Preventive Care for Chronic Illness

Exercise Options for CIDP Patients

A Review of Medicare and Disability Programs

How Patients Can CONNECT With a Community
OCTAGAM®
Immune Globulin Intravenous (Human) 5% Liquid Preparation
Initial U.S. Approval: 2004

RECENT MAJOR CHANGES

WARNINGS AND PRECAUTIONS

• IgA deficient patients with antibodies against IgA are at greater risk of developing severe hypersensitivity and anaphylactic reactions. Epinephrine should be immediately available 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.

ADVERSE REACTIONS

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-1088 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 vaccines.

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

Manufactured by: OCTAPHARMA Pharmazeutika 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

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A clear solution

Immune globulin intravenous (human) 5% liquid preparation

If you’ve been looking for an IGIV solution, take a look at octagam®.

octagam® has earned its reputation for safety and documented clinical efficacy.

To ensure tolerability, octagam® is carefully produced to retain as many of the characteristics of natural plasma as possible.

With over 40 million grams of octagam® infused world-wide, Octapharma is committed to helping PI patients live more active and healthier lives.

Ask your health care provider today about octagam® and find out if it could be the right solution for you.

For clinical or technical questions, please call our Medical Affairs team at 888-429-4535.

IMPORTANT SAFETY INFORMATION

octagam® is contraindicated in individuals with intolerance to immunoglobulins, especially in immunoglobulin A (IgA) deficiency, when the patient has IgE mediated antibodies to IgA.

Immune Globulin intravenous (Human) (IGIV) products have been reported to be associated with renal dysfunction, acute renal failure, osmotic nephrosis, and death. Other possible side effects with octagam® include: aseptic meningitis, hemolysis, transfusion-related acute lung disease (TRALI) and thrombotic events.

Immune Globulin Intravenous (Human) products have been reported to be associated with various minor reactions, such as headache, chills, backache, chest pain, fever, allergic reactions, arthralgia, dizziness, changes in blood pressure, cutaneous reactions and/or nausea and vomiting. Cases of reversible aseptic meningitis and migraine and isolated cases of reversible hemolytic anemia and reversible increases in liver function tests have been observed with octagam®. Immediate anaphylactic and hypersensitivity reactions are a remote possibility.

As with all medicines made from human plasma, the risk of spreading infectious agents, including viruses, cannot be completely eliminated.

Some types of blood glucose testing systems falsely interpret the maltose contained in octagam® as glucose. This has resulted in falsely elevated glucose readings and, consequently, in the inappropriate administration of insulin, resulting in life-threatening hypoglycemia.

See brief summary of PI on facing page.

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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MATTHEW DAVID HANSEN, DPT, MPT, BSPTS
Physical Therapist and President,
Allied Healthcare Staffing and
Consulting Agency

Exercise for CIDP
“Appropriate exercise is a vital part of any CIDP intervention plan because of its potential to improve strength and endurance, thereby minimizing muscle shrinkage and improving function and mobility.”

MARK T. HAGGARD
High School Teacher, Football Coach
and Parent of PIDD Children

Patient-to-Patient:
Connecting to a Larger Community
“Historically, patients were leery of going out in public because of the prospect of getting sick, but the Internet now allows scores of people to connect with each other in a community that stretches far beyond city limits, state lines and national borders.”

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

Humoral Components: The Role of the IgG Antibody
“Antibodies are typically thought to be one of the more critical components of immune function.”

JENNIFER KESTER
Freelance Writer

A Review of Medicare and Disability Programs
“If an individual meets the criteria for a disabled person, there are two ways they can receive benefits.”

KRIS MCFALLS
Patient Advocate, IG Living magazine

What Does Your Gut Say?
“Although currently there is no cure for either IBS or IBD, there are effective treatments.”

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 two-per-month, hosted, toll-free teleforums will be held, as well as what topic relevant to the IG community will be discussed.
- The moderated, hour-long calls will be filled on a first-come, first-served basis and will be limited to 15 readers.

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

Sign up for the Teleforums now by emailing kmcfalls@IGLiving.com or calling (888) 433-3888, ext. 1349.
Recently, a reader shared how her son, when he was in kindergarten, described her disease to his teacher: “My mommy has an invisible disease; it’s on the inside, so only Superman can see it!” Leave it to a child to come up with such an apt description for a primary immune deficiency disease (PIDD).

Recognized by the medical community only 50 years ago, PIDD remains mysterious in many ways. Those unaffected find it difficult to understand a disease that does not outwardly present itself; those suffering from a PIDD look just fine, but they’re not. And, with research in immunology so new, physicians struggle to identify the symptoms and find the causes, treatments and cures for the increasingly complex set of what is now more than 100 PIDD diseases.

But struggle they do, and with an awe-inspiring tenacity. I witnessed this at the Clinical Immunology Society’s (CIS) first annual PIDD National Conference held in May, which represented a new momentum in the quest to unveil this invisible disease. Why now? According to Dr. Charlotte Cunningham-Rundles, allergist and immunologist at the Mount Sinai Medical Center in New York, and co-chair of the conference, there had previously “been no U.S.-based forum for physicians and researchers to meet to present and discuss topics about PIDD. We thought that a meeting that concentrated on this topic alone, with a mix of the best scientists [and] with faculty and trainees in an open-access meeting, was required to fill this void.”

For three days, some of the top immunologists from all over the world collaborated on their research and challenged each other in discussions that were intended to help them learn from each other. How dynamic the field of immunology is; and, how diverse the way of thinking and treating!

Much of the research discussed focused on the connection between PIDD and autoimmunity. In addition, the need for updated registries was repeatedly acknowledged. According to Cunningham-Rundles, “Some of the most essential questions about PIDD can only be answered if we all collect our data into such a format. These diseases are rare, and each of us, working at one center, is only able to see a proportion of these patients over the years. Because of this, the data that is published is based on one person’s or one medical center’s experience and viewpoint. With a large registry, with multiple centers and geographic regions entering data, the combined information will give a truer picture of complications, best treatments and, ultimately, quality of life.”

Invisible disease? Not if these doctors can help it! As CIS members, they are dedicated to achieving the society’s mission, which, according to CIS Executive Director Anne Krolikowski, is “to enhance the competence of healthcare providers who use immunodiagnostics, treat patients with immunologic disorders and prescribe immunotherapy with the ultimate goal of improved patient care and management.” Thanks to the CIS, with more conferences like this, the mysteries of PIDD are sure to be unveiled.

To your health,

Ronale Tucker Rhodes, MS, Editor
Faces of IG Living

There are many faces in the IG Living community, representing hundreds of immune-mediated diseases and countless medical and lifestyle issues. IG Living’s magazine, website, Facebook page and blog are the ways in which our community can connect with one another. Be a part of the community by interacting with us. Your comments could lead you to others who share similar issues, and vice versa. Monday through Friday, we pose a new question on the IG Living Facebook page. Here is what some of our faces of IG Living are saying in response.

Question: Patient safety should be job No. 1 for providers supplying life-saving medication. How do you know your immune globulin or any of your other medication is authentic and unadulterated? Have you ever requested a pedigree from your provider?

Keli Carter
I was with a wonderful specialty pharmacy during my first year of infusions. I’m [an SCIG] patient [who] drew up my own syringes and was able to keep a running log of the lot numbers and expiration dates.

Abruptly, at the beginning of this year, we found out we were switched to a new specialty pharmacy because our insurance company [had] not [re-signed] a contract with the previous one. I was not notified by my insurance carrier of the change until it was already made; some people would have been able to continue with the old pharmacy if they had switched the insurance coverage at renewal time in October/November, but they did not have the heads-up either. I found out from the pharmacy we were with what they had done. Plus, I had just qualified for another hardship grant with our previous pharmacy.

The new pharmacy started sending me pre-drawn syringes, two weeks’ worth at a time [without] lot numbers or expiration dates, in the first shipment. I requested the info be sent and asked why they were drawing it up. They gave me some lame excuse and I went on about my business, until I started noticing that my energy level was dropping off — badly — after the second week. I called and they said they had never heard that before. Then, I called my immunologist and the manufacturer of the IG I receive. The manufacturer said they would not recommend using the IG after a 24-hour period of being drawn up in the syringe. After that, the pharmacy started sending me the vials again. I am back to myself again....

I can’t tell you how much I’ve wondered if it was all in my head or not. But, I know how I felt and how I’m feeling now. It’s a night and day difference.

Question: What aspects of healthcare reform are you excited about, and what would you like to see covered in IG Living?

Laura Guenther
I’m excited about the no lifetime maximums. I was in a high-risk state pool for years and ended up nearing my $1 million lifetime max, forcing me to take a new job simply for the benefits, and now have an insurance plan that has no lifetime max. But, it would be nice to have the freedom of feeling I could pursue what I’d like to career-wise without choosing based solely on these factors!

Question: What sections of each IG Living magazine do you look most forward to reading?

Suzanne Todd Revell Colville
I’ve only received two issues so far, but [I] enjoyed them so much that I went online and looked back at almost every one that was there. Great info: teaching articles, recent news on research, what’s going on concerning PIDDs in general. In fact, I ordered it for my pulmo/immuno’s office. I like it that much; I told the doctor that I did so.
IN PREVIOUS COLUMNS, I discussed components of the immune system that typically may not be considered part of immune protection. But the immune system also is made up of more traditional components, which can be broadly divided into cellular components and humoral components. In this column, I will focus on one of the humoral components.

Humoral Components Defined
The term “humoral” dates back to an earlier period when the fluids of the body were spoken of as “humors.” The body was said to have four fluids, or humors — blood, phlegm, yellow bile and black bile — that were thought to define the physical and emotional health of the individual. Today, the term humoral persists to define components found in the serum or plasma. For the immune system, these consist of antibodies, complement and complement-related proteins, and protein, carbohydrate or lipid factors, which affect immune system activation or otherwise regulate immune function (e.g., cytokines, immunokines, prostaglandins, leukotrienes).

Antibodies: The First Humoral Component
Antibodies are typically thought to be one of the more critical components of immune function, in part because their levels can be readily measured in a person’s serum, and specific antibody levels can be evaluated to measure how well a person’s immune system is working. Further, individuals deficient in antibody levels and/or function can be treated by replacement therapy with intravenous immunoglobulin (IVIG) or subcutaneous immunoglobulin (SCIG).

As is typical in medicine, antibodies may go by several names. They may be called gamma-globulins (based on their discovery) or immunoglobulins (because they are serum proteins, or globulins, with immune function). Because they are infused, antibodies may be called IVIG or SCIG by some as a descriptor of their replacement method.
Sometimes antibodies may be called Igs (for immunoglobulins) or IgG (specifically for immunoglobulin G).

IgG is one of the major classes of antibodies. The others are IgA, IgM, IgD and IgE. Each has more or less specific roles.

**The Role of IgG**

IgG is the major class of antibodies found in the blood. A typical normal adult value is approximately 1,000 mg/dL (10 g/L), with a normal range of 700 to 1,300 mg/dL (depending on the reference laboratory). IgG is the only antibody to pass through the placenta, which occurs during the last trimester. Therefore, a full-term infant will be born with IgG antibodies at a level equivalent to the mother’s serum level, which should be approximately 1,000 mg/dL. However, if a mother has an antibody deficiency, she will need extra infusions of IgG during the last trimester to ensure that her infant will be born with an adequate level.

