PIDD Patients
The Different Faces of the Disease

SCIG Needle Anxiety: How to Overcome the Fear

Pemphigus & Pemphigoid: Rare Autoimmune Diseases

Understanding Advance Health Care Directives

Parenting: Teaching Children to Care for their Health
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.
- Hyperproteinemnia, with resultant changes in serum viscosity and electrolyte imbalances may occur in patients receiving IGIV therapy.
- 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.

ADVERSE REACTIONS
- PI - The most common adverse reactions (=5%) with intravenous use of GAMUNEX-C were headache, cough, injection site reaction, nausea, pharyngitis and urticaria. The most common adverse reactions (=5%) with subcutaneous use of GAMUNEX-C were infusion site reactions, headache, fatigue, arthralgia and pyrexia.
- ITP - The most common adverse reactions during clinical trials (reported in =5% of subjects) were headache, vomiting, fever, nausea, back pain and rash.
- CIDP - The most common adverse reactions during clinical trials (reported in =5% of subjects) were headache, fever, chills, hypertension, rash, nausea and asthenia.

To report SUSPECTED ADVERSE REACTIONS, contact Talecris Biotherapeutics, Inc. at 1-800-520-2807 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

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.

Talecris Biotherapeutics, Inc.
Research Triangle Park, NC 27709 USA
U.S. License No. 1716
08939771/08939782-BS
Revised: October 2010
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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AMY EHLERS, BS, PHARMD, BCPS
Director of Pharmacy, NuFACTOR Specialty Pharmacy

Overcoming SCIG Needle Anxiety
“Needle anxiety is an understandable concern for patients new to SCIG therapy, and discomfort sometimes can be experienced.”

ARIANNA KAZEMI
IG Teen Patient

Teen Talk: The Disease that Makes “Me” Me
“My disease has made me a stronger, more confident individual, but it also has its downsides.”

MICHELLE GREER, RN
Vice President of Sales, NuFACTOR Specialty Pharmacy

Understanding Pemphigus and Pemphigoid
“It is important to raise awareness about these autoimmune diseases so that earlier detection and intervention can be possible.”

ANNABEN KAZEMI
IG Living’s Patient Advocate

A Day in the Life of PIDD Patients
“While a PIDD shouldn’t keep patients from living a normal life, the degree to which their illness affects their lifestyle does vary widely.”

DENNA MCGREW
Professional Children’s Advocate

IG Chronicles: Here’s How You Can Help
“It seems easier for loved ones to rally around an acute and time-limited problem, but harder to wrap their arms around the fact that chronic illness is the family’s reality every day.”

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

IG Living isn’t just a magazine; it’s an interactive community of people with interesting stories.

Our blog: www.igliving.com/blogengine
Our Facebook page: www.Facebook.com/IGLivingMagazine

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.

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

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

Trying to Cover It “All”

Seven years ago, IG Living magazine was created as a resource to help patients treated with immune globulin (IG) and your caretakers better understand the complex diseases you deal with. The goal was — and still is — to empower you with solutions to the questions you raise, to share your stories about coping with chronic illness, and to connect you with others in similar situations.

Each issue, it’s a challenge to decide which topics to cover, but even more so how to cover them. The reason: Each and every one of your experiences is different — from the stage of your illness, to the way in which you respond to treatment, to insurance issues you face, and even the quality of your support relationships.

Having a connection with so many patients in the IG community through our magazine, website, Facebook page and blog, we see this disparity very clearly — from the calls and emails we receive from you seeking help, to comments posted in response to our articles, to the posts on our Facebook page and our weekly blogs, and more. What we also see is that it’s hard for many of you to understand why you may experience so many more, or so fewer, difficulties compared with others faced with the same or a very similar illness.

To highlight the variety of patient experience, our article A Day in the Life of PIDD Patients takes a look at the lives of three individuals diagnosed with a primary immunodeficiency and the different ways they are coping. As our patient advocate, Annaben Kazemi, points out: “There are no typical PIDD patients, and PIDD patients seem to have no typical days.” There are simply too many variances in the way in which PIDD presents in patients for a comparison to be made between one patient and another. You may see yourself in one of these individuals, or you may see yourself somewhere in between.

Also in this issue, we focus on one aspect of subcutaneous IG (SCIG) therapy. While not all of you may be considering SCIG therapy, some may be grappling with the fear of having to insert your own needles versus having them inserted by a nurse. In our article Overcoming SCIG Needle Anxiety, Pharmacist Amy Ehlers discusses the ways this apprehension can be minimized or even overcome.

The diseases featured in this issue are the rare and difficult-to-diagnose autoimmune disorders pemphigus and pemphigoid, which affect less than 200,000 people in the U.S. and are effectively treated by high doses of IVIG. Our goal is to raise awareness about them in hopes of shortening the time to diagnose, which can avert permanent complications.

There are so many complexities associated with IG-treated illnesses that affect all of you differently, so it’s impossible for all of our articles to be helpful to everyone. But, eventually, we want to cover it “all.” So, if you’re interested in having us write about something specific in IG Living’s magazine, website or social media platforms that we have yet to address, please let us know by emailing us at editor@IGLiving.com.

Ronale Tucker Rhodes, MS, Editor
Enter IG Living’s Essay Contest!

**IG Living is hosting its third annual essay contest open to IG patients and their caregivers ages 18 and older.**

### How to Submit an Essay
- Start your essay with this phrase: “My life turned around and improved when…”
- Write no more than 600 words
- Type and double space your entry
- Include an essay title, your name, complete address, email, phone number and word count
- Submit your entry electronically as a Microsoft Word attachment to editor@IGLiving.com, or submit it by mail to IG Living Essay Contest, 41093 County Center Drive, Temecula, CA 92591, Attention: Carla Schick
- Mail your entry by June 1, 2013 (must be postmarked by that date)

### How Your Essay Will Be Scored
*IG Living’s* judges will rate the entries on a scale of one to 10 on five criteria:
- Organization (the writing flows logically with clear structure)
- Mechanics (spelling, capitalization and punctuation are correct)
- Content (subject is discussed clearly, and the reader is left with a finished feeling)
- Creativity (content is compellingly interesting for our audience)
- Effectiveness (the whole entry is effective in its purpose for our audience)

### Winners Announced on July 1st!

- **1st Place:** iPad Mini
  - essay published in IG Living magazine
- **2nd Place:** $50 gift card
  - essay published on the IG Living blog
- **3rd Place:** $50 gift card
  - essay published on the IG Living blog

### Sample of 2011 Winner’s Essay
If I knew then what I know now, I would have … not changed a thing. A lesson I learned in college about existentialism still resonates with me after all these years. One of philosopher Jean-Paul Sartre’s beliefs is that all the events that have happened in our lives, good or bad, have brought us to this moment. If we can say we are happy now for one moment, why would we want to change all the events that led up to that moment’s joy?

— Stacy Oliver

### Sample of 2012 Winner’s Essay
The last time I did something for the first time, I … walked slowly down the ramp into the 90-degree therapy pool for my first strengthening and toning class. As I moved deeper into the water, I was amazed at how soothing it felt. I thought: Why on earth did it take me so long to get here?

— Patricia Carroll

*Read the full text of the previous years’ winners essays in the August-September 2011 and 2012 issues in the IG Living magazine archives: www.igliving.com/ArchivedIssues.aspx*

**More information:** Editor@IGLiving.com
AN APPROXIMATELY 3-year-old boy was referred for evaluation of possible immunodeficiency. In the previous month, the child had more than 30 visits with his primary care physician for fevers and respiratory symptoms. It was during the winter months, and despite the number of visits, his primary care physician did not feel the child was too ill. Rather, he thought the child’s viral illness was one continuous illness that wasn’t improving.

Pertinent features of the child’s history are: 1) He began having recurrent respiratory illnesses between 3 months and 6 months of age. This is the time frame during which the maternal antibodies that pass through the placenta into the fetus during the last trimester of the pregnancy disappear from infants, creating a loss of protective maternal antibodies. This loss causes all infants to become susceptible to infections (in a certain context, immunodeficient) as they begin to acquire their own immunity against infectious organisms. On average, it is expected that all infants and young children will become ill with approximately six respiratory infections per year (generally worse through the winter months) until their immunity matures. By 6 years to 12 years of age, the immune system usually has matured to the point that fewer respiratory infections occur.

2) The child is reported to have been given courses of antibiotics every four to eight weeks. At 3 years of age, he had received approximately 15 courses of antibiotics. Therefore, the child had been taking antibiotics for seven or eight months of his 36 months of life. In persons with normally functioning immunity, most infections are due to viral illnesses and, therefore, will not benefit from an antibacterial antibiotic. But in some people, especially those with antibody deficiencies, the viral illness will injure the tissue in the respiratory passages and result in swelling of the tissues. This, in turn, may block the normal mucous drainage passages from the sinuses and/or middle ears,
thereby trapping bacteria, which may result in a secondary bacterial infection for which antibiotics may be required. However, in many cases, adequate drainage can be of great benefit. For example, the placement of ear ventilation tubes to allow drainage from the middle ear can resolve many cases of recurrent ear infections.

The swelling of the respiratory tissues can be compounded in many ways. Unfortunately, one of the more common reasons for respiratory tissue irritation is exposure to cigarette smoke. Due to the increased respiratory rate of infants, their close proximity to a smoker (typically being held by the smoker) may result in them actually smoking more than the person holding them. Even exposure to the smoker’s clothing or the car in which someone has smoked can be the equivalent of infants actually smoking several cigarettes. Allergies are another irritant of respiratory tissues. A greatly underappreciated irritation occurs from acid reflux from infants’ stomachs. A great portion of infants’ lives is spent reclining. And because the stomach contents readily flow through the esophagus into the upper respiratory tract, mouth, ears and sinuses, this can result in irritation of the respiratory tissues and contribute to bacterial infections of the sinuses and ears.

This child was not being exposed to cigarette smoke, but evaluations for allergies and acid reflux were to be included in his workup.

Next time, we will continue with the patient’s history and evaluation for possible antibody deficiency.

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

Disclaimer: This case reported is intended as an illustration for education purposes only. It does not necessarily represent an individual or precise information from patient files.
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Sorrento Therapeutics Acquires Rights to Production of rIVIG

Sorrento Therapeutics Inc., a development-stage biopharmaceutical company, has acquired the rights to apply its G-MAB antibody library technology to produce recombinant intravenous immune globulin (rIVIG). Currently, IVIG preparations are derived from human donor blood products pooled from more than 1,000 healthy individuals. However, increasing demand for IVIG products already exceeds available supply worldwide, limiting broader clinical applications. According to the company, the use of Sorrento’s proprietary G-MAB library technology for the production of rIVIG may circumvent many disadvantages and obstacles currently encumbering IVIG preparations, such as potential supply constraint, costly production due to fractionation, and purification and quality control challenges. Potential advantages of rIVIG include reduced batch-to-batch inconsistencies, minimal contamination risk, increased potency and no need for continued human blood donor supply.

“Our acquisition of the proprietary rights to the rIVIG technology, we now have access to a multibillion-dollar market opportunity applying our G-MAB antibody libraries to address unmet medical needs,” said Henry Ji, president and CEO. In addition, says Tien Lee, MD, one of the co-inventors of the rIVIG technology, “Partnering our rIVIG invention with the unique G-MAB technology may produce novel and highly differentiated IVIG products.”

