, these effects can easily be dealt with by using compresses and creams.

Many studies have documented the safety and effectiveness of SCIG therapy, and they validate that SCIG is a therapeutic equivalent to IVIG.

Studies Demonstrate the Benefits of SCIG

Many studies have documented the safety and effectiveness of SCIG therapy, and they validate that SCIG is a therapeutic equivalent to IVIG.

In one study, 165 patients who switched from IVIG to SCIG experienced reduced systemic adverse events, as well as reduced cost. The researchers concluded that not only was SCIG convenient, safe and cost effective, but it could be successfully administered to patients who had previously experienced severe reactions.1

Another study, conducted to determine the efficacy and safety of SCIG therapy versus IVIG, concluded that SCIG therapy provides acceptable trough levels of IgG, a low incidence of side effects, efficacy similar to IVIG infusions, better health-related quality of life, higher levels of treatment satisfaction, and faster functional recovery with less time off from school and work.2

Finally, a third multinational study of 16 children and 44 adults that evaluated the safety and effectiveness of switching from IVIG to SCIG showed that high IgG levels were easily maintained with SCIG, which resulted in very good protection against infections. Out of a total of 2,297 administered infusions, 28 (1 percent) systemic adverse reactions occurred, and none was severe. The most common reaction was mild to moderate tissue-based reactions at the site that declined over time, usually within eight to 10 weeks.3

Matching SCIG to the Correct Patients

SCIG is not for all patients; however, it is an appropriate mode of therapy for particular patients. It improves quality of life, provides flexibility and results in stable IgG levels. Yet, while the research is clear that the benefits of SCIG therapy outweigh the risk, it is important to match this mode of therapy to the correct patients.

ANNABEN KAZEMI is the patient advocate for IG Living magazine.

References
EMED
The new Soft Glide SCIG infusion sets use coating technology to provide significantly less skin penetration force when inserting the needle and to maximize patient comfort throughout the infusion. Needles come in short (24 gauge) and long (27 gauge) sets to meet specific clinical requirements, and in 4mm, 6mm, 9mm, 12mm and 14mm lengths to accommodate all age groups and skin types. Soft translucent wings facilitate placement and patient comfort, and safety wings are available to encapsulate the needle upon removal. The sets allow for optimal flow performance with a wide range of fluid viscosities.  
(888) 550-6500; emedicaldevices.com

IntraPump
The newly FDA-approved needle set, called neria multi for large-volume infusions, has the benefit of a flexible finger grip for easy insertion, an adhesive with a window to view the site and a pre-attached adhesive, excluding the need for extra tape/Tegaderm.  
(866) 211-7867; www.neria.com/index.asp?pageid=H3513

MarCal Medical
MarCal’s Sub Q and Safety Sub Q right-angle infusion sets feature easier needle insertion and flexible wings for optimum viewing of insertion site; integrated wings on the needle to lay flat against the skin; and central position of the needle for stability and comfort. A variety of needle lengths and gauges are available (24 gauge, 27 gauge, 6mm, 9mm, 12mm and 14mm), and specialty gauges and needle lengths are available. Sets come with colored side clamps for easy identification for pull back on each site, as well as transparent dressing in a sterile package.  
(800) 628-9214; www.marcalmedical.com/subQsafetySubQ.htm

Norfolk Medical
The ClearView Sub-Q Standard Infusion Sets feature a 90-degree needle on a clear, flexible anchoring disk for easy visualization of the insertion site. The sets are suitable for most adults with adequate subcutaneous tissue and are offered in a variety of needle and tubing lengths. The ClearView Pediatric Sets are designed specifically for the smaller patient with less subcutaneous tissue. They feature a smaller, softer, one-half-inch clear anchoring disk and shorter 4mm needles. The Clearview-MS Needle Set is a multiple site subcutaneous delivery system with 4mm, 6mm, 9mm and 12mm needle lengths. All needles are 27 gauge unless special ordered, and all infusion and extension sets are individually packaged sterile. Custom sets are available.  
(847) 674-7075; www.norfolkm edical.com