After birth, babies begin to make their own IgG. The half-life (a measure of the loss of a serum protein) of IgG is approximately 21 to 28 days. This means that for any IgG molecule made, half of what was made is removed from the body over the ensuing three to four weeks. For example: If a person who could not make IgG receives an infusion of IgG that brings their serum level to 1,000 mg/dL, their level would fall to 500 mg/dL in the next three to four weeks, then to 250 mg/dL in the second three to four weeks, then to 125 mg/dL in another three to four weeks, and so on. Therefore, between three to six months after birth, the baby will have lost most of the maternal, transplacentally derived IgG. Further, an infant with the inability to make IgG (such as a male child with X-linked agammaglobulinemia) may not start showing signs of illness until after three to four months of age. Indeed, this is also why most infants do not get ill until after three to four months of age. And, ironically, this is when most infants are placed into a daycare setting — after they have lost their protective maternal IgG, but before they have had time to make adequate levels of their own.

**Antibodies Continued**

There is much more to discuss about antibodies in the next issue.

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

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**Have You Visited Our IG Living Blog Yet?**

Whether you follow blogs already or are new to social media, the IG Living blog is destined to become one of your favorites. We tackle topics that range from touching and thought-provoking to downright funny, and new blogs are posted every Friday. Check it out at www.igliving.com.

Happy reading and be sure to leave comments; we want to hear from you!
Correction

FDA Warns About WinRho Use for ITP

In the June-July issue of IG Living, we incorrectly reported that the FDA had issued a warning about intravenous immune globulin (IVIG) use to treat ITP. While the FDA did issue a warning, it was about using human intravenous immune globulin (WinRho SDF). In March, the U.S. Food and Drug Administration (FDA), Baxter Corp. and Cangene Corp. notified healthcare professionals that cases of intravascular hemolysis (IVH) and its complications, including fatalities, have been reported in patients treated for immune thrombocytopenic purpura (ITP) with WinRho(r) SDF [Rho(D) Immune Globulin Intravenous (Human)]. Fatal outcomes associated with IVH and its complications have occurred most frequently in patients 65 years of age and older with co-morbid conditions. In addition, serious complications that include severe anemia, acute renal insufficiency, renal failure and disseminated intravascular coagulation also have been reported.

The notification comes in the form of a boxed warning that informs healthcare professionals that 1) patients should be closely monitored in a healthcare setting for at least eight hours after IVIG administration; 2) a dipstick urinalysis should be performed at baseline, two hours and four hours after administration, and prior to the end of the monitoring period; 3) patients should be alerted to and monitored for signs and symptoms of IVH, including back pain, shaking chills, fever and discolored urine or hematuria (absence of these signs and/or symptoms of IVH within eight hours does not indicate IVH cannot occur subsequently); and 4) if signs and/or symptoms of IVH are present or if IVH is suspected after administration, post-treatment laboratory tests should be performed, including plasma hemoglobin, urinalysis, haptoglobin, LDH and plasma bilirubin (direct and indirect).

Grant

IDF Awarded Five-Year NIH Grant

In April, the Immune Deficiency Foundation (IDF) was awarded the “Resources to Assist Investigations in Primary Immunodeficiency Diseases” grant by the National Institutes of Health. The grant will allow IDF to continue to advance the detection, understanding, diagnosis and treatment of primary immunodeficiency diseases (PIDD) through its United States Immunodeficiency Network (USIDNET).

USIDNET is a team of leading immunologists whose purpose is to advance knowledge in the field of PIDD. USIDNET solicits, develops, evaluates and implements clinical research strategies to advance the detection, understanding, diagnosis and treatment of PIDD.

Medicine

Two New Oral MS Drugs on the Horizon

Two new oral drugs — fingolimod, manufactured by Novartis, and cladribine, manufactured by Merck Serono — may soon be available to combat multiple sclerosis (MS). According to the results of three Phase III studies, published in the online Jan. 20 edition of the New England Journal of Medicine, the drugs reduce relapse rates in people with relapsing-remitting MS. Both drugs work by altering the immune system response. However, there are concerns about side effects, including an increased risk of serious infection and, possibly, cancer.

The current treatments for MS are all injectable medications, which Dr. John Richert, executive vice president of research and clinical programs for the National Multiple Sclerosis Society, says are sometimes a barrier for people to start early treatment. And, since treatment is more successful when it is started early in the course of the disease, Richert is hoping that having oral medications will help people start treatment sooner.
**Medicine**

**New Website on Nutritional Supplements**

WellWise.org is a nonprofit foundation launched in March to provide an authoritative website on supplements, the science behind them and other health strategies. The site offers information on a range of supplements and the latest on their impact on various health conditions, with links to scientific research behind them. The site also features blogs by scientists, food and nutrition writers, researchers and doctors. According to the site’s editor-in-chief, James Townsend, “It’s not easy to find real evidence-backed information on supplements and nutrition on the web. The vast majority of it is thinly veiled marketing for a specific brand of supplement. We feel the public needs something more objective in order to make informed decisions about health.” Each month, the site will highlight a featured nutrient, offering in-depth reporting on the value of and recent science behind the nutrient, as well as video and audio interviews with newsmakers in the world of supplements and nutrition.

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**Research**

**Genetic Signature Predicts Autoimmune Diagnosis**

Researchers have identified a cellular genetic signature that predicts prognosis in two different autoimmune diseases. Conducted at the Cambridge Institute of Medical Research, the researchers looked at patients with AAV, a type of vasculitis characterized by inflammation of the blood vessels, and lupus, a disease caused by antibodies acting against the body’s own tissues. The team took blood samples from patients before treatment and isolated the different populations of immune system cells, including T cells, B cells and neutrophils. They then looked at a specific subgroup of T cells, known as CD8+ T cells, and found that the patients could be separated into two distinct groups based on their long-term prognosis, with one group having more disease relapses or flare-ups than the other.

The study does not explain how the CD8+ T cells contribute to the severity of the disease or why these particular genes might be switched on in patients with poor prognosis. However, it is hoped that by identifying a genetic signature that predicts poor disease outcome, doctors may one day be able to use the information to tailor drug therapy according to the individual patient.

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**Research**

**Discovery in Immune System Development**

Researchers at National Jewish Health have discovered a fundamental step in the development of the immune system, one that allows B cells to mature and fight disease by producing effective antibodies. Immunologist Roberta Pelanda, PhD, and her colleagues demonstrated that immature B cells in the bone marrow must receive a positive signal before they can migrate to the spleen, where they mature and are activated. “Our work demonstrates that the immune system uses both positive and negative selection processes to create an effective population of B cells,” says Pelanda. “A defect in either selection process could lead to autoimmunity or immune deficiency.” The research was reported on in the March 15 issue of the *Journal of Experimental Medicine.*
Mutations in the CD81 gene have been identified as a new genetic cause of antibody deficiency. Researchers at Erasmus MC, University Medical Center Rotterdam, Netherlands, studied a patient who had impaired antibody responses and an absence of CD19 expression on B cells (the immune cells that produce antibodies). No mutations were detected in the patient’s CD19 genes; however, mutations were detected in both copies of the CD81 gene, which is associated with a complete lack of CD81 expression on immune cells in the blood. Further analysis using B cells from the patient revealed that CD81 is required for CD19 membrane expression, providing mechanistic insight into the antibody deficiency caused by CD81 mutation. The research appears in the Journal of Clinical Investigation.

People and Places in the News

Two researchers whose work led to a groundbreaking new treatment for arthritis and other autoimmune diseases have been jointly awarded one of Germany’s most prestigious sciences prizes. Emeritus Professor Sir Ravinder Maini and Professor Marc Feldmann, from the Kennedy Institute for Rheumatology at Imperial College London, are the winners of the 50,000 pound Ernst Schering Prize. Established in 1991 by the Ernst Schering Research Foundation, this annual international prize is awarded for particularly outstanding basic research in the fields of medicine, biology or chemistry.

Professor Yehuda Shoenfeld, MD, has joined SQI Diagnostics Inc.’s Scientific Advisory Board as chairman to support the development of the company’s IgXPlex assay and platform development pipeline. The IgXPlex product pipeline for 2010 includes the highest-demand autoimmune test panels for celiac disease, vasculitis, lupus and irritable bowel disease (Crohn’s disease, ulcerative colitis). Shoenfeld is currently head of the Department of Medicine B and Center for Autoimmune Diseases at the Sheba Medical Center, and incumbent of the Laura Schwarz-Kipp Chair for Research of Autoimmune Diseases, Tel-Aviv University, Israel.

Dr. Michael Gresser has joined Trillium Therapeutics Inc.’s board of directors as an independent director. Gresser most recently was vice president of research inflammation at Amgen Inc., where his team worked on molecular targets, introducing numerous small molecules, human antibodies and other proteins into development. Trillium Therapeutics is a biopharmaceutical company developing innovative immune-based biologics.

Compugen Ltd. has discovered an experimental validation of a novel membrane protein, CGEN-15001, for the treatment of autoimmune disorders. The in vivo validation of CGEN-15001 indicates that it might have therapeutic utility for the treatment of multiple sclerosis and other autoimmune diseases, such as rheumatoid arthritis, systemic lupus erythematosus, inflammatory bowel disease and type 1 diabetes. These are among the most common autoimmune diseases, affecting an estimated 25 million people in the U.S. alone.

Mary K. Crow, MD, has been appointed physician-in-chief and chair of the Division of Rheumatology at the Hospital for Special Surgery. Crow, who is an internationally recognized research scientist and a leader in the field of rheumatology and autoimmunity research and clinical practice, has been the associate chief of the division of rheumatology and director of rheumatology research at the hospital since 2001. She succeeds Stephen Page, MD.
Link Found between Sunlight and MS

Because sunlight is more abundant near the equator, many researchers have wondered whether high levels of vitamin D engendered by sunlight could explain the reduced prevalence of multiple sclerosis (MS) in those regions. Now, researchers have found that not only may vitamin D reduce the symptoms of MS, but the ultraviolet (UV) portion of sunlight may play a bigger role than vitamin D in controlling the disease.

The study, published in the *Proceedings of the National Academy of Sciences* in March, was designed to distinguish the role of vitamin D and ultraviolet light in explaining the high rate of MS away from the equator. Using mice that are genetically susceptible to MS-like disease, the researchers triggered the disease by injecting a protein from nerve fibers. They then exposed the mice to moderate levels of UV radiation every two to three days for a week. The UV exposure (equivalent to two hours of direct summer sun) did not change how many mice got the MS-like disease, but it did reduce the symptoms of MS. In addition, the researchers found that although the UV exposure did increase the level of vitamin D, that effect, by itself, could not explain the reduced MS symptoms.

It’s possible that these findings could eventually lead to a new mode of treatment. “There are several ways this could go,” says Hector DeLuca, Steenbock research professor of biochemistry at the University of Wisconsin, Madison. “If we can find out what the UV is producing, maybe we could give that as a medicine. In the short term, if we can define a specific wavelength of light that is active, and it does not overlap with the wavelengths that cause cancer, we could expose patients who have been diagnosed with MS to that wavelength.”

Kawasaki Disease Reduced with Steroids

A study published in the October issue of *Pediatrics* shows that the probability of heart damage in children with Kawasaki disease is reduced to a considerable level when steroids are combined with the standard treatment of aspirin and intravenous immunoglobulin (IVIG). The study was conducted to identify the discrepancy in previous guidelines that state that steroid treatment has no benefits. In addition, the study showed that by combining steroids with aspirin and IVIG, the probability of inflammation development of the heart blood vessels is reduced by half. It is hoped a multi-center study currently underway will provide further evidence of the benefits of steroid treatment for Kawasaki disease, as well as provide more evidence about the most effective types and doses of steroids.

AARDA Launches Fund Raising Campaign

In honor of National Women’s Health Week, May 9-15, the American Autoimmune Related Diseases Association (AARDA) launched a mobile giving campaign titled “Text My Cure.” The campaign features YouTube videos of patients telling their stories of autoimmune diseases and is headlined by the AARDA national spokesperson, actress Kellie Martin (“ER” and “Life Goes On”). The funds raised by the campaign will be used to increase awareness of autoimmune diseases by physicians and patients, fund research in autoimmunity and autoimmune diseases and provide patient education and services. For more information, visit the AARDA website at www.aarda.org.