Neuropathy Action Awareness Day Scheduled for Los Angeles

The 7th annual Neuropathy Action Awareness Day is scheduled for June 20 in Los Angeles. This is the first time in the event’s six-year history that the event will be held in Southern California rather than Northern California. Hosted by the Neuropathy Action Foundation, the event is expected to attract more than 300 neuropathy patients, family members, healthcare providers and others to learn about neuropathy, policy issues and patient advocacy.

Throughout the day, there will be educational sessions and an exhibit area; there also will be a sit-down luncheon, as well as a celebrity speaker, elected officials and others. Educational sessions include Partnering with Your Doctor; Making Sense of Medicine; Diet, Health and Nutrition; The Whole Body Experience; The Human Element; Neuropathic Pain and Pain Treatments; Traumatic Neuropathies and Nerve Repair; and more. The exhibit hall will feature companies, organizations and government entities that will educate neuropathy patients about their services and products — from understanding insurance and the rights to appeal decisions to learning how to alleviate neuropathic pain.

For more information about the event and to obtain a registration brochure, visit the Neuropathy Action Foundation website at www.neuropathyaction.org.
A team of researchers at the Garvan Institute of Medical Research in Sydney, Australia, has identified a weak link in the immune system and “the exact conditions under which an infection can trigger an autoantibody response.” According to the researchers, who specialize in the study of how immune B cells produce self-attacking rogue antibodies, their finding “explains a lot about how autoimmune conditions that target particular organs such as the heart or nervous system could develop after an infection.”

Immune cells go through processes when they are first formed that ensure they are able to identify self and, therefore, avoid self-attack. But the antibody-creating B cells go through a second phase of development when the body is engaged in trying to fend off disease or infection. To cope with the immeasurable range of microbes in our environment, B cells have evolved the ability to mutate their antibody genes randomly until they produce one that sticks strongly to the invader. At that point, the successful B cells proliferate and flood the system with these new antibodies. This “high affinity antibody” generation process occurs rapidly within specialized environments in the lymph system known as germinal centers. Most of the time, germinal centers help fight disease and build up a protective armory for the future. But the urgency and speed at which B cells mutate within the germinal center, as well as the random nature of the process, sometimes create an antibody that also happens to match self and has the potential to cause autoimmune activity.

The researchers, who developed sophisticated mouse models to investigate when and how this happens, found that when the invading antigen is abundant and generally present throughout the body, rogue autoantibody-generating B cells are deleted and autoimmunity is avoided. But when the target antigen is located only in a tissue or organ remote from the germinal center, B cells capable of reacting against both antigen and self are able to escape the germinal center and produce autoantibodies. Essentially, the researchers say, they’ve shown there’s a hole in self-tolerance when it comes to cross-reactive autoantibodies that can attack organ-specific targets. Their findings suggest that if enough is known about the disease and the molecular messaging systems involved, it may be possible in the future to modulate the germinal center response. They plan to continue to use their new mouse model to study the various molecular reactions involved in the progression of an autoimmune response.

The study was published in the Nov. 8 edition of the journal *Immunology*. ❘

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**Research**

**First Linked Fore a Cure Benefits AARDA**

The first Linked Fore a Cure benefit for the American Autoimmune Related Diseases Association (AARDA) held to raise awareness for autoimmune diseases raised $38,000. The event, which was held at the Water’s Edge Golf Club in Worth, Ill., included a four-person scramble, lunch, cocktails and hors d’oeuvres, as well as a silent auction, raffle and awards ceremony. “We hope to spread awareness of autoimmune diseases and raise research funds to bring us closer to a cure,” said AARDA board member and treasurer John Kaiser. “We hope that these actions are the beginning of annual collaborations with AARDA to raise research funding aimed at eradicating autoimmune diseases.” ❘
Research

Merck Doubles Investment in Autoimmune Therapy Research

Merck has promised $600 million in milestones to buy into Lycera’s development work on small molecules that target T-helper 17 (Th17) cells, which the company believes offers a new, first-in-class approach to treating major autoimmune diseases such as rheumatoid arthritis, psoriasis, inflammatory bowel disease and multiple sclerosis. This amount is double the company’s original agreement of up to $295 million.

Merck was founded on the work that Gary Glick — Lycera’s CSO — did at the University of Michigan. Based on some original animal studies, Glick believes that inhibiting the retinoic acid-related orphan receptor (RORγt), the company can take the Th17 path to reining in IL-17, a well-known player in autoimmune ailments.

Lycera is following a different path for its in-house work on autoimmune diseases, according to CEO Kathleen Metters. The company should move into the clinic with its own candidate in the next 12 to 18 months. 

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Did You Know

Advance Health Care Directives
By Trudie Mitschang

There are two universal truths about death. The first is that everyone is going to die. The second is that no one really behaves as if they believe it will happen to them. Whether someone is young and healthy or living with the challenges of chronic disease, they are probably reluctant to initiate a conversation about end-of-life care with loved ones. While broaching the subject can be emotionally uncomfortable, clarifying end-of-life choices today can significantly ease the burden on family members who may later be called upon to make decisions on the deceased’s behalf. Discussing one’s wishes with loved ones is an important first step, but the second and most significant step is preparing a document called an advance health care directive (AHCD). An AHCD offers the best assurance that decisions regarding future medical care will reflect a person’s personal values and beliefs.

What Is an Advance Health Care Directive (AHCD)?
The AHCD informs the physician, family and friends of one’s healthcare preferences, including the types of special treatment wanted or not wanted at the end of life. It also may include the desire for diagnostic testing, surgical procedures, cardiopulmonary resuscitation and organ donation. By considering options early, individuals can ensure the quality of life that is important to them and avoid having their family “guess” their wishes or make critical medical care decisions for them under stress or in emotional turmoil.

Why an AHCD Is Important
In an article featured in the San Jose Mercury News, writer Lisa Krieger shared the story of her father’s final days, and the resulting financial burden his hospital stay put on the family. She suggests that planning for the end of life should begin much sooner than it typically does. “For me, planning should have begun the day my dad got his Alzheimer’s diagnosis, four years before his death,” Krieger says. “Certainly when he broke his hip. By the time we got to the ER, trying to combat an infection in my frail 88-year-old dad, it was far too late.”

Krieger’s family ended up owing $323,000 for unnecessary procedures performed during a 10-day hospital stay that did not ultimately extend her father’s life. Sadly, her experience is not unique. In a perfect world, everyone would die peacefully at home surrounded by loved ones; unfortunately, that is not always possible. For the terminally ill or elderly patient who dies in a hospital setting
as Krieger’s father did, an AHCD is essential so that family members do not become embroiled in emotionally charged disputes with hospital physicians and staff regarding the best treatment options. Decisions like this made under duress take precious time away from families wanting to share final hours with their loved one. Many families have learned the hard way that it is virtually impossible to discern a dying person’s wishes unless the topic has been discussed ahead of time and those wishes are in writing.

The Physician’s Involvement
The conversation about end-of-life directives should begin with family members but should also include the individual’s primary care physician. The physician, likely to be the one caring for them when their instructions become relevant, is much more inclined to honor requests that have been clearly communicated. The physician can:

- Help phrase requests in a way that makes sense to medical professionals
- Point out any illogical requests; sometimes refusing one kind of treatment makes it illogical to pursue another kind of treatment
- Advise if there are aspects of their requests that the physician cannot honor because of personal, moral or professional constraints

Creating an AHCD
AHCDs are not complicated, but the content can be complex and should be thought through very carefully. In the U.S., most state governments have designed forms individuals can complete on their own by filling in the blanks. Keep in mind most states do require witnessing or other specific signing formalities. While legal advice to prepare an AHCD is not required, it may be a good idea to ensure that the actual instructions for one’s wishes are stated accurately.

What to Include in an AHCD²
- Who should make decisions for the individual if they are unable to make their own (including financial matters and healthcare decisions)
- What medical treatments and care are acceptable to them and which ones are not
- Opinions regarding artificial nutrition and hydration and any other measures that prolong life
- The wish to be or not to be resuscitated if they stop breathing and/or their heart stops
- The desire to be hospitalized or stay at home, or somewhere else, if they are seriously or terminally ill
- How care will be paid for (is there adequate insurance?)
- What might have been overlooked that will be costly at a time when loved ones are distracted by grieving over the person’s condition or death

Planning Ahead
Planning ahead with an AHCD can give caregivers, family members and other loved ones peace of mind when it comes to making decisions about a person’s future health care. It lets everyone know what is important to the patient, and what is not. When loved ones are clear about preferences for treatment, they’re free to devote their energy to what matters most: making the most of precious time with the one who is dying.

TRUDIE MITSCHANG is a staff writer for IG Living magazine.

Reference
Anxiety and discomfort are common occurrences when transitioning to SCIG therapy, but these issues can be minimized or resolved.

By Amy Ehlers, BS, PharmD, BCPS
When considering the transition from intravenous immune globulin (IVIG) therapy to subcutaneous IG (SCIG) therapy, patients commonly experience needle anxiety. With IVIG infusions, patients may have used a port or have had a skilled infusion nurse who was able to start a peripheral IV quickly, easily and with minimal discomfort. But to obtain the benefits of SCIG, patients need to be responsible for placing their own needles, the number of which can range anywhere from one to six. Considering this, patients often question if SCIG is right for them. Yet, while anxiety about needles and any discomfort that may arise is common, there are multiple options to address these concerns to allow for a successful conversion from IVIG to SCIG therapy.

Talking About Anxiety
First and most importantly, patients need to have an open dialogue with their healthcare team about their concerns regarding needles. Even when initiated and desired by patients, transitioning to SCIG therapy can be overwhelming. For some, being able to talk through their apprehensions may be helpful. Patients who are stressed, anxious and fearful may be more likely to experience pain than those who have a relaxed and positive outlook. And, with children, it may be a little more of a challenge. If children are old enough, allowing them to touch and handle the supplies should be considered. Some of their fears may be eased when they discover an SCIG needle is different from a peripheral or port needle. SCIG needles are of a higher gauge (27 gauge), which is thinner or finer than a peripheral or port needle (for example, a 22 gauge or 24 gauge). They also are shorter in length, which often results in less discomfort upon insertion. Caregivers of children should consider using a proven distraction when possible and try to not allow their own fears to transfer to children. If caregivers don’t appear anxious or concerned, children’s anxiety levels likely will also be low.

Learning Proper Technique
Proper needle placement technique is critical for minimizing needle discomfort. Patients transitioning to SCIG often are scheduled for three to four teaching and training visits during which they will learn about needle placement, among other things. Learning to place the needle swiftly and at the appropriate angle will help ensure the needle goes into the desired subcutaneous space. It also is important to allow the antiseptic skin prep to dry, especially if an alcohol pad is used. If the skin is still wet with alcohol when the needle penetrates, there is a higher likelihood of stinging and burning.

If needle technique assessment is correct and other external factors have been addressed, using a topical skin anesthetic such as EMLA cream (lidocaine 2.5%/prilocaine 2.5%) is a standard first-line therapy to medically manage needle discomfort. To receive maximum benefit, it is important to use the correct method. Patients should apply the cream at least one hour prior to needle placement. While some numbing effect may be noticed as early as 15 minutes after its application, this is typically only the surface layer of the skin; it takes time for the medication to reach the underlying tissues. In most patients, the level of analgesia achieved at one hour is sufficient to place the SCIG needles without pain. If this is not the case, patients should attempt leaving the cream on longer; the maximum effect of EMLA is usually seen two to three hours after its application.