RMS Medical Products
The RMS HlgH-Flo Safety Needle Sets are 26 gauge needle sets available in 6mm, 9mm and 12mm lengths for use in single, double, triple and quad configurations. The sets can be used with the company’s low-residual “Y” connector for up to an eight-needle configuration. They deliver maximum flow rate with less residual volume and distribute equal volumes to all needles in multi-needle sets. A closing feature safely secures the needle after use, and a sterile Tegaderm cover dressing by 3M is included.  
(800) 624-9600; www.rmsmedicalproducts.com
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Contact us to arrange a trial with our free sample program.
These organizations provide information about various disease states, which can be found by conducting a search of the disease state name.

- Advocacy for Patients with Chronic Illness: www.advocacyforpatients.org
- The Alliance for Biotherapeutics (fair access to plasma therapies): www.bioalliance.org
- American Autoimmune Related Diseases Association (AARDA): www.aarda.org
- American Chronic Pain Association (ACPA): www.theacpa.org
- Band-Aides and Blackboards: www.lehman.cuny.edu/faculty/jfleitas/bandaides
- eMedicine from WebMD: emedicine.medscape.com
- FamilyDoctor.org: www.familydoctor.org
- Johns Hopkins Medicine: www.hopkinsmedicine.org
- KeepKidsHealthy.com (pediatrician’s guide to children health and safety): www.keepkidshealthy.com
- Mayo Clinic: www.mayoclinic.com
- National Committee for Quality Assurance (detailed report cards on health plans, clinical performance, member satisfaction and access to care): www.ncqa.org
- National Heart, Lung and Blood Institute: www.nhlbi.nih.gov/health/health-topics/by-alpha
- National Institutes of Health: health.nih.gov/see-all-topics.aspx
- National Organization for Rare Disorders (disease-specific support groups and virtual communities for patients and caregivers): www.rarediseases.org
- Office of Rare Diseases Research: rarediseases.info.nih.gov
- Patient Advocate Foundation (patient access to care, maintenance of employment and financial stability): www.patientadvocate.org
- WebMD (medical reference): www.webmd.com

**IG MANUFACTURER WEBSITES**
- Baxter: www.baxter.com
- Bio Products Laboratory: www.gampmaplex.com
- CSL Behring: www.cslbehring.com
- Grifols: www.grifolsusa.com
- Kedrion: www.kedrionusa.com
- Octapharma: www.octapharma.com

**Disease-State Resources**

### Ataxia Telangiectasia (A-T)

**WEBSITES**
- A-T Children’s Project: www.atcp.org

### Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

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

### Evans Syndrome

**ONLINE PEER SUPPORT**
- Evans Syndrome Research and Support Group: www.evanssyndrome.org

### Guillain-Barré Syndrome (GBS)

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

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

### Idiopathic Thrombocytopenic Purpura (ITP)

**WEBSITES**
- ITP Support Association — UK: www.itpsupport.org.uk
- Platelet Disorder Support Association: www.pdsa.org

### Kawasaki Disease

**WEBSITES**
- American Heart Association: www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsOfChildhood/Kawasaki-Disease_UCM_308777_Article.jsp#.T1T2boePWEO
- Kawasaki Disease Foundation: www.kdfoundation.org

### Mitochondrial Disease

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

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Sources

For a more comprehensive list of resources, visit the Resources page at www.IGLiving.com.
Multifocal Motor Neuropathy (MMN)

WEBSITES
- Neuromuscular Disease Center at Washington University: neuromuscular.wustl.edu
- The Neuropathy Association: www.neuropathy.org

Multiple Sclerosis (MS)

WEBSITES
- All About Multiple Sclerosis: www.mult-sclerosis.org/index.html
- Multiple Sclerosis Association of America: www.msaa.com
- 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
- International Myositis Assessment and Clinical Studies Group: www.niehs.nih.gov/research/resources/collab/imacs/main.cfm
- The Myositis Association, www.myositis.org, is devoted exclusively to all types of myositis, which affects upwards of 20 million Americans. The Association’s mission is to increase public awareness of the nature and extent of PN, facilitate information exchanges about the disease, and advocate the need for early intervention and support research into the causes and treatment of neuromyopathies. (202) 887-0088