Did You Know?

People with asthma who have suspected or confirmed influenza should be strongly considered for antiviral medications because of their increased risk of developing a complication such as bacterial pneumonia.

— The Journal of Allergy and Clinical Immunology
Infusion Pumps: New FDA Initiative Launched to Reduce Device Risks

By Ronale Tucker Rhodes, MS

With more than 700 deaths linked to infusion pumps in the past five years, the U.S. Food and Drug Administration (FDA) is taking steps to address problems through its Infusion Pump Improvement Initiative.

Pumps and Problems

According to the FDA’s white paper on the initiative, between 2005 and 2009, the FDA received approximately 56,000 reports of adverse events associated with the use of infusion pumps, including numerous injuries and deaths. During this time, 87 infusion pump recalls were conducted by firms to address safety concerns, 70 of which were designated as Class II (a category that applies when the probability of serious health consequences is remote) and 14 were classified as Class I (situations in which there is a reasonable probability that use of the recalled device will cause serious adverse health consequences or death).

Between 2005 and 2009, the FDA received approximately 56,000 reports of adverse events associated with the use of infusion pumps, including numerous injuries and deaths.

The adverse events and recalls are not limited to any specific manufacturer, type of pump or infusion environment, but instead have occurred across multiple manufacturers and pump types. And while some adverse events have been the result of human error, the FDA says that many of the reported events are related to deficiencies in device design and engineering, which can either create problems themselves or contribute to user error.

Software defects, user interface issues and mechanical or electrical failures are the most common problems that have been reported. One software defect is known as “key bounce,” which occurs when a healthcare worker enters one number into the keypad, but it is actually recorded twice, which can cause the release of too much medication. According to Jeffrey Shuren, the director of the FDA’s device division, “When I punch 10 digits in my cell phone, I don’t get 11 or 12, and we should have the same expectation for infusion pumps.”

The most common user interface issues are confusing or unclear on-screen user instructions, which can lead to improper programming of medication doses or infusion rates. And, mechanical failures are often related to components, such as pump housings...
Did You Know?

that break under routine use, premature battery failures and sparks or pump fires.

**The Initiative**

The newly launched initiative represents a major shift in the FDA’s approach to medical device safety. In the past, the agency has taken actions to respond to issues that have arisen on a largely case-by-case basis. But, because the same types of problems continue to occur, it is taking a more proactive and comprehensive approach.

The first draft of the Infusion Pump Improvement Initiative suggests that manufacturers submit additional design and engineering information during premarket review of their devices; recommends that manufacturers explain the steps they have taken to reduce the risk of their device at each stage of its life cycle, from design to actual operation; and recommends tests of infusion pumps be conducted in the environment for which they are intended, such as the hospital or home, to account for real-life issues. It also states that the FDA may, in certain circumstances, exercise its authority to withhold premarket clearance of an infusion pump until the manufacturer’s facility has been inspected. While these are merely recommendations as of this writing, the FDA is currently taking steps, including a public comment period, to convert the draft guidance document into a special controls guidance and regulation that will require manufacturers of new and existing pumps to comply with the recommendations.

**Risk Reduction Strategies**

Prior to the implementation of the new initiative, the FDA is providing clinicians and patients with information and strategies to mitigate the risks associated with the use of existing infusion pumps. Its new infusion pump website (www.fda.gov/MedicalDevices/ProductsandMedicalProcedures/GeneralHospitalDevicesandSupplies/InfusionPumps/default.htm) features basic information about infusion pumps and commonly seen problems. In addition, the site has risk reduction strategies for users in all infusion environments, including clinicians, home health nurses and patients. These strategies cover planning ahead, checking pump settings, usage instructions, reporting problems and more.

**Taking an Active Role**

Healthcare professionals and patients are urged to do their part to help reduce the risks associated with infusion pumps. To help the FDA develop a better understanding of the risk-benefit profile, it is encouraging all users to file a voluntary report about any problems that are encountered at www.fda.gov/MedicalDevices/ProductsandMedicalProcedures/GeneralHospitalDevicesandSupplies/InfusionPumps/ucm202503.htm.

RONALE TUCKER RHODES, MS, is the editor of IG Living magazine.
Individuals with an immune-mediated disease often are so focused on tending to their chronic illness-related health needs that they neglect to practice preventive healthcare. In fact, not remembering to regularly see their primary care physician is the norm simply because there are so many other frequent visits to their various “ologists.” But, having a chronic illness can put patients at a higher risk for other diseases, making it even more important to be proactive about their overall health. This includes having regularly scheduled visits with a primary care physician (PCP), getting regular screenings and keeping up to date on vaccinations.

The Specialist-PCP Connection

Regular checkups with a PCP are often referred to as “well visits,” whereas visits to a specialist typically are “chronic care visits.” What’s the difference and why is it important? First, well visits are generally more comprehensive and involve planning and education on the part of the patient and the physician. For instance, at the basic health level, most individuals are unaware of what screenings or vaccinations are needed and when, and they don’t know that they should ask their doctors what is recommended. On the other hand, many PCPs may not be as well-versed about the patient’s chronic condition as they could be, so communication between the PCP and the patient is especially necessary.

When it comes to chronic care, specialists and PCPs have very different roles and, often, there is a lack of communication between the two. Dr. Marc Reidl, MD, MS, assistant professor of medicine, clinical immunology and allergy at the University of California, Los Angeles,
explains that chronically ill patients often have multiple, complex medical conditions, and it is the specialist’s role to provide detailed recommendations and advanced management of a specific condition. Ideally, this will optimize the patient’s health with regard to that specific issue. In addition, as medical diagnosis and treatment become increasingly complex, specialization is necessary to keep up with the vast amount of information in any specific area. “This focus allows a specialist to develop expertise through education and experience that the PCP may not possess,” says Reidl. “However, the specialist care is no substitute for the overall comprehensive care provided by the PCP, and sometimes, specialists can miss ‘the big picture,’ because they are so focused in one area.”

According to Annette Zampelli, MSN, CNRP, medical science liaison at CSL Behring Biotherapies for Life, the PCP is viewed as the “gatekeeper” of the patient’s health information, including making sure that redundancy is not occurring regarding labs, tests, etc. And while, ultimately, this should be the PCP’s responsibility, often it is the patient who communicates the information that avoids duplication. Ideally, says Reidl, there is regular communication between the specialist(s) and the PCP, and it is often the practice of specialists to report their findings and/or recommendations to the PCP. However, he adds, “there is no formal protocol for this interaction. My general advice to patients is to be their own advocate [and] make efforts to get all their clinic records and reports to ensure [their] PCP has information from all involved specialists.”

**Screenings Are Twice as Important**

Chronic illness predisposes patients to certain other diseases, including cancer. A recent survey conducted as part of the Gallup Healthways Well-Being Index, and based on telephone interviews with more than 350,000 adults in the U.S., found that individuals who have had a heart attack or chronic illness may raise the risk of being diagnosed with cancer. Those with high blood pressure, high cholesterol or diabetes were about twice as likely to have cancer as healthy people without these chronic illnesses.

Cancer screenings of particular importance to immune-mediated disease patients include the prostate-specific antigen test to screen for prostate cancer in men, which should be conducted annually in high-risk patients beginning at age 40 to 45; annual mammograms and Pap smears to test for breast, pelvic and cervical cancer in women beginning at age 40; and colonoscopies and fecal occult blood screenings to test for colon cancer in both men and women, with the first normally recommended at age 50 and then every 10 years, depending upon personal medical and family histories.

Other annual screenings also are of importance. Vision should be regularly checked (especially for those with autoimmune disease), as should hearing (particularly for those with chronic ear infections). Because obesity and being overweight are major risk factors for chronic disease in the general population, patients with immune diseases who are already at higher risk for chronic diseases especially need to maintain a healthy weight. Studies show that extra fat is a reservoir for toxic chemicals that can have an adverse effect on many cell types and, thus, is a cancer promoter. Therefore, body mass index should be regularly measured.

**When it comes to chronic care, specialists and PCPs have very different roles and, often, there is a lack of communication between the two.**

Individuals with immune-mediated diseases also are more prone to cancers such as lymphoma and skin cancer. Those with either a primary immune disease or an autoimmune disease who are on immune-suppressing therapy have an impaired ability to control or kill infectious agents and environmental toxins, putting them at higher risk for lymphoma. As a precaution, the lymph nodes should be regularly checked. And, patients on immune-modulating medications and frequent antibiotics have a higher risk of sun-related skin damage from UV radiation exposure. So, it becomes even more important to annually scan for and document any changes in skin pigmentation and growths.

Blood work is often part of both the specialty visit and the PCP visit. Patients on immune-modulating medications usually will have a complete blood count (CBC) test and a chemistry panel every three to six months depending on their medication. Blood tests performed at the yearly
screening that are not normally included in the specialist screening should include a CBC with differential and platelet count, a uric acid test, erythrocyte sedimentation rate test, C-reactive protein test and a lipid panel that tests for cholesterol levels.

Which Vaccinations, Why and When?

All individuals require some vaccinations at intervals throughout their lives, regardless of their health status. Vaccines can be grouped into four major categories: live, attenuated; dead, inactivated; component or conjugate; and toxoid. PCPs may avoid giving immunocompromised individuals live viral vaccines, such as measles-mumps-rubella (MMR) or chickenpox (varicella) vaccines, because a compromised immune system can’t recognize and fight off bacteria, viruses or other germs the way a healthy immune system can. However, which vaccines are recommended for those with a weakened immune system depends on their particular condition.

Live, attenuated vaccines usually are created from the naturally occurring germ itself. The germs used in these vaccines still can infect people, but they rarely cause serious disease. These vaccines include:

**Chickenpox.** Also known as varicella, chickenpox is extremely contagious, spread through the air when people sneeze or cough, or through an infected person’s chickenpox sores. Most children are vaccinated against chickenpox at 15 months old. People ages 13 years and older who were not vaccinated as children need two doses of the chickenpox vaccine. Once chickenpox is contracted, it is very rare but possible to get it again. It’s more common for people who have had it to develop shingles, caused by a reactivation of the same virus, later in life.

**Measles, mumps and rubella.** Most adults today are immune to measles, mumps and rubella either because they have had the diseases as children or they have been vaccinated against them. People born in or after 1957 have likely received at least one dose of the measles-mumps-rubella (MMR) vaccine. However, those born before 1957 who don’t think they’ve been vaccinated should be, and healthcare workers and individuals who travel outside of the U.S. are advised to get a second dose.

**Shingles.** Although shingles, caused by a reactivation of the chickenpox virus, typically occurs in adults later in life, adults of all ages have been known to get this disease. One dose of the shingles vaccine is recommended only for adults age 60 and over.

**Rotavirus.** Rotavirus is the most common cause of severe diarrhea among children, and approximately 55,000 children in the United States are hospitalized each year, while more than 600,000 children die from it annually worldwide. The disease is characterized by vomiting and watery diarrhea for three to eight days, and fever and abdominal pain frequently occur. Repeat infections can occur, but they tend to be less severe than the original infection. A series of three doses of the rotavirus vaccine is recommended in children at ages 2, 4 and 6 months. Infants diagnosed with severe combined immunodeficiency syndrome (SCID) should not receive rotavirus vaccine.