A thick layer of cream should be applied to the planned needle sites, and it should not be rubbed in. The area(s) with an occlusive dressing such as Tegaderm should be covered to allow for maximum penetration of the medication. Just prior to inserting the needles, the occlusive dressing is removed, the cream is wiped with a gauze pad, and the area is cleaned with an antiseptic skin prep. By following these steps, the most benefit from the analgesia will be had, since the effects last approximately one to two hours after the cream is removed from the skin.

If patients are unable to use a topical anesthetic cream, another option is to apply ice to the infusion site prior to needle insertion. The drawback to icing the area is that it may slow the absorption of the medication into the subcutaneous tissue.
Managing Infusion Effects

Needle anxiety also may develop over time due to the discomfort or irritation that occurs during the SCIG infusion. Again, patients should be encouraged to have open and prompt communications with their pharmacist or physician to discuss the specific concerns and possible solutions. Assessing and adjusting the SCIG needle length, the specific sites of infusion and the total number of infusion sites can minimize or eliminate needle discomfort.

SCIG needles are available in several lengths: 4 mm, 6 mm, 9 mm and 12 mm. The longer the needle, the deeper into the skin and subcutaneous tissue it is able to go. The length of the needle should be long enough to reach the subcutaneous tissue but not so long as to reach and irritate muscle or nerves. The size of the patient and preferred infusion sites often are the biggest influences on needle size. For example, a patient who is 5 feet 4 inches, weighs 160 pounds and prefers to infuse in the abdominal area needs a longer needle than a patient who is 6 feet 2 inches, weighs 160 pounds and prefers to infuse in the upper arms. The first patient would benefit from a longer needle (9 mm or 12 mm) because the subcutaneous layer is thicker in the abdominal area and the shorter needle may irritate the intradermal layer of the skin, in addition to causing some site leakage. The second patient would not benefit from these longer needle lengths because the patient has a leaner build and a 9 mm or 12 mm SCIG needle has a higher risk of entering muscle or nerve tissue in the upper arms.

Needle anxiety also may develop over time due to the discomfort or irritation that occurs during the SCIG infusion.

Patients are typically able to infuse in almost any place they can “pinch an inch,” with common areas being the abdomen, hips, thighs and upper arms. The more subcutaneous tissue that is available, the less the risk of needle discomfort. Care should be taken to avoid areas with scars, stretch marks or other types of skin breakdown. When selecting sites in the abdominal area, a comfortable distance should be kept from the belly button. Spacing needles at least two inches apart and rotating sites also are important. Patients sometimes use a birthmark, mole or other identifying feature on the skin for a reference point and unknowingly use the same sites infusion after infusion. Over time, this may cause the subcutaneous tissue to become irritated or damaged.

The total number of infusion sites also should be considered. Patients should use the number of sites that allow the best absorption of the SCIG medication in the desired infusion time. This is highly patient-specific and may take several weeks or months of infusions to determine. It is best to start with the recommended number of sites determined by the healthcare team and then adjust up or down based on response.

Ensuring SCIG Success

Needle anxiety is an understandable concern for patients new to SCIG therapy, and discomfort sometimes can be experienced. What is important for patients to know is that with open and honest conversations with their physician, nurse or pharmacist, many if not all issues can be resolved or minimized. Addressing these concerns either initially or as they occur will allow patients the best chance for success to enjoy the benefits of using SCIG therapy to manage their disease.

AMY EHLERS, BS, PharmD, BCPS, is the director of pharmacy at NuFACTOR Specialty Pharmacy. She has a bachelor’s and doctorate degree in pharmacy, and is board certified in pharmacotherapy.
If you are a Hizentra patient or caregiver
Get connected through 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).

Find out what a Voice2Voice advocate can do for you.

A Voice2Voice advocate is someone you can share your story with. They can help answer your non-medical questions* and connect you to helpful resources. In addition, Voice2Voice advocates can share their own real-life treatment stories and offer encouragement as only someone who’s “been there” can do.

*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

Voice 2 Voice™

Sign up for Voice2Voice.
Log on to Hizentra.com/V2V to get connected with an experienced Voice2Voice advocate for helpful peer-to-peer support.

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, or call 1-800-FDA-1088.

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

**Initial U.S. Approval: 2010**

**INFORMATION FOR PATIENTS**

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

-----------------------------------INDICATIONS AND USAGE-----------------------------------

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

**INFORMATION FOR PATIENTS**

This patient package insert summarizes important information about Hizentra. Please read it carefully before using this medicine. This information does not take the place of talking with your healthcare professional, and it does not include all of the important information about Hizentra. If you have any questions after reading this, ask your healthcare professional.

**What is the most important information I should know about Hizentra?**

Hizentra is supposed to be infused under your skin only. DO NOT inject Hizentra into a blood vessel or vein.

Do not take Hizentra if you have too much proline in your blood (called “hyperprolinemia”) or if you have had reactions to polysorbate 80. Tell your doctor if you have had a serious reaction to other immune globulin medicines or if you have been told that you also have a deficiency of the immunoglobulin called IgA. Tell your doctor if you have a history of heart or blood vessel disease or blood clots, have thick blood, or have been immobile for some time. These things may increase your risk of having a blood clot after using Hizentra. Also tell your doctor what drugs you are using, as some drugs, such as those that contain the hormone estrogen (for example, birth control pills), may increase your risk of developing a blood clot.

**How should I take Hizentra?**

You will take Hizentra through an infusion, only under your skin. Make sure that the infusion is not into a blood vessel. You will place up to 4 needles into different areas of your body each time you use Hizentra. The needles are attached to a pump with an infusion tube. It usually takes about 60 minutes to do one infusion. You will need to have infusions once a week.

Do not use Hizentra by yourself until you have been taught how by your doctor or healthcare professional.

**What should I avoid while taking Hizentra?**

Vaccines may not work well for you while you are taking Hizentra. Tell your doctor or healthcare professional that you are taking Hizentra before you get a vaccine. Tell your doctor or healthcare professional if you are pregnant or plan to become pregnant, or if you are nursing.

**What are possible side effects of Hizentra?**

The most common side effects with Hizentra are:

- Redness, swelling, itching, and/or bruising at the injection site
- Headache/migraine
- Nausea and/or vomiting
- Pain (including pain in the chest, back, joints, arms, legs)
- Fatigue
- Diarrhea
- Stomach ache/bloating
- Cough
- Rash (including hives)
- Itching
- Fever and/or chills
- Shortness of breath
- Dizziness

Tell your doctor right away if you have any of the following symptoms. They could be signs of a serious problem.

- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain, swelling, warmth, redness, or a lump in your legs or arms. These could be signs of a blood clot.
- Numbness or weakness of an arm or leg or one side of your face. Sudden confusion, or trouble speaking or understanding.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of a brain swelling called meningitis.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a blood problem.
- Chest pains or trouble breathing.
- Fever over 100°F. This could be a sign of an infection.

Tell your doctor about any side effects that concern you. You can ask your doctor to give you more information that is available to healthcare professionals.

**How do I use Hizentra?**

Infuse Hizentra only after you have been trained by your doctor or healthcare professional.

Revised: October 2011
No two PIDD patients are the same. All are affected differently by their disease, and their responses to IG therapy vary widely.

By Annaben Kazemi
Three patients, one disease, yet none functions the same as the other — despite all being treated with immune globulin (IG) therapy. This can be said for the thousands of individuals affected by primary immunodeficiency disease (PIDD). Some are highly functioning and living normal lives with only an occasional infection; others are marginally functioning due to frequent infections; and yet others struggle with so many complications that they are rarely able to function at all. Why does PIDD present so differently in patients?

The truth is that no two PIDD patients are the same. There are more than 180 types of immunodeficiency, each with a wide spectrum of symptoms affecting many systems of the body. Physicians diagnose PIDD based on poor or absent response to immunization, serum concentration of IgG (IgA, IgE, IgG and IgM) and the number of infections that are classified as recurring, persistent, debilitating and chronic.

IG is the standard therapy with the goal of allowing PIDD patients to lead full lives. But because the disease is so complex and varied, IG therapy varies in its ability to achieve adequate IgG trough levels to allow patients to fight off infection. For instance, one person may become severely ill with an IgG trough level of 700, whereas another might become that sick with a level of 600. This makes it a challenging task for immunologists to prescribe the optimal dosage of IG therapy to ensure an adequate IgG trough level for each patient. Complicating the dosage issue are the side effects and the infusion method — intravenous (IV) versus subcutaneous (SC), the former of which results in a greater variance in the IgG level during the infusion cycle.

In essence, there are no typical PIDD patients, and PIDD patients seem to have no typical days. So much depends on how they are feeling, what their complications are, how their bodies are responding to therapy, as well as where they are in the infusion cycle. This is illustrated by the stories of three PIDD patients whose lives are as varied as the disease itself.

Robert’s Story

Robert, a busy 42-year-old executive and avid baseball fan from New York, was diagnosed as an adult with PIDD, but he doesn’t think of himself as sick. Robert wakes up early each morning and gets in a 30-minute walk with his golden retriever before starting his busy day. He tries to maintain a healthy lifestyle by eating only organic foods, taking supplements and exercising daily. He power naps most afternoons and is religious about getting enough sleep. He is able to go to work every day, help coach his son’s baseball team and live “pretty normally.”

Before Robert was diagnosed with a PIDD, he saw countless doctors for most of his adult life for recurring allergies, ear infections and chronic sinus infections. At one point, Robert’s sinuses were so bad that he was told to have ear, nose and throat surgery. Within a month of his surgery, Robert had three more sinus infections. His allergist tested for an immune deficiency, and he was diagnosed with common variable immune deficiency (CVID). A year after being diagnosed, Robert began treatment with IVIG therapy, which he now receives at an infusion clinic every four weeks.

There are no typical PIDD patients, and PIDD patients seem to have no typical days.

Robert hasn’t had a serious infection since starting IVIG, but he still checks in with his immunologist twice a year. When the occasional infection does occur, it is resolved with a prolonged course of antibiotics, but it doesn’t impede his lifestyle as it used to. He states that the time he spends in the clinic (four to six hours a month) to get IVIG has completely revolutionized his world. “Sometimes having this disease is really horrible, and it can get you down,” explains Robert. “The risk of infection is real; subsequently, a healthy or unhealthy amount of fear has accompanied my disease. But I’ve learned life goes on and it’s mostly pretty good.”

Now that he has experienced a prolonged period of wellness, Robert admits he is not as much of a germaphobe as he was when first diagnosed with CVID. He used to open every public door with a paper towel in his hand. He says he realizes that it could be much worse. “Much more serious infections like pneumonia can occur in CVID, but I’ve been rather lucky in that department,” he says. “Those antibodies appear to have shown up for work.”

He coaches his son’s baseball team and is either out on the field or in the batting cage most nights of the week.
“I love baseball. I’ve either been playing the game or coaching just about my whole life. It’s what I love to do,” Robert emphasizes. “I hope that other people with immune disorders will see that I am an adult living vibrantly with the disease and use me as an example that it is possible to enjoy an active lifestyle despite being diagnosed with CVID.”