Online Peer Support

Peripheral Neuropathy (PN)

WEBSITES
- Neuropathy Action Foundation: www.neuropathyaction.org
- Calgary Neuropathy Association: www.calgaryneuropathy.com
- Texas Chapter of the Neuropathy Association: www.handsfeetheart.org

Primary Immune Deficiency Disease (PIDD)

WEBSITES
- The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/health/topics/Primary_Immunodeficiency.cfm
- American Academy of Allergy, Asthma & Immunology: www.aaaai.org
- International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: www.ipopi.org
- New England Primary Immunodeficiency Network: www.nepin.org
- Rainbow Allergy-Immunology: www.uhhospitals.org/rainbow/services/allergy-immunology
- Team Hope (for families and patients in New England): www.teammhope.info

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


The Jeffrey Modell Foundation, www.info4pi.org, is dedicated to early and precise diagnosis, meaningful treatments and, ultimately, cures for primary immunodeficiency. (212) 819-0200
**ONLINE PEER SUPPORT**
- IDF Common Ground: www.idfcommonground.org
- IDF Discussion Forum: http://idffriends.org/forum
- IDF Friends: http://idffriends.org
- Jeffrey Modell Foundation Message Board: www.info4pi.org
- Michigan Immunodeficiency Foundation: www.facebook.com/groups/108048062584350
- Rhode Island peer group: health.groups.yahoo.com/group/RhodeIslandPIDD

**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 foraums/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.livingwithspss.com
- Stiff Person Syndrome: www.stiffpersonsindrome.net

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**EDUCATION AND DISABILITY RESOURCES**
- Americans with Disabilities Act of 1990: www.ada.gov Provides protection for people with disabilities from certain types of discrimination, and requires employers to provide some accommodations of the disability.
- Individuals with Disabilities Education Improvement Act of 2004: idea.ed.gov/explore/home
- National Disability Rights Network: www.ndrn.org This website offers a search tool to find resources in your state to assist with school rights and advocacy.
- Social Security: www.ssa.gov/disability
- U.S. Department of Education Website: www2.ed.gov/parents/landing.jhtm l?exp=4 This federal government website offers a parents section titled “My Child’s Special Needs.”

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

**FOOD ALLERGIES**
- Allergic Disorders: Promoting Best Practice: www.aaaai.org
- American Partnership for Eosinophilic Disorders: www.apfed.org
- Food Allergy and Anaphylaxis Network: www.foodallergy.org
- World Allergy Organization: www.worldallergy.org

**PRODUCT INFORMATION**
- Influenza and the influenza vaccine: www.cdc.gov/flu or call (800) CDC-INFO: (800) 232-4636
- IVIG Flebogamma 5% DIF and 10% DIF: www.grifols.com/portal/en/US/bioscience?div=8883
- IVIG/SCIG Gammagard Liquid: www.gammagardliquid.com
- IVIG Gammagard S/D: www.baxter.com/patients_and_caregivers/products/gammagard_sd_5.html
- IVIG/SCIG Gammaked: www.gammaked.com
- IVIG Gammaplex: www.gammaplex.com
- IVIG Privigen: www.privigen.com
- SCIG Hizentra: www.hizentra.com

**PUMP AND INFUSION SETS WEBSITES**
- EMED Corporation: www.safetymedicalproducts.com
- Marcal Medical Inc.: www.marcalmedical.com
- Intrapump Infusion Systems: www.intrapump.com
- Micrel Medical Devices: www.micrelmed.com
- Norfolk Medical: www.norfolkmedical.com
- RMS Medical Products: www.rmsmedicalproducts.com
- Smith Medical: www.smiths-medical.com/brands/cadd

Have something to add to these pages? Please send your suggestions for additions to the IG Living Resource Directory to editor@IGLiving.com.
The **Products you need when you need them.**

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- Coagulation Products
- Hyperimmunes
- Albumin
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