**Influenza (nasal spray).** The nasal spray flu vaccine (sometimes called LAIV for live attenuated influenza vaccine) is a vaccine made with live, weakened viruses that cannot grow at normal body temperature. Given via a nasal sprayer, this vaccine was approved for seasonal influenza viruses in 2003 and tens of millions of doses of the vaccine have been given in the United States. LAIV is approved for use in healthy people 2 to 49 years of age who are not pregnant. People with illnesses that weaken the immune system, or who take medications that can weaken the immune system, should not be vaccinated with LAIV.

Inactivated (killed) vaccines cannot cause an infection, but they still can stimulate a protective immune response. Viruses are inactivated with chemicals such as formaldehyde. Inactivated vaccines include:

**Influenza.** Because flu viruses change all the time, flu vaccine is reformulated every year to provide protection against the three most prevalent virus strains in circulation. Flu shots are needed between September and mid-November each year to give an individual’s body time to build the proper defense. The flu shot both prevents and controls the flu.

**Polio.** While polio has been eliminated from the Western Hemisphere, it has not been eradicated in the rest of the world. Most children are vaccinated against polio in four different doses, the first three before 18 months of age, and the last before 6 years old. Teens who have not completed their series of polio vaccines and are not yet 18 years old are advised to complete them.

Conjugate vaccines are made by using only parts of the
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viruses or bacteria. These vaccines cannot cause disease, but they can stimulate the body to produce an immune response that protects against infection with the whole germ. Five of the newest vaccines are made this way:

**Pneumococcal disease.** Pneumonia is a serious disease affecting the lungs, and the bacteria that form from this disease can attack other parts of the body, including the brain, which can cause meningitis. The Centers for Disease Control and Prevention (CDC) recommends that people over the age of 65 get the pneumococcal vaccine one time. If an individual has received this vaccine before age 65, and it has been longer than five years, the CDC recommends a second shot. Individuals ages 2 through 64 who have diabetes or chronic heart, lung, liver or kidney disorders should also get a pneumococcal vaccine.

**Haemophilus influenzae type b.** Hib disease, caused by a type of bacteria, usually strikes children under 5 years old. A child can contract Hib disease by being around other infected children or adults, as the germs spread from person to person. If the germs stay in a child’s nose and throat, the child probably will not get sick. But if the germs spread into the lungs or the bloodstream, they can cause serious problems. Before the vaccine, Hib disease was the leading cause of bacterial meningitis among children under 5 years old in the United States. All children should be immunized with the Hib vaccine. And while children over 5 years old usually do not need Hib vaccine, some older children or adults with special health conditions should get it.

**Meningococcal disease.** Meningitis has seen a resurgence in the U.S. Approximately 2,600 people in the U.S. are diagnosed with meningitis, and about 10 percent to 15 percent of those people die. Meningitis is a viral or bacterial infection of the fluid of a person’s spinal cord and the fluid that surrounds the brain. It is spread through the exchange of respiratory and throat secretions (i.e., coughing, kissing). Symptoms include high fever, headache and stiff neck, which can develop over several hours, or they may take one to two days to appear. Other symptoms may include nausea, vomiting, discomfort looking into bright lights, confusion and sleepiness. All young people ages 11 through 18, as well as college freshmen living in dormitories and individuals with special medical conditions, should be vaccinated against this disease.

**Hepatitis A and B.** Approximately 12.5 million Americans have been infected with hepatitis B virus at some point in their lifetime, and about 5,000 people die each year from hepatitis B-related liver disease. The number of new infections per year is declining due to routine hepatitis B vaccination in children and adolescents. But, even if vaccinated as a child, vaccines to protect against hepatitis A and B are recommended for individuals in high-risk groups, such as healthcare workers, those who live in households and/or have sex with people with chronic hepatitis B, those with multiple sex partners, people with a recently acquired sexually transmitted disease, men who have sex with men, and injecting drug users.

**Human papillomavirus.** HPV, known to cause cervical cancer, is now believed to cause other women’s cancers like vulvar and vaginal. The three-dose HPV vaccine is recommended for all females ages 11 to 26, and could be given to females as young as 9 years old.

Toxoid vaccines are made by treating toxins (or poisons) produced by germs with heat or chemicals, such as formalin, to destroy their ability to cause illness. Even though toxoids do not cause disease, they stimulate the body to produce protective immunity just like the germs’ natural toxins. These include:

**Diphtheria, tetanus and pertussis.** While tetanus is not spread from person to person, it is caused by a toxin that enters through the skin. Common signs include headache and muscle stiffness in the jaw initially, and then stiffness in the neck, difficulty swallowing, muscle spasms, sweating and fever. Diphtheria, on the other hand, is also caused by a toxin, but it can also spread from an infected person to the nose or throat of others. In some instances, it can lead to breathing problems, heart failure, paralysis and some-
times death; in others, it can cause sores on the skin that are painful, red and swollen. Pertussis (whooping cough) has recently been on the rise, and is easily spread and most dangerous in babies younger than 1 year old. In 2008, 19,000 whooping cough cases in adolescents and adults were reported to the CDC. Most children receive a combined diphtheria-tetanus-pertussis (DTP) vaccine, and adolescents and adults need a Tdap vaccine followed by a Td booster every 10 years.

Many organizations, including the CDC, Immunization Action Coalition, American Academy of Pediatrics and most state health departments, publish immunization schedules. While some are more in-depth than others and list medical conditions as factors, all follow the same general guidelines.

**Keeping Up-to-Date Records**

No central repository of records exists, so to keep current on screenings and vaccinations, patients need to keep up-to-date records. Some PCPs encourage patients to keep records by providing vaccination record cards for patients to track immunizations and the dates they were received. This card should be kept in a safe place, while a copy of the card should be kept handy in a purse or wallet.

> **When the lab tech is drawing blood, patients should request that their PCP be sent copies of all lab results.**

If individuals can’t locate a record of whether they’ve been immunized for a particular disease, “there is a blood test called the antibody titer that can detect the presence of antibodies against the disease in question. If the level of these antibodies is high enough, it is a good indication that they have immunity to the disease and do not need another vaccination.”

Blood work also should be logged. When the lab tech is drawing blood, patients should request that their PCP be sent copies of all lab results. By writing both the PCP’s and specialist’s fax numbers on the lab sheet, patients can help to ensure the lab sends the results to both physicians. This will also help to decrease the risk of unnecessary duplicate testing at the well visit.

A free service from Google — Google Health — may offer an easy solution to maintaining immunization records, as well as a complete repository for all kinds of personal health information. At [www.google.com/health](http://www.google.com/health), it’s possible to create online personal health profiles, including health conditions, medications, allergies and lab results by importing medical records from hospitals and pharmacies. Sharing health records within an approved care network is another feature, as well as browsing an online health services directory to find services that are integrated with Google Health. Competitor offerings include Microsoft’s HealthVault and the Indivo project (an open-source project managed by the Children’s Hospital Informatics Program).

**The Patient as Advocate**

The need for regular well visits, in addition to specialist visits, to ensure overall good health cannot be overemphasized for patients with immune-mediated diseases. This dual supervision makes certain that the whole person is treated, rather than just the chronic illness. Routine screenings and immunizations at well visits help keep routine, simple problems from being overlooked. They also can help to prevent serious infections in chronically ill patients.

Maintaining good communication between the patient’s PCP and “ologists” is essential to keeping the PCP current with the patient’s chronic illness, plus helps to eliminate duplicate testing and, even, medication errors. But, patients need to take responsibility, too. They must act as their own advocate, making regular appointments, encouraging communication between physicians, asking questions and maintaining up-to-date and accurate healthcare records.

RONALE TUCKER RHODES, MS, is the editor of IG Living magazine, and Kris McFalls is IG Living’s full-time patient advocate.

**References**

Individuals with CIDP can improve function and mobility by engaging in appropriate exercises for the upper and lower extremities.

By Matthew David Hansen, DPT, MPT, BSPTS

Anyone who has experienced local anesthesia or has had an arm or leg “fall asleep” knows how frustrating loss of feeling and/or impaired control of a body part can be, even for a short while. Our brains have an inherent desire to know the status of our body parts at all times. When something is preventing — or interfering with — the signals that travel through our nervous system to and from our brain, it can have a dramatic effect on our behavior and our ability to function.

Think about the last time that you were given a shot of Novocain before having dental work performed. Can you remember what you probably began doing even before you left the office? You might have run your tongue along the side of your mouth that was numb; rubbed, pinched or tapped your face; or even gently bit your cheek to see how hard you could squeeze before feeling something. Why? The impulse comes from our brains as they try to figure out what’s going on and to regain control of the situation. But, drawing a comparison between a trip to the dentist and the symptoms experienced by someone with chronic inflammatory demyelinating polyneuropathy (CIDP) is by no means a fair association. However, it may help those who have never suffered from a neuropathy to have some concept of what is experienced day after day by someone who has.

What Is CIDP?

CIDP, commonly considered the chronic equivalent of a similar condition named Guillain-Barré syndrome, is a disease of the peripheral nervous system that is caused by an abnormal immune response that mounts an attack on myelin (a fatty covering that protects nerve fibers and allows for a signal to be relayed quickly). As a result of the lost myelin, the affected nerves respond weakly or not at all to stimuli, resulting in progressive muscle weakness, fatigue, loss of deep tendon reflexes and atypical nerve sensations (tingling, burning, numbness and/or pain). Symptoms are usually symmetrical and frequently cause difficulties with walking and the coordination of other movements. The autonomic nervous system also may be
involved, leading to complaints of dizziness when changing positions, heart symptoms and trouble with bowel and bladder function.

In most patients, the course of CIDP is slowly progressive; however, it is not uncommon for periods of recovery, lasting weeks to months, to occur between relapses. Although there is currently no known cure for CIDP, symptoms can be treated via corticosteroids to reduce inflammation, plasmapheresis to remove harmful antibodies from the blood, intravenous immune globulin (IVIG), immunosuppressant drugs (in some severe cases) and exercise. Early medical treatment is important to confine nerve damage to the myelin sheath and to prevent harm to the axons (nerve fibers) themselves.

**How Exercise Can Help**

Appropriate exercise is a vital part of any CIDP intervention plan because of its potential to improve strength and endurance, thereby minimizing muscle shrinkage and improving function and mobility. Understanding some of the recommendations that have emerged from scientific research for those exercising with a peripheral neuropathy can help to establish a proper program.

First, patients should always visit with their medical doctor before beginning an exercise regimen. This is an important principle for any population; however, it is even more essential for those with a peripheral neuropathy, because the wrong exercise parameters can actually make a bad situation worse rather than better. The possibility of the autonomic nervous system being involved also means that the body may not respond to exercise in a typical manner.

Second, patients shouldn’t overdo it! The damage caused by CIDP to myelin, and the possible damage to axons, results in the body’s ability to recruit fewer muscle fibers to perform a task. Consequently, those muscle fibers that are engaged are at greater risk of being overworked. Some soreness after exercise may be expected, but it should dissipate within 12 to 48 hours. If pain persists, is exaggerated or is coupled by a loss in strength, the patient likely did too much.

Submaximal exercise is frequently recommended for peripheral neuropathies. A doctor and/or a properly trained physical therapist can help patients find the exercise prescription (frequency, intensity, time and type of activity, known as the F.I.T.T. principle) that is currently right for them. Low-impact exercises like walking, swimming, riding a recumbent bike or performing “open-chain” arm and leg exercises (without bearing weight through the extremity) might also be good alternatives to high-impact activities like running or jumping.

Third, patients need to be aware of their physical limitations. Activities that put them at undue risk of falling or other physical injury should be avoided. And, they shouldn’t hesitate to ask someone to help, or at least accompany, them during their workouts.

Fourth, muscle strengthening and aerobic conditioning are important. Science has demonstrated that strength exercise programs can improve muscle force in patients with peripheral neuropathies. However, it also has been shown that aerobic conditioning is important in combating fatigue and other impairments, particularly in the later stages of recovery.