Sarah’s Story

Sarah, a 19-year-old college student who lives in the Pacific Northwest, had been sick her entire childhood and spent her adolescence filled with tests and doctors’ appointments. After a serious hospitalization with pneumonia, Sarah was finally diagnosed with hypogammaglobulinemia when she was 14 years old, after it was determined that the pneumonia and flu vaccines had no effect on her immune system. Upon diagnosis, Sarah began treatment with IVIG therapy every three weeks. But because Sarah suffered severe migraines after IVIG therapy that kept her in bed for 24 hours at a time, affecting both her studies and lifestyle, she was transitioned to a twice-weekly regimen of SCIG.

On a typical day, Sarah wakes up and studies for a while, checks in with her online friends and updates her infusion log with how she is feeling. Sarah chose to go to a college close to where her parents live so that she can continue to live at home rather than in a dorm, where illness can spread quickly among even healthy students. Her mom is her biggest supporter and ally, and she checks in with her regularly. But her mom encourages Sarah to take ownership of her illness.

Sarah’s SCIG infusions take about one hour. She prefers SCIG to IVIG because she feels it gives her more control and freedom. With her busy college schedule, she likes being able to infuse when it suits her time frame rather than scheduling an appointment. She also likes that it takes less time (one hour at a time as opposed to four to six hours each time in the clinic). However, there is also a downside. “It’s kind of a drag to stick yourself or have someone else stick you at multiple sites; it’s just not something you look forward to,” Sarah explains.

Sarah has to see her immunologist every three months to monitor her levels while on SCIG. She sees her primary care doctor even more often, and she is especially susceptible to infections when she gets run down around the middle and end of each semester. Not being able to take rests when she needs them has taken its toll as well. She was treated for a respiratory infection last fall and ended up on a nebulizer for several weeks. But it’s the constant aches and pains that wear her out. According to Sarah, “Fatigue has been my worst enemy. On bad days, thinking too hard is like competing in an intense sporting event, and there are times I just don’t want to get up off the couch.”

While Sarah admits that since transitioning to SCIG her energy levels have been much more stable, she is still not as fully functional as she’d like to be. She doesn’t go out as much as other co-eds, and she has to constantly turn down their invitations. It’s hard when they say things like “But you look so good today” and “I can’t even tell you’re sick; aren’t you better now?” No matter how hard she tries, she can’t make her college friends understand how exhausting it is for her or that her disease isn’t something that can be cured.

Jenny’s Story

Jenny, a ”58 and fabulous” grandmother from Nevada, went undiagnosed and misdiagnosed until she was in her early 50s. “I had been told for many years that I had other conditions instead,” she said.

For years, Jenny had long periods of normal health and then was suddenly struck by high fevers, pneumonia or sinus and throat infections that she couldn’t shake for months at a time despite several rounds of antibiotics. She eventually needed intravenous antibiotics. While her ear, nose and throat doctor was baffled, she continued to struggle through life, dragging herself to work, wondering...
why she got sick so easily and so often. According to Jenny, the illnesses would just seem to “hit all at once,” leaving her fatigued and chronically worn out.

Unfortunately, while Jenny fought for a diagnosis, she faced a number of difficulties, among them misunderstandings and discrimination by her employer. Many people thought it was “all in my head,” Jenny says. Eventually, she had to go on disability because the constant exhaustion was causing her to miss so much work. In addition to missed workdays, the doctors’ visits, treatments and hospitalizations all were having a significant financial impact on Jenny’s family. And the insurance companies “just didn’t understand or seem to care.”

Jenny was diagnosed five years ago with acute chronic sinusitis, but it wasn’t until she still didn’t respond to treatment and was undergoing her fourth surgery that the underlying PIDD diagnosis was made. When treated with IVIG, she developed severe muscle pain, headaches and back pain. She was then switched to SCIG therapy, but she still experienced problems and could not tolerate the medication in large doses. Through a lot of trial and error, Jenny and her doctor decided she needed SCIG infusions on a daily basis at a very slow rate, which takes four to five hours.

Jenny has been receiving daily SCIG infusions for four years now, and she still struggles with fatigue, although it’s not as debilitating as it used to be. Her days are centered around her infusions, and she always takes an afternoon nap. She does try to do a little bit of light housework, read or play “Words With Friends,” and she cooks dinner for her husband. But she misses out on a lot of social and family events because she just can’t deal with the aftermath of exhaustion and infection. It has been a frustrating journey, and Jenny says “the worry has turned even my healthy periods into times of great anxiety.”

While Jenny doesn’t feel she has a “normal” life because she is isolated and rarely leaves the house, she concedes that it’s better than it was. “I’ve had to struggle with having this disease and all the discouragement that goes along with having it,” she says. “I still can’t do the things others can, but I lean on my faith as a source of support and encouragement.”

**An Individual “Normal”**

PIDD patients work, play, marry and have families just like people unaffected by disease. While a PIDD shouldn’t keep patients from living a normal life, the degree to which their illness affects their lifestyle does vary widely. As these stories illustrate, every person is affected differently.

A patient’s level of functioning depends upon the type and degree of antibody deficiency and the treatment regimen prescribed. While some patients, like Sarah, start to feel better and are able to begin normal activities after beginning treatment, other patients, like Jenny, continue to be greatly impacted by symptoms and complications even with treatment. Then, there are patients like Robert: His lifestyle is relatively unaffected by his disease; he’s never had to stop working, and his treatments are far less time-consuming than Sarah’s or Jenny’s. It’s important for PIDD patients to understand that it’s normal for different people to experience differing levels of functioning.

Because of the extreme variance in the disease, as well as in the response to treatment, patients need to be aware of their condition and advocate for access to trained specialists who understand their disease and are aware of the most recent developments in managing symptoms. Equally important is for patients to stay informed about obtaining and keeping health insurance coverage and about the laws and regulations that govern insurance. Education and awareness are key components to helping PIDD patients make informed choices so they can live their lives to their fullest potential.

**ANNABEN KAZEMI is the patient advocate for IG Living magazine.**

*Editor’s note: The patients in this article are fictitious, developed through a series of composite interviews with identifying details changed to protect the privacy of individuals.*
Understanding Pemphigus and Pemphigoid

By Michelle Greer, RN

These autoimmune diseases are so rare that they often are difficult to diagnose, and while there is no cure, there are effective treatments to put them in remission.
Mike woke up one morning with pain on the right side of his mouth. Looking at it in the mirror, he saw what looked like a small sore, and he brushed it off. But after a week, it was getting worse. He went to a dentist, who ordered an antifungal rinse, which did nothing for it. The dentist referred him to a periodontist, who promptly removed all of his wisdom teeth and ordered a different type of rinse. A month later still with no relief, he noticed a small sore beginning in the lower lid of his left eye. At first, he didn’t connect the two sores, but after some time, both were progressing.

Eight months and as many physicians later, Mike received yet another diagnosis: mucous membrane pemphigoid. Not only was he confused because he didn’t know what it was, he was also frustrated that it took so long and so many doctors to figure out what the sores were. Yet he also was relieved because he thought he would finally get the appropriate treatment.

Unfortunately, the relief he felt only lasted a little while, and the frustration and confusion continued. Mike was prescribed prednisone, a powerful steroidal anti-inflammatory that seemed to help temporarily. But each time the dose was tapered, the sores would flare up again. The high-dose steroids eventually caused diabetes, and for a few weeks, he required insulin to control his high blood sugar. Then, he was prescribed an immunosuppressant, and he ended up in the hospital with a rare fungal infection. Finally, he saw a dermatologist with expertise in his disease who prescribed intravenous immune globulin (IVIG) therapy in addition to the prednisone and immunosuppressant. At last, the blisters healed, and he was able to discontinue all other medicines after six months. Unfortunately, it was not before Mike had permanent scarring in his left eye, leaving him with partial visual difficulties. He also was very underweight because the sores in his mouth made eating painful for him. At one point, Mike weighed only 170 pounds, not a lot for his 6-foot-1-inch frame. While his doctors wanted to put in a feeding tube, he refused.

How Are Pemphigus and Pemphigoid Diagnosed?
Pemphigus and pemphigoid are diagnosed through special testing. One test, immunofluorescence, uses a dye to look for the presence of antibodies that are responsible for the attack on self. There are two types of immunofluorescence: direct and indirect. With indirect immunofluorescence, blood is drawn to look for the presence of antibodies. With direct immunofluorescence, the tissue is stained with a dye and examined under a special microscope to look for antibodies. The ELISA test measures the levels of autoantibodies to specific skin molecules targeted by pemphigus and pemphigoid. Another diagnostic measure may include rubbing an area near a blister. In pemphigus, it is likely the top layers of skin will be rubbed off when this is done (known as Nikolsky’s sign).
How Are Pemphigus and Pemphigoid Treated?

Once a diagnosis is made, treatment almost always begins with high-dose oral steroids. These steroids are powerful anti-inflammatory drugs that suppress the immune system. In many cases, this is enough to control the disease and stop the attack on self so that blister formation ceases and existing blisters heal. Unfortunately, as in Mike’s case, this treatment has significant side effects that can be so severe that the therapy is not tolerable.

Immunosuppressants often are prescribed to suppress the immune response, but this treatment may not result in the attack on self subsiding. Immunosuppressants also can cause significant side effects that result in undesirable immune suppression, which can then result in unusual infections. And these infections lead to additional treatments with antibiotics and other therapies and may result in hospitalization.

IVIG in high doses can be a very effective treatment. IVIG in high doses can be a very effective treatment. although some physicians may prescribe the dosing differently. Severe side effects of IVIG include renal failure, increased risk of blood clots, and aseptic meningitis. The

| Table 1. Types of Pemphigus and Pemphigoid |

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<thead>
<tr>
<th>Pemphigus Type</th>
<th>Description</th>
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<tbody>
<tr>
<td><strong>Pemphigus vulgaris</strong></td>
<td>Most common type. Blisters are soft and fragile and may form in the mouth first and then spread to the skin and even genitals. Blisters are frequently painful but not itchy, and in the mouth make chewing and swallowing difficult.</td>
</tr>
<tr>
<td><strong>Pemphigus foliaceus</strong></td>
<td>Less severe type. Blisters may form on the scalp and face first and then spread to the chest and back. Blisters are not usually painful and are superficial and form crusts.</td>
</tr>
<tr>
<td><strong>Pemphigus vegetans</strong></td>
<td>Thicker sores mainly in groin and under arms.</td>
</tr>
<tr>
<td><strong>IgA pemphigus</strong></td>
<td>Caused by IgA (an antibody) binding to the epidermal cells. May resemble pemphigus foliaceus or may appear as small pustules.</td>
</tr>
<tr>
<td><strong>Paraneoplastic pemphigus</strong></td>
<td>Associated with certain forms of cancer. Blisters form inside the mouth and may affect the lungs, leading to a fatal outcome.</td>
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<tr>
<th>Pemphigoid Type</th>
<th>Description</th>
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<tr>
<td><strong>Mucous membrane pemphigoid</strong></td>
<td>Affects the eyes, mouth and throat. A clinical form called cicatricial pemphigoid can result in blindness if it involves the eyes, and respiratory compromise if it involves the deeper parts of the throat.</td>
</tr>
<tr>
<td><strong>Bullous pemphigoid</strong></td>
<td>Limited to the skin with blisters presenting predominantly on the abdomen, groin, back, arms and legs. The blisters may itch and be painful.</td>
</tr>
<tr>
<td><strong>Gestational pemphigoid</strong></td>
<td>Blistering rash starting around the navel and spreading to the entire body, typically in the second trimester.</td>
</tr>
<tr>
<td><strong>Epidermolysis bullosa acquisita</strong></td>
<td>Blistering rash on the skin without involvement of mucosal surfaces. Blisters are usually smaller than in pemphigoid.</td>
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risk of these side effects can be minimized by assessing risk factors prior to therapy, as well as selecting an appropriate IVIG brand, premedicating properly and running the infusion at an appropriate rate. Other less severe side effects include headache, nausea, vomiting, fatigue, malaise and blood pressure fluctuations. These side effects are most often rate- or batch-related. The rate-related complication can be managed by slowing or pausing the infusion and then resuming it. The batch-related complication can be managed by premedicating prior to and after the infusion. Premedications are usually acetaminophen and diphenhydramine.

**There is currently no cure for pemphigus or pemphigoid, only remission.**

A relatively newer therapy is rituximab, which can be used alone or in combination with IVIG. Again, this will depend on the prescribing specialist. Rituximab is a monoclonal antibody, meaning it is made from immune cells that are all identical to one parent cell. Monoclonal antibodies target specific immune cells. Rituximab targets B cells. Depleting B cells is effective in halting immune attacks such as pemphigus and pemphigoid. But side effects due to induced B cell immunodeficiency can be serious and require testing for certain infections such as hepatitis B prior to therapy, as drugs such as rituximab can reactivate these conditions. Reported side effects of rituximab include renal failure and other severe reactions. Another severe side effect called progressive multifocal leukoencephalopathy (PML) is a progressive condition that results in death. Although these side effects are concerning, most people receive this therapy without issue. There are proper infusion precautions that can be taken, as well as up-front assessment of any risk factors.

There is currently no cure for pemphigus or pemphigoid, only remission. The goal of any therapy is to achieve quick remission with the least amount of steroidal medication. Once initial control of the disease is achieved, doses of steroids are tapered with the ultimate goal of discontinuation. A person is considered to be in full clinical remission when he or she does not need to take medications to remain lesion-free.

**Awareness Increases the Odds**

Pemphigus and pemphigoid are conditions that are so rare that they are not in the forefront of a physician’s mind when assessing what appears to be a simple blister or rash. That’s why it is important to raise awareness of these autoimmune diseases so that earlier detection and intervention can be possible (see The Awareness Campaign sidebar). A prompt and accurate diagnosis will lead to faster initiation of the appropriate treatment and, ultimately, to better outcomes that will decrease or eliminate the chance of long-term and/or permanent complications.

**The Awareness Campaign**

The International Pemphigus & Pemphigoid Foundation (IPPF) is currently funding the Awareness Campaign with the goals of increasing pemphigus and pemphigoid knowledge, reducing diagnostic timelines and improving treatment protocols. The campaign includes training lectures for the 62 dental schools in the U.S., where an estimated 4,500 new dentists graduate each year; two videos, one from expert clinicians and one based on patient testimonies; a fellowship program (funded by IPPF) that will allow side-by-side training with leading pemphigus and pemphigoid experts; and a consensus meeting (hosted by IPPF) that will bring the greatest pemphigus and pemphigoid minds together to redefine existing diagnostic and treatment practices to result in improved patient care. Collectively, the videos, training, fellowship program and consensus meeting underscore the need for early detection and intervention. The videos will be available on www.pemphigus.org.

**MICHELLE GREER, RN, is the vice president of sales at NuFACTOR Specialty Pharmacy.**
Parenting: Teaching Children to Care for Their Own Health Needs

It’s never too early for parents to teach their children about their disease, how to live a healthy life and how to take an active role in their treatment.

By Mark T. Haggard

CHRONICALLY ILL CHILDREN face greater challenges than other kids their age. One of the biggest challenges is to learn to care for their own special health needs so they are prepared for adulthood. And, parents play a crucial role in this process. It starts by educating children about their condition and what is happening inside their bodies, and then teaching them how to treat themselves and be proactive about their care.

Educate Them About Their Disease

Education is key to helping kids understand what they are going through. Fortunately, there are many educational resources. Many pediatric immunologists are able to translate the advanced language of immunology into a language that kids can understand as they grow older. The Immune Deficiency Foundation has a “Kids Connection” page on its website to educate children (primaryimmune.org/patients-and-families/idf-kids-connection). IG Living recently launched “IGL Teen,” a section on its website for teens being treated with immune globulin (IG) therapy to connect (www.IGLiving.com/IGLTeen.aspx). The Jeffrey Modell Foundation website offers literature at www.info4pi.org. And, author Sara Le Bien has published a book titled Our Immune System.

Teaching children to understand their bodies is important. Knowing which signs may signal that they are becoming sick allows them to seek treatment immediately before a small illness gets out of hand. Parents can detect certain signs that even kids might not recognize. For instance, when one of my children becomes irritable for no reason, it is an indication of a coming infection; my son used to burst out in tears if I just looked at him wrong because he knew I thought it meant he was getting an infection. I also know that when one of my kids wakes up on the couch, they are likely getting an infection.

Also, once children know what is going on in their bodies, they understand why the medications and their unique delivery systems are important. (It doesn’t make an injection any easier, but it helps to understand its importance.)

Teach Them to Live a Healthy Lifestyle

It is especially important for parents to teach kids to be proactive about their condition rather than reactive. A healthy lifestyle decreases opportunities for pathogens to reach kids’ immune systems. Therefore, parents should be models of healthy lifestyles by maintaining a healthy diet, washing hands with soap and water, using hand sanitizer, brushing and flossing.
teeth, refraining from smoking, avoiding secondhand smoke, fastening seat belts in the car, and using protective equipment while riding bicycles or engaging in other outdoor sports.

The National Institutes of Health (NIH) provides a number of suggestions for ways parents can help kids learn to live healthy lifestyles: 1) Involve the entire family in activities such as walking, hiking, bicycling or other recreational sports; 2) Take a trip to the zoo or the park, which can involve a lot of walking; and 3) Make children responsible for household chores that involve vigorous activity such as mowing the lawn, walking the dog or washing the car. Most importantly, kids need to see their parents having fun performing these activities; if parents look like they are enjoying the activities, the kids will likely enjoy them.

Unfortunately, “screens” have become a huge roadblock to healthy lifestyles. Getting children out from in front of a screen, whether it’s a television, computer monitor or a handheld game, is important. The NIH recommends limiting television watching and not allowing televisions in children’s rooms. They further recommend that families eat dinner together with the television off, have family game nights with the television off, and not use television or video games to reward children. Parents need to be mindful about how much they watch television and use computers themselves, since children’s lifestyles will likely mimic theirs.

To promote physical activity, parents can look for outdoor fun in their hometowns. Fortunately, our local area offers a plethora of opportunities, including fishing, biking, hiking and skiing. While my wife and I are leery of the play structure at the local fast-food restaurant, we let our kids do as much as possible otherwise. For instance, my daughter is a ballet dancer and my son plays football. We know there is the possibility that the slightest injury can be aggravated by an infection, so we keep a specialized first-aid kit on the sideline during practices and games, and give our son extra padding to cushion any potentially injurious blow. Obviously, kids should abstain from activities that their doctors prohibit, but if there are no concerns, the upside of improved immune health from being physically fit far outweighs the possibility of an infection from an injury.

Teach Them to Be Active in Their Treatment

Involving kids in their treatment is another way for parents to prepare kids to care for themselves. Rather than letting Mom or Dad do all of the work, children should be encouraged to take charge themselves by researching what is occurring inside their bodies, scheduling infusions and making their own contacts. Consistency is important. Children can keep their own calendars and infusion logs and begin setting their own appointments.

Of course, there may be a limit to how much children can be involved in their treatment, depending upon whether they are prescribed intravenous IG (IVIG) or subcutaneous IG (SCIG). For instance, my son’s IVIG requires a home health nurse, so he is limited in how much he can manage in his treatment. My daughter started SCIG when she was 6 years old. For now, I put the tubing together and prime the pump, my wife accesses my daughter, and my daughter de-accesses herself when she is finished. We hope to eventually give her the reins entirely, but at 10 years old, she is not yet able to handle everything.

It’s entirely possible that while children are learning to be active in their treatment, they will have a setback. For instance, Michael, who has selective IgA deficiency and takes antibiotic prophylaxis, was given the reins to be responsible for taking his medicine soon after he turned 13. His mother stopped giving him daily reminders. And, while he was responsible for a few weeks, his regimen became sporadic for another week, and he got a sinus infection. After a few rounds of higher-potency antibiotics, Michael recovered. The lesson he learned has made him more loyal to his prophylaxis regimen, and he has not been struck by any more major infections.

Like teaching kids to brush their
teeth, parents can’t harp on them every morning and night. A once-in-a-while reminder doesn’t hurt, but if parents do that too often, they start to sound like Charlie Brown’s teacher: “Wah wah, wah wah wah.” As a teacher, I know that kids learn best on their own, and they will eventually learn to be responsible. For Michael, it was a hard lesson, but it was a lesson learned.

Don’t Dash Their Dreams
Parents have to allow their kids to dream as well. From the time my son started talking, he wanted to be a U.S. Navy Blue Angel pilot. His story is similar to another child with common variable immune deficiency (CVID) who desperately wanted a career in the Navy. But conventional wisdom told his mother that the Navy would not take a candidate with CVID, and she told him as much. “Mom,” he replied, “it’s OK for someone else to take away my dream, but it’s not OK for you to do it.” He didn’t need his mother’s advice; he needed her unconditional love and support. In reality, this is all parents can do for their children: Encourage them, not tell them what the world says they cannot do.

It’s Never Too Early for Independence
Some parents want their children to perpetually remain 5 years old. Other parents feel they need to protect their children from their illness, thereby making their children dependent on them. It’s not OK to do either of these things. Children will eventually leave the nest, and they will have to act on their own. Success or failure will be up to them. It is never too early to begin teaching children to care for their own health needs. If they fail as children, their parents can help them; if they fail as adults, most times, their parents cannot.

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.
Reader: My husband recently had a five-day intravenous immune globulin (IVIG) treatment, which was finished four days ago. At the IV site, there is an approximately 1-inch-by-3-inch mass under the skin that is somewhat swollen and painful to the touch. It is better today than yesterday, and there is no fever. We are wondering if this is a common reaction or something about which we should seek medical treatment.

Michelle: This is more of a reaction to the infiltration rather than the IVIG itself. The vein, as well as the tissue, becomes irritated when fluid seeps outside the vein. Normally, warm compresses help. If there is no improvement or if it worsens or other symptoms appear such as rash, redness or increased pain, your husband should see a physician.

Reader: I'm aware of the drug low-dose naltrexone (LDN) being studied for the treatment of autoimmune diseases, especially multiple sclerosis (MS). Is it possible that LDN might help to treat chronic inflammatory demyelinating polyneuropathy (CIDP)?

Michelle: I have checked with several neurologists who are actively engaged in studying different treatment protocols for CIDP. None of them is aware of any current studies using LDN for the treatment of CIDP at this time. I also checked the National Institutes of Health clinical trials website (www.clinicaltrials.gov), and it appears that most of the current trials using LDN are targeting MS, Crohn's disease and addiction disorders.

Reader: I receive my intravenous immune globulin (IVIG) infusions at an infusion center. Our sons live on the West Coast in Washington and Oregon, and we would like to take extended visits there. How can I arrange to receive my treatments every three weeks? I don't know any doctors where they live.