And last, patients should wait until a muscle can work against gravity before stressing it against additional resistance. Fortunately, under normal circumstances, myelin and peripheral nerve fibers can regenerate, with muscle control gradually returning as it does. However, with CIDP, residual damage is not uncommon and recoveries can take some time. Therefore, it is important to progress exercises in a systematic way in order to avoid overstressing muscles and joints.

**Appropriate exercise is a vital part of any CIDP intervention plan because of its potential to improve strength and endurance, thereby minimizing muscle shrinkage and improving function and mobility.**

**Choosing the Appropriate Level of Exercise**

Those experiencing an immunological disease exacerbation probably find it difficult to imagine themselves exercising. The biggest hindrance may not be the weakness they are experiencing, but instead, the popular misconception that exercising means performing a workout à la Jane Fonda,
Billy Blanks or even Richard Simmons. The reality is that there are multiple levels of exercise difficulty, each as achievable and as genuine a workout for those to whom they are prescribed as a typical exercise video would be for a fully able-bodied recreational athlete. For CIDP patients, the following exercise progression levels can be used for particular exercises, but which level is appropriate will depend upon what the patient is ready for.

Passive exercise: Gentle movement of the body (usually the limbs) is performed by a properly trained individual, without effort on the patient’s part. Passive movement can be beneficial for maintaining or improving blood circulation and range of motion. Thinking about the movement and trying to assist may also help to re-establish nerve connections in cases where actual damage has occurred to the nerve and regeneration is under way.

Active-assisted exercise: Assistance is still required from another person, but the patient is able to participate in movement to some degree. Actual activation of the muscle(s) is occurring; however, it is still not strong enough to move the limb independently.

Active exercise (gravity eliminated): Independent movement is possible in a gravity “eliminated” position, but not against gravity. For example, a patient may be able to lift their knee toward their chest (hip flexion) while lying on their side in bed (gravity eliminated position), but not while standing (against gravity).

Active exercise (gravity reduced): Movement is possible against some gravity, but not against its full pull. To use the same example of hip flexion, a patient may be able to bring a knee toward their chest while lying on their back (gravity reduced), but not while standing (against gravity).

Active exercise (against gravity): Movement is possible in all planes (including standing for hip flexion), but without additional resistance.

Resistive exercise: The highest level of progression, but also the most variable level, limited only by the potential of the conditioned human body to produce force. Resistance may take the form of weights, resistive bands, household items, one’s own body weight, etc.

**Lower-extremity exercises.** The lower-extremity exercises predominantly involve the hip, knee and ankle. Hip flexion (Figures 1-4) is the action performed when lifting the leg to walk forward or step up onto something. In addition to hip flexion, two other hip actions — extension and abduction — are fundamental to an individual’s ability to walk and maintain balance.

- **Figure 1.** Active-Assisted or Passive Hip Flexion
- **Figure 2.** Active Hip Flexion while lying on your side (Gravity Eliminated)
- **Figure 3.** Active Hip Flexion (Gravity Reduced)
- **Figure 4.** Active Hip Flexion (Against Gravity)

**Lower- and Upper-Extremity Exercises**

There are several upper- and lower-extremity exercises for some of the most important gross motor (large muscle) actions performed by the body. The figures in this article provide one example of the progression levels for each exercise. These exercises also can be performed while patients are lying on their back, stomach, sides, sitting and standing.
Hip extension (Figure 5) is the motion that helps to pull the leg back and propel the body forward after taking a step. Hip and knee extension also are needed to stand up from a chair, the floor or a squatting position, to jump or to climb stairs. For instance, an individual lifts their foot to the next step via hip flexion, but they progress, or pull, themselves up to the step via hip extension.

Hip abduction (Figure 6) is most easily visualized as an open-chain exercise. However, the prime muscle of hip abduction, the gluteus medius, performs most of its work in a weight-bearing mode as a “closed chain” exercise when the leg may not even be moving. To experience this phenomenon, place an open hand over your hip; not the boney area at your waist line (that’s part of your pelvis), but the area just below it. Now, if you are able to, lift the opposite leg (while stabilizing yourself against something with your free hand) so that you are standing on one foot. Did you feel anything happen under the hand that is placed over the hip? That’s the gluteus medius contracting. If it didn’t, your body would fall to the side of the leg that is being held up off of the floor.

The inability to dorsiflex the ankle/foot (Figure 8) is a frequent complication of peripheral neuropathies. The condition becomes especially troublesome when the toe drags or catches the floor when walking, causing a patient to stumble or making it impossible to advance the limb forward without using an assistive device and/or dramatically changing the way that they walk.

Besides its use in standing, jumping and climbing, knee extension (Figure 7) also is the motion of kicking — perhaps not something that individuals still do every day, but it was probably more important to people at some time during their lives!

Upper-extremity exercises. The upper-extremity exercises predominantly involve the shoulder, elbow and wrist. Shoulder flexion (Figure 9) is the chief shoulder action used by individuals to reach for something in front of their body or over their head (such as shaking someone’s hand or getting something down from a shelf).
HIGHLIGHTS OF PRESCRIBING INFORMATION

These highlights do not include all the information needed to use GAMUNEX®, Immune Globulin Intravenous (Human), 10% Caprylate/Chromatography Purified, safely and effectively. See full prescribing information for GAMUNEX.

GAMUNEX (Immune Globulin Intravenous [Human], 10% Caprylate/Chromatography Purified) 10% Liquid Preparation

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 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. GAMUNEX does not contain sucrose.
- Administer IGIV products at the minimum concentration available and the minimum infusion rate practicable.

INDICATIONS AND USAGE

GAMUNEX is an immune globulin intravenous (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. Epinephrine should be 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.
- 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 GAMUNEX 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 infectious agents, e.g., viruses and, theoretically, the Creutzfeldt-Jakob disease agent.

ADVERSE REACTIONS

- PI – Most common drug related adverse reactions during clinical trials were headache and cough.
- ITP – Most common drug related adverse reactions during clinical trials were headache, vomiting, fever, and nausea.
- CIDP – Most common drug related adverse reactions during clinical trials were headache and fever.

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

USE IN SPECIFIC POPULATIONS

- In patients over age 65 or in any patient at risk of developing renal insufficiency, do not exceed the recommended dose, and infuse GAMUNEX at the minimum infusion rate practicable.
- Pregnancy: no human or animal data. Use only if clearly needed.
Important Safety Information for GAMUNEX

Gamunex, Immune Globulin Intravenous (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).

Immune Globulin Intravenous (Human) products have been reported to be associated with renal dysfunction, acute renal failure, osmotic nephrosis and death. Patients predisposed to acute renal failure include patients with any degree of pre-existing renal insufficiency, diabetes mellitus, age greater than 65, volume depletion, sepsis, paraproteinemia, or patients receiving known nephrotoxic drugs. Especially in such patients, IGIV products should be administered at the minimum concentration available and the minimum rate of infusion practicable. While these reports of renal dysfunction and acute renal failure have been associated with the use of many of the licensed IGIV products, those containing sucrose as a stabilizer accounted for a disproportionate share of the total number. Gamunex does not contain sucrose. Glycine, a natural amino acid, is used as a stabilizer.

Gamunex 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.

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

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, and/or known or suspected hyperviscosity. Hyperproteinemia, increased serum viscosity, and hyponatremia may occur in patients receiving IGIV therapy.

Gamunex 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.

In clinical studies, the most common adverse reactions with Gamunex were headache, fever, chills, hypertension, rash, nausea, and asthenia (in CIDP); headache, cough, injection site reaction, nausea, pharyngitis, and urticaria (in PI); and headache, vomiting, fever, nausea, back pain, and rash (in ITP). The most serious adverse reactions 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 full Prescribing Information.
Elbow flexion (Figure 10) is used to lift and carry (such as carrying a box) and to bring objects that are grasped closer to the body. It also is used to simply lift something to one’s mouth to eat or drink.

There are also many fine motor (small hand muscle) exercises that can be performed to increase grip strength and improve precision handling of objects. For instance, the wrist extension (Figure 12) is a functional position used while eating, handwriting, keyboarding, driving, grasping objects and performing a number of other tasks.

Tailoring the Program to Each Individual

The muscles targeted by the exercises presented in this article are just a few of the 640 skeletal muscles found in the body, but they are some of the most important to everyday function. While it’s true that CIDP and other immunological diseases certainly can be disabling, appropriate exercise to improve strength and endurance can lead to better daily function. Individuals with CIDP are capable of doing a lot, and there is a level of exercise that’s right for everyone!

References


MATTHEW DAVID HANSEN, DPT, MPT, BSPTS, is a practicing physical therapist in Utah, and president of a healthcare consulting and fitness product company, SOMA Health, LLC. The company’s latest product, the Freedom2Move exercise program and video series, was inspired by an IG Living Readers Teleconference. Matt completed his formal education at the University of Utah in Salt Lake City.

Illustrations by Veronica Hansen
A Review of Medicare and Disability Programs

People with chronic illness who can no longer work have options for receiving disability payments. Here’s how to get them and what to expect.

By Jennifer Kester

Cathy Vargas worked full time as a legal secretary, part time as a real estate agent and, somehow, also found time to be a coordinator for the Special Olympics on the side. But when the 50-year-old was diagnosed with hypogammaglobulinemia, her busy work life came to a halt. Having already exhausted her time off through the Family Medical Leave Act, she was told by her employer that if she got sick again within the next 30 days, she’d be fired. Her place of work offered her an alternative: long-term disability.

Many people with a chronic disease, both young and old, find themselves unable to work and have to turn to disability benefits. According to the Social Security Administration (SSA), more than three million Americans will apply for disability benefits this year. Knowing what to expect can help.

What Benefits Are Available?

The SSA will deem individuals disabled if they are not working or if they are earning less than $1,000 a month; if they have a severe condition that hinders basic work-related activities; and if their affliction is on its list of medical conditions. If the medical condition isn’t on that list, individuals must prove that the condition interferes with their ability to perform the work they did previously and that they’re unable to adjust to other tasks.

If individuals meet the criteria for a disabled person, there are two ways they can receive benefits. Supplemental Security Income (SSI) pays money to disabled adults and children who have limited income and resources. Those 65 and older without disabilities who are in financial need also can qualify for the monthly payments. Social Security Disability Insurance (SSDI)
provides benefits to disabled people who are “insured” through workers’ contributions to the Social Security trust fund. The difference between the two is that SSI is paid based on financial need, and SSDI is paid only to those who worked long enough and paid Social Security taxes.

The formula for payment differs between the two as well. The SSI amount is calculated using the Federal Benefit Rate (for 2010, it’s $674 for a qualified recipient), and the SSDI figure is based on the worker’s lifetime average earnings covered by Social Security. For SSI, the health insurance coverage provided is Medicaid, while those under SSDI will qualify for Medicare. One last key difference is that many states will give SSI recipients an added state supplement, whereas there aren’t any extras for those receiving SSDI.

How to Apply for Benefits

It can be tricky for individuals to figure out whether they are eligible to receive SSI or SSDI. Some help can be found with Social Security’s Benefit Eligibility Screening Tool at http://connections.govbenefits.gov/ssa_en.portal. Based on their answers to the online questionnaire, the tool will explain which benefits they might be eligible to receive, as well as how to qualify and apply for them.

When ready to apply for benefits, they will need to complete an application for Social Security benefits and an Adult Disability Report, both of which are available online (http://www.socialsecurity.gov/applyfordisability/adult.htm). Also on that site is a comprehensive checklist of the items needed for their application. This includes things like the names, addresses and phone numbers of all doctors and hospitals visited; birth certificate; and a W-2 form from the previous year.