Leslie: Traveling when being treated with IVIG can be difficult. Your physician may have some contact with physicians in the locations where you are traveling who may be able to coordinate your infusions. However, it can be quite difficult to coordinate single infusions. Your physician will be unable to prescribe your IVIG to be received in a facility in another state, since he or she won’t have “admitting” privileges for that facility. Instead, the local physician will have to prescribe your IVIG, and he or she may first require you to come in for an office visit. There also may be insurance challenges if the infusion center is not contracted with your insurance company.

To accommodate travel, an option is to consider changing to the weekly subcutaneous route of administration. If this is an option for you, I suggest discussing it with your physician as soon as possible to allow time for learning how to self-administer prior to traveling. A home infusion or specialty pharmacy company will provide you with all of the IG, supplies and pump needed to complete your infusions, and they will send a nurse to your home during the first several infusions to teach you to self-administer.

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MICHELLE GREER, RN, is the vice president of sales for NuFACTOR Specialty Pharmacy.

Have a question? Email us at editor@IGLiving.com. Your information will remain confidential unless permission is given.
Let’s Talk!
By Trudie Mitschang

Deb Stanzak is the founder of RonWear Port-able Clothing. After several years of personal tragedy, Deb founded RonWear with the hope of helping people who receive infusion treatments. RonWear is a specially designed line of clothing with zippered openings to accommodate ports for dialysis, infusions or chemotherapy.

**Trudie:** Tell us about how you came up with your idea for RonWear.

**Deb:** My brother, Ron, was a noncompliant diabetic with complications that included renal failure. He had three dialysis treatments a week and usually wore a short-sleeve shirt. He often complained of being very cold in treatment, and he would put many blankets on him at the dialysis center to keep warm. I am a seamstress and have a background in textiles and apparel in the retail market, and I went home and made a makeshift shirt of warm fleece with a zipper in the arm for the port. He loved it and so did the nurses.

**Trudie:** Once you came up with the original design, how did you turn that into a growing company?

**Deb:** Ron wanted me to make more samples, and other patients were asking to purchase them. I had a lot on my plate at the time, and after some initial research, I put the idea on the shelf, where it sat for years. Before Ron passed away in 2005, he made me promise I would develop the idea, and in 2008, I went back to the drawing board to find out what more needed to be addressed so that this garment could fully accommodate those going through dialysis or any type of infusions. I added things like dual-tab zippers that allow port access with openings in compliance with government regulations just small enough to allow the site to be seen by the attendant. Then, I put zippers in all the areas patients could have a port: both sides of the chest and both arms. I created pants with openings on both legs for femoral ports. This way, if one port gets infected or has to be moved, the patient does not have to purchase a new item.

**Trudie:** What other features are unique to RonWear?

**Deb:** The material is antimicrobial and doesn’t retain odors. I also made it water-repellent and stain-resistant. I added a media pocket in the front to securely house a cell phone or a digital device (since they fall out of pockets when the patients lie down). I put elastic in the waistband of the pant for comfort; the jacket only has elastic in the back and a flat front, so if a patient is bloated, the flat front is more flattering. The companion pant matches the jacket to make it look like an outfit — more like a stylish jogging suit. Many doctors say: “This is your new normal.” But in my experience, most patients want their “old normal” back.

I tried to give them that with RonWear.

Many doctors say: “This is your new normal.” But in my experience, most patients want their “old normal” back.
most patients want their “old normal” back. I tried to give them that with RonWear.

**Trudie:** What was the biggest obstacle you had to overcome?

**Deb:** The biggest obstacle was learning the manufacturing business. That was something all of my retail, textile and management training did not include. Manufacturing is a dying business in the apparel industry in the U.S., for a variety of reasons. Although I really wanted to produce my goods in the U.S., I gradually accepted that it was not financially possible, nor was it affordable to the customer if we manufactured here.

**Trudie:** What advice do you have for those who have an idea for a business?

**Deb:** My advice is: “Go for it!” Don’t hesitate, just do it! But, you need to do your homework. You cannot look back, and you cannot quit. You will make mistakes, and you will spend cash that, looking back, you would have used differently. But I don’t know one business owner who doesn’t have the same story. In the end, it is worth it. The other suggestion I would give is to do a lot of networking. Meeting people who can help or who know people who can help you will move the business forward. Networking is key!

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**Trudie:** What inspires you?

**Deb:** The first time I spoke with a patient who wore my RonWear outfit and he told me how it changed the whole treatment experience for him and thanked me profusely, I knew I had to do this. My customers motivate me!

**Trudie:** What are your goals for the future?

**Deb:** I want RonWear to give patients options in their treatment wardrobes. I want RonWear to make patients feel normal again in a very abnormal environment. I am expanding the line to more apparel options and accessories, and making our website a “one-stop-shop” for those in treatment. Expanding to the pediatric market is on our radar also. Caregivers want to “save” their loved one or friend. They want to cure them and heal them. And although that is not possible, they can make their treatment experience much easier with RonWear.

For more information on RonWear, visit [www.ronwear.com](http://www.ronwear.com).

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Prior to passing away, Deb’s brother made her promise to turn her prototype into a business, which she calls RonWear, that gives patients options in their treatment wardrobes.

**TRUDIE MITSCHANG** is a staff writer for IG Living magazine.

If your life depends on immune globulin and you have a unique experience to share, we want to feature you in this column! Email us at editor@IGLiving.com.
I AM A 14-YEAR-OLD freshman in high school who loves to dance, cheer, listen to music and hang out with friends. Three words I use to describe myself are **hardworking**, **outgoing** and **friendly**. But there is one thing that many people don’t know about me: I live with a disease called primary immune deficiency disease (PiDD). My disease has made me a stronger, more confident individual, but it also has its downsides. It’s difficult to be as active as my friends, and I worry about confiding in others about my disease. Yet one thing I have learned is that PiDD is what makes “me” me. If I didn’t have this disease, I wouldn’t be the person I am today.

PiDD dictates what I can and can’t do, such as whether I can go to social events, dance practice, hang out, etc. I take steps so that I don’t get sick, but being in crowded high school classes, it’s sometimes unavoidable. I’ve learned that when I’m tired, it’s more important to take some time out. Unfortunately, a lot of people don’t know or understand what I’m going through. They think I just need to get more sleep by going to bed earlier or that I should drink an energy drink.

PiDD also makes me miss a lot of school. Now that I’m in high school, I’ve learned I have to communicate with my teachers. I’ve found out that it’s important to have a good relationship with my teachers and to email them individually when I’m out. This makes them more understanding and patient about missing school, and it has helped me to stay on top of the workload and helped them to know what I go through.

The hardest part about having this disease is having to tell other people — especially because of how people have reacted in the past. I’m a very social girl, and I love hanging out with my family and friends. But when I tell people about my disease, they treat me differently. I don’t like the pity and sympathy they give me; I just want to be treated the same as anyone else. That’s the hard thing to accept about my disease: I can’t be.

I also worry about telling my current friends about my disease. I worry that some will not want to be friends anymore. They might think I’m contagious and not want to be around me, or that I’m too fragile to have fun, or that they couldn’t handle the scary parts of me being in the hospital. It’s frightening to think someone you trust, care about and love might leave you. I also feel that classmates at my school whom I’m not close with will judge me — call me a freak, weird or disgusting. I’ve experienced bad reactions, and they make me very private about my disease. However, I know that my true friends accept me for who I am, and they will never leave just because I have a disease. They try really hard to look past my disease and treat me the same as before they knew about it. These friends have become my biggest supporters.

But even these great friends don’t get some things about my disease. That’s why connecting with others who can share the same fears and hopes is really important. I don’t know anyone in my town or high school who shares my same disease, but I have online friends I can talk to who get it. I’m super excited to join IG Living’s teen group and hope other teens will too so that we can support each other.

**ARIANNA KAZEMI** is the daughter of IG Living’s patient advocate, Annaben Kazemi. She enjoys helping others and teaches dance to young beginning dancers. She is a member of the IG Living website’s teen group.
IG Living magazine is excited to announce the debut of its newest web page, IGL Teen. The site is filled with inspiration, motivation and opportunities for readers to connect with other teens in the IG community.

Log on to IGL Teen to find cool links, tools and tips, inspiring videos, blogs, reader photo uploads, book reviews, poems, playlists and much more! There are teen-targeted resources too – our Get the 411 quickly connects visitors with articles, advice and information specially geared toward our younger audience.

“Being a young adult is tough enough, and having a chronic illness doesn’t make it any easier, but the resources on this website are helpful and can be used in our everyday lives. I’m super excited!” says IG Living reader Callie Hines, 17.

“I really like the Motiv8 section because the videos and poems by other teens are inspirational,” adds Arianna Kazemi, 14. “The tabs make it easy to get around and find things – I like that it’s just for teens.”

The new website went live on January 18, and it’s already generating plenty of buzz! The celebration kicked off with a week of online contests and promotions on the IG Living Facebook and Teen Group pages.

Know an inspiring teen we should profile? Tell us about them! Are you an aspiring teen writer, photographer or artist? Send us your work so we can publish it. IGL Teen is also on the lookout for bloggers and columnists. Send inquiries and writing samples to editor@IGLiving.com.

“Being a young adult is tough enough, and having a chronic illness doesn't make it any easier, but the resources on this website are helpful and can be used in our everyday lives. I'm super excited!”
I HAVE A ... um... a... port-a-catheter — better known as a “port.” There, I said it. Yes, I’m a proud owner of not just a port, but a PowerPort! Why this gutsy move, you ask? I was tired of being a human pin cushion, so I bit the bullet and acquired a first-class, able-to-withstand-contrast PowerPort, and I’ve never been poked more than once since.

I know, I know. Port is a four-letter word in our community. Implanting a port is quite controversial, or possibly even scandalous. If you get one, don’t be surprised if port-placement gossip rears its ugly head upon your post-procedure appointment. But you are now part of an elite group of patients who simply would not agonize through one... more... inhumane... IV... insertion. Period. Come to think about it, immune-compromised patients contemplating a port, or patients who are having ports placed behind their physicians’ backs, could make
for great reality television!

Getting a port is just not a popular medical decision, nor is it for the faint of heart. Why the brouhaha? I think the answer lies in another four-letter word: germ (and we all know what kind of havoc germs like to cause). But, between you, me and the infection-infested fly on the wall, I get sick breathing. Frankly, port-a-catheters are for those like me who have tried everything in the book to get their veins to cooperate on infusion day. Our veins roll, flatten, hide and disappear like a 2-year-old caught with her hand in the cookie jar. We are an army of folks who have drunk an ocean of water (minus the salt, of course), fought off our favorite adult libation and forgone our beloved salt shakers in order to produce plump, luscious veins for the next day's needle placement — all, of course, to no avail. Some think of me as a brave and tenacious self-advocate for insisting my doctor approve a port, while others think I've gone completely postal for placing an “infection waiting to happen” in my body. However, a recent trip to my handy-dandy surgeon's office had me thinking I wasn’t the one who had spent too much time around microwaves.

“So, what body part brings you in today, Cheryl?” a cheery voice chirped, interrupting my pain threshold.

“It’s my right shoulder, Dr. Kay,” I said while measuring up the resident physician.

“Oh, please forgive me for not introducing you to our new resident, Dr. Janik.”

Dr. Janik and I exchanged pleasantries while Dr. Kay fumbled through my medical records. Dr. Janik, a ruddy sort of gent who was finishing the last of his orthopedic residencies, was getting used to our balmy weather but not our Idahoan naïveté.

“So, whaddaya think?” I asked, gripping my right shoulder.

“Wewhhheellllllll, let's take a look-see, shall we?” Dr. Kay sing-songed, making his way toward me. A sharp pain stabbed my bicep, sending a throbbing wave through my entire right arm. It was as if my whole right side was protesting Dr. Kay’s impending inspection.

“Dr. Janik, see if you can find Ms. Haggard’s latest films, OK?”

“Absolutely. I’ll be right back,” Dr. Janik announced with a certain caution in his voice.

As Dr. Kay inspected his future surgical appointment (my right bicep’s tendon and a few other anatomical disasters caused by my aggressive autoimmune disease), Dr. Janik knocked on the door, let himself into the room and made his way to the computer screen.

“We’re up ‘n runnin', Doc,” Dr. Janik declared casually as if he were Dr. Kay’s Bugs Bunny to his Elmer Fudd.

They whispered and discussed; they hemmed and hawed.

“When did you get that pacemaker placed, Cheryl?” Dr. Kay asked.

“Um… that's not a… ur… uh…” I bumbled, looking over my shoulder as best I could.

Am I supposed to correct a very-well-respected surgeon by telling him he has his medical implants completely confused, or do I keep my mouth shut and hope to the high heavens he figures it out? I argued in my mind. I looked over to see what Dr. Janik was doing, hoping he was going to do the dirty work for us both. No such luck; Dr. Janik appeared to have been struck by lightning and absolutely unable to speak, let alone walk.

It seemed as if a lifetime warped until I was able to eek out a “Well, uh…” all the while wondering what happened to my smarty-pants resident.

“That’s all you gotta say…well, uh…?” Dr. Kay mocked. I didn’t mind as I had racked up a lot of frequent flier miles with him and other physicians at his practice. These folks knew me quite well as a kidder with decent insurance, except this time, no one was kidding about my port gone pace.

I looked again at Dr. Lightning, just hoping for some sort of life, and all I got was an imaginary talk bubble above his head with absolutely nothing inside of it.

“So, when did you have this pacemaker put in, and for what?” Dr. Kay asked, a bit more impatiently.

“Well, uh, Dr. Kay, that's not exactly a pa—”

“Pacemaker,” Dr. Wonderful Resident decided to chime in and save the day as I was ever so grateful I didn’t have to say a four-letter word in my surgeon’s office.

And I am happy to report my right bicep’s tendon is feeling better than ever and, yes, they did use my PowerPort for surgery, thank you very much. ☻

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.
AT LEAST ONCE a week, someone asks me, “What can I do to help?” You see, both of my children have complex medical problems, and together they have spent hundreds of days and nights in various hospitals from New York to New Orleans. My “zebra” is 11-year-old Ethan, who has common variable immune deficiency; my 6-year-old Jenna has lymphatic malformation.

Usually the conversation goes like this:

Friend: “Sorry that you guys are having a hard time (again). What can I do to help?”

Me (while silently running through the list of things that would be helpful but not wanting to admit weakness): “Um, I don’t know. Thanks for asking.” This scenario plays out time after time, and rarely does anyone outside of the immediate family actually end up helping. I’ve thought a lot about this phenomenon: Clearly, my friends want to help, but they don’t know how to make it happen. It seems easier for loved ones to rally around an acute and time-limited problem, but harder to wrap their arms around the fact that chronic illness is the family’s reality every day.

That’s why I wrote this on behalf of all of us who are stressed out, worn out and maxed out! Here are some things that friends and family can easily do to make a difference:

1. Provide respite: What that means is to take the kids so that the parents can have a break and focus on protecting their marriage. Call and ask if you can take the kids on a specific day at a specific time. Tell the parents that you have a great activity that the kids would love. This eliminates the ever-present parental guilt.

2. Take the sibling(s): Take the non-sick sibling on a special outing. It is hard to watch your brother/sister suffer day after day. Give these kids a fun break and some special time too.

3. Buy groceries: Call and say, “I’m headed to the grocery store. What can I pick up for you?” Then drop off the requested items, and don’t stay. Don’t make the family feel that they need to entertain you.

4. Make dinner: Call and say, “I’m making lasagna for dinner. I would love to make two pans and drop one off at your house.” Follow the drop-off protocol described above.

5. Provide funding: Prior to an out-of-town medical trip, anonymously give the family a gift card for gas, hotel costs and food. Most families with special needs children spend a shockingly large percentage of their income on quality medical care and access to specialists, prescriptions and special educational needs.

6. Do yard work: Show up and start mowing!

7. Tutor/help with homework: Sick kids get behind in school. Offer to come by and help with homework while the parent goes for a coffee break.

8. Be a friend: Don’t let the fear of the family’s pain scare you away. Be a friend who sticks around. Be a friend who replies to emails and text messages when the family uses precious emotional energy to reach out. Be an encourager.

I would be remiss if I didn’t mention the one major don’t. Don’t judge the medication and treatment choices made by the parents. Unless you’ve had a child with a chronic illness, you really don’t know what it is like — it is not like your healthy child’s broken arm.

I challenge you to reach out and provide encouragement to a parent of a special needs child. Your one gesture could be the thing that keeps a family together and moving forward for the sake of their child.

DENNA MCGREW is a professional children’s advocate with a regional nonprofit organization in northeast Louisiana. Her 11-year-old son, Ethan, was diagnosed in 2007 with common variable immune deficiency after being sick since birth.
FINDING THE JOY in life can sometimes be difficult when I have to deal with health complications that are overwhelming. Whether it is a new diagnosis, an upcoming procedure, the onset of a new symptom or even a change in medication or treatment, out of fear, I take to Google like a moth to a flame looking for answers and information. But then, suddenly, I find myself bombarded with the most heinous of all possible explanations!

I remember a few years ago when I was suffering from severe tension headaches. I Googled my symptoms, and I came across pages and pages of brain tumor diagnoses. I was convinced I had one. I was going blind, and I could have a stroke at any time. Looking back, it seems funny, but in the moment, I was horrified. The laundry list of all the disturbing — yet at the time what seemed like inevitable — outcomes was so not funny. I became sad and then angry and then went through every other stage of grief before I even knew what was wrong with me. I had no diagnosis, yet I had a sense of impending doom that I couldn’t shake. Like Alice, I had fallen down a rabbit hole of negativity and fear — with no joy in sight! It had been only 15 minutes, and I had fallen so far that I didn’t know how to climb out.

It has taken me years to come to terms with the fact that I can’t take everything I read on the Internet at face value. I’ve had to learn to turn off the screen, step away from the endless abyss of negativity and realize that my symptoms probably are not that bad — not even close to what I will find online. Instead, what I need to do in this circumstance is take a painkiller and email my doctor. I need to release my tension by accepting that whatever the outcome is, it is out of my hands. Around every corner, I will face challenges — some of which will be easier to overcome than others. But the only way I can control my fear is to deal with it by letting go.

So how do I let go? Most importantly, it’s never by not caring. I care, because if I didn’t, I wouldn’t feel fear. Instead, I don’t worry; I take action. I remind myself that what I’m experiencing could be much worse, and then I find the joy around me. Even in the worst situations, I know I have so much to be grateful for and that I can’t get bogged down by worrying about it all.

It starts with baby steps. I consciously make the choice to see all that is good. I think about all the things and the people I love. I think about the one thing at that moment that can lift my spirits. And I go for it — guilt-free — like a manicure, a peppermint hot chocolate, a nap, new lip gloss or anything involving glitter. It may seem trivial, as though I’m putting a Band-Aid on a bigger problem, but I truly believe that by allowing myself those little bits of joy into my life, I can slowly allow myself to see the more profound joy that is around me all the time.

EVER FECSKE MAZZA was diagnosed with CVID and interstitial lung disease in 2004. She is a fashion design student, loves spending time with her husband, family and bulldog, Dunkin, and can’t get enough of writing, cake decorating and anything that sparks!
WHEN IT COMES to obtaining the nutrients an individual’s body needs, the best possible source is food, especially fruits and vegetables. However, circumstances may prevent some people from eating optimally every day. This is especially true for patients with a chronic illness, who may explore the use of vitamins and supplements to optimize their health. The key for these patients is to choose the right vitamins and supplements.

Determining the Need

The primary reason for taking supplements is to protect against gaps in the diet. Research shows that some important vitamins (B and D particularly) and minerals are protective against disease in amounts that may be difficult to obtain through diet alone, no matter how conscientious one is.

Another reason for taking supplements is to make up for the vitamin and mineral deficiencies caused by some medications. For example, patients prescribed methotrexate, a drug used in chemotherapy and for autoimmune issues, may be required to take folic acid. In cases of deficiencies, a physician may prescribe vitamins or supplements.

Ensuring Quality and Safety

Unlike prescription and over-the-counter medications, vitamins and supplements are not regulated by the U.S. Food and Drug Administration (FDA). However, there are organizations that test these products for strength, purity, dissolution and disintegration. A label that indicates testing by U.S. Pharmacopeia or another reliable source such as ConsumerLab.com or NSF International can indicate whether a supplement is effective, safe and of good quality. If a supplement has not undergone examination by one of these organizations, it is preferable to switch to a supplement that has passed one of these organizations’ standards.

It is important to read the product label and check the dosage of a vitamin or supplement. For instance, there is no standard multivitamin formula. The term “multivitamin” applies to any combination of vitamins and minerals in any strength, as long as they are listed on the label. Labeled ingredients are listed in descending order of amount used. If all the natural ingredients are at the bottom of the list, chances are they make up only a small percentage of the product. All of Pharmacopeia’s products contain at least 99 percent natural ingredients. If there are any doubts, a pharmacist should be consulted.

It’s also important to note that price is not a predictor of quality. ConsumerLab.com reported that some multivitamins that sell for less than 10 cents a day perform better in tests than those selling for 50 cents or more.

Talk with a Healthcare Professional

The FDA suggests consulting with a healthcare professional before using any dietary supplement. Not only can a physician discuss with patients the safety of a particular product and whether that product is appropriate, but some vitamins and supplements may be contraindicated with current medications.

ANNABEN KAZEMI is the patient advocate for IG Living magazine.
An Epidemic of Absence: A New Way of Understanding Allergies and Autoimmune Diseases

Author: Moises Velasquez-Manoff
Publisher: Simon & Schuster Inc.,
www.simonandschuster.com

An Epidemic of Absence explores the promising but controversial “worm therapy” — deliberate infection with parasitic worms — in development to treat autoimmune disease. It explains why farmers’ children so rarely get hay fever, why allergy is less prevalent in former Eastern Bloc countries, and how one cancer-causing bacterium may be good for people. It also probes the link between autism and a dysfunctional immune system, and investigates the newly apparent fetal origins of allergic disease — that a mother’s inflammatory response imprints on her unborn child, tipping the scales toward allergy.