Individuals can apply online for SSDI at https://secure.ssa.gov/apps6z/iClaim/dib. If help is needed, they can call Social Security at (800) 772-1213 to have someone take their application over the phone or to make an appointment for in-person assistance. For SSI, a large portion of the application can be completed at www.socialsecurity.gov. However, it is necessary to call (800) 772-1213 to make an appointment with a Social Security representative for help in person or over the phone.

If professional help is needed to navigate through all of the paperwork, hiring a disability lawyer is a viable option. However, anyone who is familiar with the rules and the system can be an advocate and help with the paperwork. When Vargas had to apply for long-term disability benefits, she wanted to avoid what her sister had gone through. Battling systemic lupus, her sister was caught in the cycle of getting a job, getting sick and then getting fired for taking too much time off to tend to her health. Vargas’ sister applied for disability twice and was denied both times. On her third try, she hired an attorney who helped her win approval. “I didn’t want the trouble she had to go through,” Vargas says. “And, I didn’t want to have to apply for work and get fired. So, I hired an attorney who specializes in Social Security disability law, and he won my case the first time around.”

How Benefits Are Approved or Denied

If individuals meet the basic requirements, the Social Security Administration will send their application to their state’s Disability Determination Services, which determines whether to classify them as disabled. The state agency will review the application and speak with their doctors. But, sometimes, the state agency needs more medical information before it makes its ruling, so it may ask applicants to go for a special examination. While they may be able to go to their own doctors, sometimes the exam will have to be performed by someone else. Social Security will pay for the exam, as well as some of the related travel expenses.

It typically takes the state agency three to five months to come to a decision, depending on the time it takes to access medical records or other needed evidence. If approved, applicants can expect to receive monthly SSI payments of $674 for an individual and $1,011 for a couple.

Applicants shouldn’t be discouraged if they’re denied benefits on the first try. The approval rate for initial disability applications is about 37 percent, according to the National Press Office of the Social Security Administration. The decision merely needs to be appealed. If going that route,
hiring a lawyer is especially recommended. “Hearings are like court, so a lawyer’s skill and experience will help,” says Jennifer Jaff, executive director of Advocacy for Patients with Chronic Illness Inc.

What to Expect After Approval

Once individuals are approved, there is still some bumpy road ahead. For instance, Social Security disability benefits will be paid beginning with the sixth full month after the date disability begins. And, individuals will automatically be enrolled in Medicare after they receive disability benefits for two years, dating from the month they became entitled to receive disability. However, during this waiting period, they will not be entitled to healthcare benefits. According to Jaff, what people do in the interim depends on their circumstances; some have spouses with insurance, and others get Medicaid or purchase COBRA, a costly temporary continuation of health insurance only available to some. However, the sad reality is that a lot of people go without coverage during that time.

During Vargas’ two-year waiting period, she purchased COBRA insurance, but after two months, she had to cancel it. “I couldn’t make the $450-a-month premium,” she says, “I just couldn’t afford it.” Instead, the Florence, Ariz., resident turned to the Arizona Health Care Cost Containment System (AHCCCS), the state-run healthcare program for low-income residents.

For others who are facing similar problems of keeping up with healthcare premiums and co-payments, Vargas recommends they check out Patient Services Inc. (PSI) at www.patientservicesinc.org, a charitable organization that helps people with chronic illnesses make these payments. PSI solicits donations to fund thousands of patients and their families every year. And, if high premiums mean having to skip treatments, Vargas suggests individuals appeal to the pharmaceutical company that makes their treatment drugs. This is especially true for patients with chronic illness who rely on immune globulin (IG) treatments. Many IG manufacturers offer patient-assistance programs. Vargas approached her drug company for assistance, and it was ready to help her out, but her insurance kicked in before she needed the company’s aid.

Other problems also can crop up. For example, when Vargas did finally receive her Social Security benefits, her long-term disability checks were decreased with no warning, something she says is commonplace in the Arizona state system. Again, she had to hire a lawyer to help her reinstate her full benefits, and it took six months to get them back. “In order to keep my house and pay my insurance and my attorney, I had to borrow from my children, my parents and my boyfriend,” she says. Later, she found out she was eligible for free legal services through the state. So, individuals should be sure to check if their state offers similar complimentary legal aid.

If individuals meet the criteria for a disabled person, there are two ways they can receive benefits.

A problem for those who rely on IG is that Medicare part B covers only 80 percent of the drug cost. Individuals can try to buy a Medigap policy to cover the other 20 percent, but people with pre-existing conditions aren’t eligible for Medigap policies in all states. An alternative is to buy a Medicare Advantage plan (HMO) to cover the whole thing, Jaff says.

Not Welcome Choices

SSI and SSDI are not welcome choices for those with chronic illnesses; they are alternatives people turn to for help when working is no longer an option. Unfortunately, patients typically have to deal with a stressful, difficult approval process — one that could leave them bankrupt in the struggle to get their benefits. And, once they do get approval for benefits, they aren’t set for life. Most people have to go through the process of again proving they are, indeed, disabled.

“I made it,” says Vargas. “I thank God every day, and people like my family and others who were there along the way.” As for the future, she’s fortunate to have a strong support system that will stick by her because Vargas’ lawyer thinks it’s likely she’ll have to fight for her benefits again in 2015.

JENNIFER KESTER is a San Diego-based writer and editor specializing in health and lifestyle issues.
After James and Connie Ramos’ two children were diagnosed with primary immunodeficiency (PIDD), the couple committed to learning everything they could about the disease. Then, they went a step further by dedicating themselves to funding research and expanding awareness to help other families facing similar challenges. With the support of their physician and All Children’s Hospital in St. Petersburg, Fla., James and Connie established the Southeast Primary Immunodeficiency Network (SEPIN) under the umbrella of the Jeffrey Modell Foundation. Starting a nonprofit organization is a huge undertaking for anyone; for the parents of two chronically ill children under the age of 10, it’s positively heroic.

We chatted with Connie to find out what fuels this wonder mom.

**Trudie:** Tell me about your family’s journey with PIDD.

**Connie:** When my daughter, Abigail, who is now 8, was born, it took a long time to confirm her diagnosis. She had severe food allergies, chronic ear infections and was always vomiting as an infant. As a new mom, I knew something was not right, but we went from doctor to doctor. She was constantly on antibiotics and just didn’t get any better. Abby was finally diagnosed with consumption immunodepression when she was nearly 3. I became pregnant with my son, Aidan, a short time later.

**Trudie:** Was Aidan sick, too?

**Connie:** Aidan was even sicker than Abby, but this time I started seeing a specialist right away. That’s when we met Dr. John Sleasman, professor and chief of the division of allergy, immunology and rheumatology at the University of South Florida’s Department of Pediatrics. He told us it was no coincidence that both of our children were sick; there was a genetic link. I was so relieved when he confirmed what I suspected. Finally, we began to get some answers.

**Trudie:** What did you find out?

**Connie:** After all the genetic blood work came back, we learned the defect came from my husband, James. As a result, both of our children have the positive trait.

**Trudie:** Are both of your children on intravenous immune globulin (IVIG) now?

**Connie:** No, just Aidan. IVIG has turned his life around, and it’s been amazing to watch him go from struggling to thriving.

**Trudie:** Tell me how you went from being the mother of two chronically ill children to becoming an advocate and fundraiser for immune deficiency awareness.

St. Petersburg, Fla., Mayor Mike Finnerty presented a fund-raising check to the Southeast Primary Immunodeficiency Network at the 2009 Wiffle ball tournament.

**Connie:** After Aidan was diagnosed and began to get stronger and healthier, I pulled myself together and said, “We need to do something about this.” It was so frustrating to go as long as we did without an accurate diagnosis, and James and I didn’t want other parents to have to
go through that.

**Trudie:** How did you establish the Southeast Primary Immunodeficiency Network?

**Connie:** I began discussing the need for an advocacy group with James and Dr. Sleasman. Before I knew it, I had a team on board. Parental passion was a big driving force, and I have to give a lot of credit to James. He’s someone who can break through obstacles and doesn’t take no for an answer.

**Trudie:** How did you arrive at the idea of a Wiffle ball fund-raiser?

**Connie:** I know it sounds funny, but James and his best friend, Derek, grew up playing Wiffle ball every July 4th. When we were in college, there would sometimes be 300 to 400 people on the beach in front of his parents’ home, and we’d play for money. James thought it would be a great way to raise money for immunology. Believe me, there was a lot of skepticism at first, especially at the children’s hospital — we really had to sell them on the idea.

**Trudie:** What challenges did you overcome to pull off your first fund-raiser?

**Connie:** First, we had to pin down the location. The hotel we selected loved the idea but did not want to host it on the 4th of July — their biggest travel weekend. Once they agreed to our date, James met with the mayor and some of the local media to get their support. The next thing I knew, I was on a plane to New York to meet with Vicki and Fred Modell at the Jeffrey Modell Foundation to ask for their support. Fred felt an immediate connection with our efforts — they really support community-based events. They put their confidence in us and were kind enough to sponsor one of the kids’ tents for our fund-raiser.

**Trudie:** Was the fund-raiser a success?

**Connie:** The turnout was fantastic, and we raised $85,000, which far exceeded our hopes and expectations. It was a phenomenal amount of work from January until June, and it was well worth the effort.

**Trudie:** What’s next for you?

**Connie:** We hosted the Wiffle ball tournament again this year at the same location. We’d love to see it become an annual event. Our main goal is to raise funds to support research and development and to increase awareness about immune deficiency. Ten years ago, I knew nothing about immunology or immune deficiency because it didn’t affect me personally. But when life throws things at you, you have to embrace it and take it on. That’s what we’ve done.

**Trudie:** Any advice for someone who would like to become more involved in advocacy?

**Connie:** I think you start with the people around you, whether you live in a small community or a big city. Get involved in your church; talk to your neighbors. There’s power in telling your story. We have to stop viewing IVIG as a negative topic — this is something that improves health and changes lives.

**Making a Difference with Wiffle Ball**

In 2009, the creators of the Southeast Primary Immunodeficiency Network, James and Connie Ramos, held their first Wiffle ball tournament to raise money for PIDD in St. Petersburg, Fla. The 2009 Wiffle ball tournament was able to fund a comfort and fun area for children at the brand-new Infusion Center at All Children’s Hospital; a community education and awareness campaign sponsorship at the new Glazer Children’s Museum in downtown Tampa, Fla.; and funding for a genetic analysis study focused on patients with PIDD. In May of this year, a Jeffrey Modell Diagnostic and Research Center was dedicated at All Children’s Hospital, one of 72 prestigious centers in the world focused on the earliest diagnosis of PIDD. The second annual Wiffle ball tournament took place on July 4, 2010, at the Don CeSar Beach Resort at St. Pete Beach in St. Petersburg, Fla. Activities included an old-fashioned barbecue, kids’ games, fireworks and, of course, Wiffle ball. For more information about the Southeast Primary Immunodeficiency Network and to find out about the funds raised in this year’s event, visit www.medicalchampion.com. 

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The first Wiffle ball tournament held July 4th at St. Pete Beach in St. Petersburg, Fla., raised $85,000 for PIDD.

**TRUDIE MITSCHANG** is a staff writer for IG Living magazine.
Reader: Why is exercise important for myositis patients, and who decides the types of beneficial exercises?

Kris: No matter what your level of fitness, anyone can benefit from some form of exercise. According to the Mayo Clinic, exercise can positively affect your mood, sleep and help control chronic disease, among other things. Keep in mind that when starting any new exercise routine, it is always best to check with your doctor first. For more specifics on how exercise relates to myositis, I asked Matthew Hansen, doctor of physical therapy, to answer your question.

Matthew Hansen: Myositis is characterized by progressive weakening and possible shortening of the muscles. Exercise is an important component of a patient's treatment regimen. It can help preserve strength and functional skills (e.g., getting up from a chair, climbing stairs and getting dressed), control weight gain and loss of bone mass as a consequence of prescribed corticosteroids, maintain respiratory health and provide an important boost to psychological morale.

After being diagnosed with myositis, it is important for a patient to consult with a medical specialist before beginning an exercise program. For a patient undergoing early treatment with corticosteroids or other medications, muscles can be particularly prone to damage from strenuous activity, and overdoing it at any point in the disease process can actually increase weakness to an already damaged muscle unit. Myositis symptoms and response to medications vary, so treatment must be individualized. Your doctor will be able to tell you when it is appropriate to start an exercise program, while a qualified physical therapist can help you to select exercises and monitor your routine. However, even when exercise should be limited, it is important to learn proper stretching techniques to prevent loss of flexibility in the joints and muscles.

Reader: My daughter will graduate college in the spring and will no longer be covered by my husband's health insurance. She requires Vivaglobin infusions to keep her healthy. What are my options?

Kris: You are wise to plan ahead. My boys are in the same stage of life, so I very much understand your concerns. I would advise you and your daughter to research this issue together. It is, after all, her disease, and she is going to have to take it with her wherever she goes.

The U.S. Department of Labor publishes a booklet titled An Employee's Guide to Health Benefits Under COBRA. On page 16, it states that a dependent child who loses his or her dependent child status may purchase COBRA insurance for up to 36 months. Your daughter would have to pay the monthly premiums, but that would be cheaper than paying for her medication. It would also give her time to find a job that will provide her with health insurance benefits. To find out what the monthly premiums would be, you will need to contact your husband's employer's human resource department that handles your daughter's insurance.

Because your daughter is prescribed Vivaglobin, there is an added layer of protection for which she would qualify. CSL Behring, manufacturer of Vivaglobin, offers a program that will provide the medication free of charge in the event there is a gap in insurance. To learn more about that program, go to https://www.cslbehringassurance.com/cslbehring_enu/start.swe?SWECmd=Start&SWEHo=www.cslbehringassurance.com.

It is great that your daughter has such wonderful support. I wish you both a bright future.

Kris McFalls has two adult sons with chronic diseases treated with IG. She is formerly a physical therapist assistant, and currently is IG Living's full-time patient advocate.

Matthew Hansen, DPT, MPT, BSPTS, is a practicing physical therapist in Washington state and president of an allied healthcare staffing and consulting agency.
RELATIONSHIPS — some that we’ve had forever come with unconditional love and support. We make these connections with our best friends, our moms, our cousins, husbands or all of these. And, we consider ourselves really lucky to have so many people who support and love us, and in turn, we do the same for them. These people accompany us on infusion days with a suitcase full of snacks; we can call them at any time of night to share news or ask a question; they will drive us to any doctors’ appointments when we just don’t feel up to it; they are our backbone; and they make our lives better. These are the relationships we cherish and feel grateful to have even if there are times we might take them for granted.

But what about new relationships — the ones we have just made or will make in the future? How much do we share with these people? How long do we wait to tell them about the complications in our lives? The answer to this varies, depending upon whether dealing with a friend versus a potential boyfriend or girlfriend.

I remember when I told some people I had just met about having common variable immune deficiency, and they immediately asked if I had AIDS. I said no, and they asked if it was contagious. In my mind, I imagined hitting them upside the head. When I run into situations like this, I tend to lose a little faith in humanity. But, then I ground myself and allow the experience to be a reminder that not everyone is understanding or patient enough to listen to your explanation.

Dating, unfortunately, is a little different. From the perspective of someone who is chronically ill, it always feels as if there is a nurse with a needle in the room — until we tell them about our illness, of course. Dating can be awkward enough between two healthy people. So, as an ill person, I know that I have to be honest. The question is: When? My advice: If it is the first date, we don’t have to even bring up our illness. There are so many other things to talk about: hobbies, work, school, family, each other. At this point in the relationship, we don’t even know if we like them, and they don’t know if they like us. So, why sabotage something with potential, or confide such personal information with people we hardly know?

Some people with chronic illness would say, “If they don’t like me for all of me, then why bother?” Here is my answer: Give this person an opportunity to get to know you first. Unfortunately, not everyone responds well to illness; many are not as strong as we are. It’s like talking about an “ex” on the first date. When we do that, we close ourselves off, and we become difficult to reach. Plus, the date will be turned off. When we talk about an illness, our date may be scared, may feel sorry for us or may shut down because it is too much to deal with.

I think people with chronic illness should either wait until the third date or when we feel comfortable. By the third date, we know that we like each other; otherwise, why would we be going out again? Then, it should be explained simply, without much detail. Then, wait. If they are interested, they will ask questions to learn more. If they aren’t, then we shouldn’t waste our time — because while our illness isn’t all of us, it is still a part of us.

We date and build new relationships because we need to find people to share with — the ones who will be in our lives forever. It’s worth the time it takes to build lifelong relationships, especially for those of us with chronic illness; in fact, most of us will continue working at it for the rest of our lives. So, it seems only right that we give others time, too. We need to find the patience to allow those whom we want in our lives to have the time to form their bond with us and, of course, with our illness. We are worth it.

EVER FECSKE was diagnosed with CVID and interstitial lung disease in 2004. She is a fashion design student, loves spending time with her fiancé, family and bulldog, Dunkin, and can’t get enough of writing, cake decorating and anything that sparkles!
My husband, Mark, and I have learned a lot from raising three kids with primary immune disease. We learned how to hold Molly still while she had IVs placed in her feet. We learned how to control Caleb when he became impossibly strong when a needle was coming his way. We learned that too many fruit snacks turned Calvin’s poop an alien-ish shade of blue. And, as middle-age greeted me, we learned I inherited bad DNA and had my own immune issues.

But, please, don’t feel too sorry for me. My infusion nurse and I like to share celebrity gossip, I have been introduced to many new slow-cooker recipes from friends who bring dinner, and when I’m feeling crummy, I’ve learned naps are guiltless pleasures. One thing is for sure: I’ve learned that being sick at age 41 doesn’t mean I can’t have fun!

Suffering with never-ending strep throat, I recently found myself at the mercy of Dr. Carz, MD, ENT (etc.). He and nurse Belle were a well-oiled machine, peppering me with questions at lightning speed. With every
inquiry about my condition, I grew more confident with the pair. After all, they probably had seen a 41-year-old woman with icky tonsils a million times before.

“Well, now that that's over, let's take a look-see, shall we?” Dr. Carz said while shining the circle wrapped around his forehead in my direction. “Open, please.”

The chain of events that followed happened so fast I can barely recollect. What I can recall is that within seconds of Dr. Carz and nurse Belle leaving the room, I was scheduled for a tonsillectomy — at age 41.

I can only describe my experience as being much like someone hollowing out the middle of my mouth — as if I were a Halloween jack-o'-lantern. I'm sure that if you had lit a small candle and placed it on my tongue, my face would have lit up. The only food I could tolerate afterward was room-temperature Jell-O, which I stopped eating after one of my Facebook friends revealed what I truly was digesting.

By recovery day eight, I called my nurse and grumbled, “I can’t take this anymore.”

“You're dehydrated. Start drinking Gatorade and you'll start feeling better by day 10,” nurse Belle answered with soothing confidence as if she'd taken day-eight phone calls a million times.

“Thank you,” I strained. “Having a tonsillectomy at 41 is really not age-appropriate. It's more for age 4.” Nurse Belle laughed as if she'd heard my wisdom before.

Grape Gatorade quickly became my best friend. The silly concoction miraculously saw me to day 10 and beyond. And if I had not gone through the “too many fruit snacks” with Calvin as a baby, I would have taken myself to the emergency room after day 11’s toilet experience.

Fourteen days into my recovery, I had my post-operative appointment. And, by the late-in-the-day appointment, I had drunk, at minimum, four bottles of my favorite grape swirl.

“OK, Cheryl. Come on in,” nurse Belle invited and then made me as comfortable as possible on the cold examination table.

“I see you're enjoying Gatorade,” she mentioned.

“Yes! It's been a lifesaver!” I enthusiastically responded.

“And, here comes Dr. Carz,” she announced as the physician waltzed into the room.

“So, how are you feeling?” Dr. Carz asked while examining my neck.

“Much better after grape Gatorade came into my life,” I giggled.

I'm not sure he even heard me because of what happened next.

“Open, please,” he commanded.

I opened my mouth slowly as the shiny disc on Dr. Carz’s forehead caught the light and flashed past my eyes. And, as if it were in slow motion, Dr. Carz’s countenance became fear-stricken and his face became ashen.

“Are you OK, Ms. Haggard?” Dr. Carz asked, swallowing hard.

“I'm fine. I mean, considering what I've been through, I should be …” I replied.

Dr. Carz interrupted my comments and invited nurse Belle over to see what the fuss was all about.

A sinking feeling gripped my stomach. Dr. Carz scraped a bit of blue goo off my tongue and readied it for the lab. He then barked orders at nurse Belle, and before I knew it, I was whisked away to the waiting room.

“Dr. Carz wants you to stay right here until we hear back from the lab,” explained nurse Belle. “Make yourself comfortable until then. Can I bring you something to drink?”

It was as if my mind was screaming at me: “Wake up, Cheryl! Could it be the Gatorade?” I grabbed the blue bottle from my purse and read the ingredients to nurse Belle:

“Natural and artificial flavor, mono-potassium phosphate … red 40, blue 1!”

“Cheryl, I think we have our answer to the mysterious blue goo!” said nurse Belle.

Telling Dr. Carz about Calvin’s poop after too many fruit snacks, and my day-11 post-poop experience was actually kind of fun. Watching my well-seasoned physician's countenance quickly change from terror to tickled pink, I concluded I was not going to be discarded as “hazardous waste.” It also was a relief to all that my lab report concluded: No notable pathogen found, just copious amounts of blue 1.

And I swore I heard Dr. Carz mutter, “I really wish people would act their age.”

Names have been changed to protect the innocent.

**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.
LifeStyle

Transitions
By Ronale Tucker Rhodes, MS

"I JUST WISH I could fix Quentin — make it all go away and make things better for him," says Matahna Erickson about her 9-year-old son. Quentin was diagnosed with common variable immune disease (CVID) as a baby. "I noticed something was wrong at 3 months old, but I thought maybe it was just teething. At 6 months, I knew," explains Matahna. "So, I pursued getting him seen by some different doctors and finally got him diagnosed at 1 year." And while the diagnosis gave them answers, it was just the beginning of what has been a frustrating experience for the family: trying to make Quentin understand why he has to endure his lifesaving treatments when he doesn’t want to cooperate. It’s a situation that, no doubt, many families with immune-deficient children struggle with. Perhaps the Ericksons’ story can help them.

From Doctor’s Office to Homecare
After diagnosis, Quentin was placed on antibiotics for the first year and a half. At age 2-1/2, he had a central line placed and he had his first infusion. "At that point, it wasn’t too bad," says Matahna. "We tried to make it fun. We had great nurses, and they were able to access his line every time the first time." But, while at first Quentin didn’t mind too much going into the doctor’s office for his infusions, he gradually began to resent them and started to develop behavioral problems.

To make infusion days more tolerable, the Ericksons started making exceptions to their ‘rules’ that television and sugar were not regularly allowed. For instance, on infusion days, Quentin could pick a movie to bring with him to the doctor’s office to watch. He also would get to pick a special treat, such as cookies or brownies, and he helped his mother make them the day before. "We had a day that was all about him," explains Matahna. “Then, after the treatment was over, we would go to a park that he would pick out, and sometimes we’d go over to Grandma and Grampa’s house, and he would pick the dinner he wanted.”