Do You Trust Me? Allowing Hope to Triumph Over Tragedy

Author: Jessica Leigh Johnson
Publisher: WestBow Press,
www.westbowpress.com

In Do You Trust Me?, Jessica Johnson gives readers a vivid and honest look at her very personal struggles with faith, prayer and trust in the midst of the most painful event of her life. In 2006, Jessica and her husband were living the life they had always planned. But several months after the birth of her third child, Jessica was faced with the question, “Do you trust me?” in a way that she had never dreamed of before. She is the mother of four boys with X-linked agamaglobulinemia (XLA), one of whom passed away from the disease. This book is about her family’s experiences with the initial sickness, diagnosis and, ultimately, the death of their son, as well as their life with three more boys diagnosed with XLA. It addresses primary immune deficiency disease (PIDD), and is intended to be beneficial for other parents who have newly diagnosed children, or who have lost a child to PIDD. Do You Trust Me? is not just for those struggling with the loss of a child, but anyone who has ever wondered, “Does God even listen when I pray? Does he truly care about his children?” Jessica hopes that after reading the book, others will discover that the answer to these questions is a resounding “Yes!”

Brain On Fire: My Month of Madness

Author: Susannah Cahalan
Publisher: Simon & Schuster,
www.simonandschuster.com

One day in 2009, 24-year-old Susannah Cahalan woke up alone in a strange hospital room strapped to her bed, under guard and unable to move or speak. Only weeks earlier, she had been on the threshold of a new, adult life: a healthy, ambitious college grad a few months into her first serious relationship and a promising career as a cub reporter at a major New York newspaper. With the help of a lucky, ingenious test, neurologist Souhel Najjar saved her life. He recognized the symptoms of a newly discovered autoimmune disorder in which the body attacks the brain, a disease now thought to be tied to both schizophrenia and autism, and perhaps the root of “demonic possessions” throughout history. Brain on Fire is the powerful account of one woman’s struggle to recapture her identity and to rediscover herself among the fragments left behind. Using all her considerable journalistic skills, and building from hospital records and surveillance video, interviews with family and friends, and excerpts from the deeply moving journal her father kept during her illness, Susannah pieces together the story of her “lost month” to write an unforgettable memoir about memory and identity, faith and love.
These organizations provide information about various disease states, which can be found by conducting a search of the disease state name.

- Advocacy for Patients with Chronic Illness: www.advocacyforpatients.org
- American 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
- Baxter: www.baxter.com
- Bio Products Laboratory: www.gammalex.com
- CSL Behring: www.cslbehring.com
- Grifols: www.grifolsusa.com
- Kedrion: www.kedrionusa.com
- Octapharma: www.octapharma.com

For a more comprehensive list of resources, visit the Resources page at www.IGLiving.com.
Idiopathic Thrombocytopenic Purpura (ITP)

**WEBSITES**

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

Kawasaki Disease

**WEBSITES**

- American Heart Association: www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsofChildhood/Kawasaki-Disease_UCM_308777_Article.jsp#.T1T2boePWE0
- Kawasaki Disease Foundation: www.kdfoundation.org

Mitochondrial Disease

**WEBSITES**

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

Multifocal Motor Neuropathy (MMN)

**WEBSITES**

- Neuromuscular Disease Center at Washington University: neuromuscular.wustl.edu
- Neuropathy Action Foundation: www.neuropathyaction.org
- 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**

- The Myositis Association: www.myositis.org

Pemphigus and Pemphigoid

**WEBSITES**

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

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**The Myositis Association**

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

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**ONLINE PEER SUPPORT**

- P.A.N.D.A.S. Network: pandasnetwork.org

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**Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)**

**WEBSITES**

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

**ONLINE PEER SUPPORT**


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**Pemphigus and Pemphigoid**

**WEBSITES**

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

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Sources

• Neuropathy Action Foundation: www.neuropathyaction.org

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

Peripheral Neuropathy (PN)

WEBSITES

• Calgary Neuropathy Association: www.calgaryneuropathy.com

Neuropathy Action Foundation: www.neuropathyaction.org

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

Primary Immune Deficiency Disease (PIDD)

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

Jeffrey Modell Foundation

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

WEBSITES

• The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/health/topics/Primary_Immunodeficiency.cfm

• American Academy of Allergy, Asthma & Immunology: www.aaaai.org

• International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: www.ipopi.org

• New England Primary Immunodeficiency Network: www.nepin.org

• Team Hope (for families and patients in New England): www.teamhope.info

ONLINE PEER SUPPORT

• Scleroderma Support Forum: curezone.com/forum/f.asp?f=404

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

Scleroderma

WEBSITES

• Scleroderma Foundation: www.scleroderma.org

• Scleroderma Research Foundation: www.srfcure.org

• Scleroderma Center: www.hopkinsmedicine.org/rheumatology/clinics/scleroderma_center.html

ONLINE PEER SUPPORT

• Scleroderma Support Forum: curezone.com/forum/f.asp?f=404

• International Scleroderma Network: www.sclero.org/support/forum/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

Rhode Island peer group: health.groups.yahoo.com/group/RhodeIslandPIDD
Other Resources

**Education and Disability Resources**

- Americans with Disabilities Act of 1990: www.ada.gov Provides protection for people with disabilities from certain types of discrimination, and requires employers to provide some accommodations of the disability.


- 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.


- World Allergy Organization: www.worldallergy.org

**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 Gammalex: www.gammalex.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

- Intra Pump Infusion Systems: www.intrapump.com

- Micrel Medical Devices: www.micrelmed.com

- Norfolk Medical: www.norfolkm edical.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.
HIGHLIGHTS OF PRESCRIBING INFORMATION
These highlights do not include all the information needed to use Octagam, Immune Globulin Intravenous (Human), safely and effectively. See full prescribing information for Octagam.

Octagam® [Immune Globulin Intravenous (Human)]
5% Liquid Preparation
Initial US Approval: 2004

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

- Renal dysfunction, acute renal failure, osmotic nephrosis, and death may be associated with Immune Globulin Intravenous (Human) (IGIV) products in predisposed patients.
- Renal dysfunction and acute renal failure occur more commonly in patients receiving IGIV products containing sucrose. Octagam 5% liquid does not contain sucrose.
- Administer IGIV products at the minimum concentration available and the minimum infusion rate practicable.

RECENT MAJOR CHANGES

Warnings and Precautions – Hyperproteinemia 8/2008

INDICATIONS AND USAGE

- Octagam is an immune globulin intravenous (human), 5% liquid, indicated for treatment of primary humoral immunodeficiency (PI).

DOSAGE AND ADMINISTRATION

Intravenous use only.

<table>
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<th>Indication</th>
<th>Dose</th>
<th>Initial Infusion rate</th>
<th>Maintenance infusion rate (if tolerated)</th>
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<td>PI</td>
<td>300-600mg/kg</td>
<td>0.5mg/kg/min</td>
<td>3.33mg/kg/min Every 3-4 weeks</td>
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- Ensure that patients with pre-existing renal insufficiency are not volume depleted; discontinue Octagam 5% liquid if renal function deteriorates.
- For patients at risk of renal dysfunction or thrombotic events, administer Octagam 5% liquid at the minimum infusion rate practicable.

DOSE FORMS AND STRENGTHS

Octagam 5% liquid is supplied in 1.0g, 2.5g, 5g, 10g, or 25g single use bottles

CONTRAINDICATIONS

- Anaphylactic or severe systemic reactions to human immunoglobulin.
- Immunoglobulin A (IgA) deficient patients with antibodies against IgA and a history of hypersensitivity.
- Patients with acute hypersensitivity reaction to corn.

WARNINGS AND PRECAUTIONS

- IgA deficient patients with antibodies against IgA are at greater risk of developing severe hypersensitivity and anaphylactic reactions.
- Epinephrine should be available immediately to treat any acute severe hypersensitivity reactions.
- Monitor renal function, including blood urea nitrogen and serum creatinine, and urine output in patients at risk of developing acute renal failure.
- Falsely elevated blood glucose readings may occur during and after the infusion of Octagam 5% liquid with some glucometer and test strip systems.
- Hyperproteinemia, increased serum viscosity and hyponatremia occur in patients receiving IGIV therapy.

- 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 has been reported with Octagam 5% liquid and other IGIV treatments, especially with high doses or rapid infusion.
- Hemolytic anemia can develop subsequent to IGIV therapy due to enhanced RBC sequestration.
- IGIV recipients should be monitored for pulmonary adverse reactions (TRALI).
- The product is made from human plasma and may contain infection agents, e.g. viruses, and theoretically, the Creutzfeldt-Jakob disease agent.

ADVERSE REACTIONS

The most serious adverse reactions observed with Octagam® 5% liquid treatment have been immediate anaphylactic reactions, aseptic meningitis, and hemolytic anemia. Most common adverse reactions with an incidence of >5% during a clinical trial were headache and nausea. To report SUSPECTED ADVERSE REACTIONS, contact Octapharma at 1-866-766-4860 or FDA at 1-800-FDA-1008 or www.fda.gov/medwatch.

DRUG INTERACTIONS

- The passive transfer of antibodies may confound the results of serological testing.
- The passive transfer of antibodies may interfere with the response to live viral vacancies.

USE IN SPECIFIC POPULATIONS

- Pregnancy: no human or animal data. Use only if clearly needed.
- In patients over age 65 or in any person at risk of developing renal insufficiency, do not exceed the recommended dose, and infuse Octagam 5% liquid at the minimum infusion rate practicable.

HOW SUPPLIED

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MANUFACTURED BY:
OCTAPHARMA Pharmaeutika
Produktionsges.m.b.H.
Oberlaaer Strasse 235
A-1100 Vienna, Austria

DISTRIBUTED BY:
Octapharma USA, Inc.
121 River Street, Suite 1201
Hoboken, NJ 07030
Tel: 201-604-1130
Fax: 201-604-1131
www.octapharma.com/usa

Revised: September 2009
CONTRAINDICATIONS

octagam® 5% liquid is contraindicated in patients who have acute severe hypersensitivity reactions to human immunoglobulin. octagam® 5% liquid contains trace amounts of IgA (not more than 0.2 mg/ml in a 5% solution). It is contraindicated in IgA deficient patients with antibodies against IgA and history of hypersensitivity. octagam® 5% liquid is contraindicated in patients with acute hypersensitivity reaction to corn. octagam® 5% liquid contains maltose, a disaccharide sugar which is derived from corn. Patients known to have corn allergies should avoid using octagam® 5% liquid.

WARNINGS AND PRECAUTIONS

IgA deficient patients with antibodies against IgA are at greater risk of developing severe hypersensitivity and anaphylactic reactions. Epinephrine should be available immediately to treat any acute severe hypersensitivity reactions. Monitor renal function, including blood urea nitrogen and serum creatinine, and urine output in patients at risk of developing acute renal failure. False elevated blood glucose readings may occur during and after the infusion of octagam® 5% liquid with some glucometer and test strip systems. Hyperproteinemia, increased serum viscosity and hyponatremia occur in patients receiving IGIV therapy. 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 has been reported with octagam® (Human) 5% liquid does not contain sucrose. (See Dosage and Administration and WARNINGS and PRECAUTIONS in enclosed Full Prescribing Information for important information intended to reduce the risk of acute renal failure.)

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References


Please see Highlights of Prescribing Information on adjacent page.

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Date of Preparation 6/12. GAMS-003-PAD-R1
Now it’s easy when
YOU CHOOSE
your delivery dates!

Visit MyFluVaccine.com to secure YOUR best delivery dates.

Choice
Select from a broad portfolio of products

Convenience
Choose your delivery dates

Safety
Count on a secure supply

YOU PICK THE QUANTITY • YOU PICK THE DATE • WE DELIVER

Brought to you by FFF Enterprises, Inc., the nation’s largest and most trusted distributor of flu vaccine and critical-care biopharmaceuticals.