Despite these special treats, Quentin’s behavioral problems grew worse. He started to kick, scream and cry hysterically. “He kicked a nurse once really hard, and I, as a mom, was incredibly ashamed,” says Matahna. “It’s hard to make him understand at 3 and 4 years old that he has to have [his treatments] and have him comprehend it. And, then tell him he needs not to throw fits.”

So, Matahna made the decision to move his treatments to the home, where she reasoned his behavior might improve. But, that turned out to be more difficult than she thought. At first, they found a home infusion nurse Quentin really liked, but she quit. After that, they went through several more nurses; Quentin didn’t like them, and they didn’t like him. "I think because he felt safe and secure at home, some of his behaviors even got worse, when I hoped they would get better,” says Matahna.

Good Nursing and Choices
Quentin transitioned back and forth twice between homecare and doctor’s office infusions. After experiencing these transitions, Matahna learned that the nurse made all the difference.

Immune globulin (IG) is a miraculous therapy, and no one knows this better than patients who receive it for an immune-mediated disease. Indeed, without IG, these individuals would fail to make the transition from living in a constant state of illness, struggling to survive, to a healthy, happy lifestyle. That is what this column is about — the unique and sometimes extraordinary stories of people of all ages whose lives have positively transitioned with the help of IG. We want to hear your stories! Email us at editor@igliving.com.
If Quentin had a nurse he could connect with, he still threw fits, but they weren’t nearly as bad. The first nurse he made this connection with made a deal with him that at every infusion, he would have a special treat for him that he could see only after he accessed the port and started the infusion. For instance, one time he brought a big projector and a white board with tracing paper and pencils. The nurse put pictures on the projector that he had taken, and Quentin would sit on the floor and draw.

According to Matahna, it makes a world of difference if the child trusts the nurse and they have a bond between them. “I didn’t have to get along with the nurse; my son had to have the connection with the nurse,” she explains. “I had to trust him coming into my home, but the connection and the bond had to be between the nurse and my son.”

Another thing that made a difference was giving Quentin a little more control over the infusions. “We asked him if he wanted us to tell him ahead of time when infusions were,” says Matahna. He did. And, now they put a big T to mark the dates on his calendar so he knows when treatment day is. “We talk about the infusion the day before, and he knows that [his infusion] is coming as soon as he gets home from school,” says Matahna. “If he knows exactly what’s coming, it’s better. Knowledge is power for him. Getting treatment — that’s not his choice. But if you can find things that they can have choices about, that works really well.” For instance, now he gets to have a snack right away while the nurse is getting ready, and when she’s ready, he likes to sit in Matahna’s lap and they’ll talk about a story or sing a song, and he just counts to three and then the nurse pokes him.

All in the Family
The Ericksons have three other children: 8-year-old Tevin, 6-year-old Makaylee and 2-year-old Karsten. Cooper, a son older than Quentin, died from heart complications when Matahna was pregnant with Quentin. According to Matahna, Karsten is quite compassionate and will pat him on the leg and say, “Shh, don’t cry Quentin,” or he’ll try to bring him a toy and console him.

Makaylee went through a phase of questioning why Quentin is sick and whether she was going to get it. Then, she thought it was a “boy” thing and wondered why Karsten didn’t get sick after he was born. Matahna bought Quentin a “feel-better” bear that has a patch on his heart and hospital clothes with various first-aid items, and they named him Cooper Bear. Before treatments start, Makaylee asks Quentin, “Do you want Cooper Bear?”

Treatment nights are on Fridays now, and they involve the whole family. Quentin gets to pick the dinner and the movie, and it’s his choice about what they eat during the movie. That, says Matahna, is popcorn 98 percent of the time.

A Better Tomorrow
Recently, Quentin outgrew his port, it became clogged and had to be removed. While at first he was glad to be rid of it because it made him feel different from the other kids at school, now that he has experienced IVs, he wants the port back. But that won’t happen soon because this summer his treatments will be temporarily discontinued so his immunoglobulin levels can be retested.

Quentin is hoping that he’ll grow out of his CVID, something doctors think is possible. If not, Matahna says Quentin will likely choose to have another port, and she’s looking on the bright side. With maturation, more control and a good nurse on Quentin’s side, she no longer has to leave the room and cry each time he is treated. She knows things are a lot better now.

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

Patient-to-Patient: Connecting to a Larger Community

The Internet, retreats and conferences offer great opportunities for PIDD patients and their families to interact with others affected by the same disease.

By Mark T. Haggard

Life can be lonely for those who live with primary immune deficiency (PIDD). We often find that we are the only person in a city or a town with this disease. There is no one else in our school, our circle of friends, our church or our place of worship who knows what we go through. That is why it is so important to make connections to a broader community of others carrying the same burden. Just knowing there are others out there can relieve the overwhelming isolation we feel. Thankfully, with a little knowledge and a few clicks of a mouse, we can connect with hundreds, and even thousands, of others in similar circumstances.

The Disease Shield
While HIPPA laws are designed to keep medical practitioners from inappropriately sharing patient information, they make it difficult for a person living with PIDD, or any other disease, to meet others with the same condition. What’s more, many PIDD patients are treated through home healthcare rather than in a hospital or clinic setting, and the number of people we see living with the same condition shrinks to zero. Help is out there, though. We are not alone.

Connecting Through the Internet
During this “information age,” the Internet has become a great tool for many with PIDD. Historically, patients were leery of going out in public because of the prospect of getting sick, but the Internet now allows scores of people — doctors, nurses, patients, advocates, writers — to connect with each other in a community that stretches far beyond city limits, state lines and national borders.

Renee Tuckwiller uses the Internet to help her keep connected. Renee is from Auburndale, Fla., and was diagnosed with IgG deficiency in 1997. Not knowing anyone else with IgG deficiency and in need of a community, she did a Google search and found the Immune Deficiency Foundation (IDF) website through which she made a connection with a volunteer and attended a retreat. She is now an IDF volunteer herself, helping at family retreats and a number of conferences.

Earlier this year, IG Living magazine added a Facebook page (www.facebook.com/IGLivingMagazine) where readers can connect with one another and see what other immune patients and caretakers are doing. Each weekday, the magazine poses a question to which readers can respond. Questions range from the thoughtful, “What advice do you have for those on Medicare/Disability?” to the sublime, “When do you tell someone whom you are dating that you have a chronic illness?” As of this writing, there are nearly 500 people connected to this community.

These are but a handful of hundreds of resources available online. And, Renee cautions that,
although the availability of information on the web has increased precipitously, a patient or caregiver needs to be careful to figure out what is truth and what is not. Anyone can create a website and put whatever they want on it. The key is to find trustworthy websites that speak fact rather than fiction.

Connecting Through Retreats and Conferences

Retreats and conferences are the perfect opportunities for making the connections that we need. For instance, we need to be made aware of the latest changes in treatments, and be given hope that research is moving forward. More than that, we need to know that someone else is living with the same condition. Connecting with others helps us to understand that we are not alone in dealing with our disease.

Branson Worthen of Syracuse, Utah, was diagnosed with CVID when he was 6 years old (see the story about Branson in IG Living’s April-May 2010 issue). His mother, Connie, was told by one nurse that he would have serious health issues when he reached puberty. But, when the family traveled to an IG Living-sponsored retreat, they talked with a number of adults living good lives with CVID who, apparently, did not have serious health issues during puberty. “Meeting adults and other children changed our family forever!” Connie exclaimed. “We redirected our focus in a more positive way, and for the first time, we weren’t afraid of the diagnosis.” Retreats give Connie and her family the satisfaction of knowing that her children will be able to live a long and full life.

Sometimes medical “stuff” can be intimidating. We might not be sure how to talk to a doctor who has multiple diplomas hanging on the wall. The matter-of-fact approach of some doctors can be one of the main obstacles to our well-being. Connie said that the “doomsday” feeling that they got from a doctor caused them great grief. Conferences can help mediate those feelings by providing valuable information in a comfortable setting. Every other year, IDF sponsors a three-day national conference that connects hundreds of attendees to each other and scores of experts — doctors, advocates, lobbyists — from across the country. The next IDF National Conference will be held in Phoenix, Ariz., in the summer of 2011.

Retreats and conferences are the perfect opportunities for making the connections that we need.

Connie notes, though, that large “national” conferences can have a couple of drawbacks: The first is that they are not very personable; the second is that, even though they are medical conferences, they can still be germ factories. “We prefer the local miniconferences,” she says. Because it is a smaller group, there is more opportunity for personal contact, including the much-sought-after one-on-one time with doctors.

Smaller conferences and events are held throughout the year. The Jeffrey Modell Foundation has sponsored KIDS Day events in various cities around the country. As the name implies, these events are directed at children with immune deficiencies, allowing them to spend time with others facing the same issues. Likewise, this also is another opportunity for parents of PIDD kids to connect. The IDF also sponsors conferences targeting specific types of immune deficiencies, as well as an annual teen conference.

Once a month, IG Living magazine offers a toll-free teleconference featuring guest speakers who are some of the most knowledgeable people in the immune-compromised community, including doctors, nurses and advocates. One can sign up to receive an invitation to a readers’ group teleconference at www.IGLiving.com.

The Need for Connection

Human beings are not designed to carry their burdens alone. As social beings, we need to help carry the burdens of others, as well as have others help us carry ours. We have to connect with others or else our disease can crush us — in both health and spirit. There may be a few roadblocks along the way, but the tools are out there to make connections to a broader community that will surely give us a lift.  

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.
**GASTROINTESTINAL (GI) SYMPTOMS** are a common problem among patients with an immune disease. As a result, many patients are diagnosed with either irritable bowel syndrome (IBS) or inflammatory bowel disease (IBD). Both IBS and IBD present with symptoms such as abdominal pain, cramping, bloating, diarrhea or constipation. These symptoms are the gut’s way of screaming for help. Yet, despite what their gut is trying to tell them, many patients are embarrassed to talk about IBS and IBD symptoms, no matter how debilitating they might get. But, not talking about symptoms can raise patients’ stress levels, which in turn, will make symptoms worse. So, it is a conversation patients should have with their doctor.

**Effective Treatments**

Although currently there is no cure for either IBS or IBD, there are effective treatments. Tracking symptoms and responses to treatments is one of the first things patients’ doctors may ask them to do. Keeping a simple log of all symptoms, along with infusions, allows a doctor to spot any patterns associated with symptoms. NuFACTOR specialty pharmacy has a very simple, complimentary health diary developed especially for immune globulin patients, which can be requested at https://secure.nufactor.com/pages/ig_infusion_log_form.html.

Tracking symptoms also may help to reveal certain patterns associated with infections. Because the intestinal system is full of billions of bacteria, known as “gut flora,” it can easily be upset by chronic use of antibiotics needed to fight infections. Out-of-balance flora can lead to overgrowth of some bacteria. Recent research suggests small intestinal bacterial overgrowth may contribute to IBS. If patients have shown a pattern of increased symptoms after antibiotics, they may be asked to take another antibiotic such as rifaximin or neomycin to help get the gut flora back in balance. These particular antibiotics are used because they are not absorbed in the stomach and, therefore, are thought to work better on the bacteria in the small intestine.

Keeping gut flora in balance is thought to be a key strategy for better overall health. Some doctors recommend their patients maintain good balance through diet and supplements that include probiotics. Many food products, such as yogurt and cheese, already carry natural-forming probiotics.

Despite their best efforts to eat a well-balanced diet and to exercise, patients still can have severe GI symptoms requiring more involved treatments. IBD, such as Crohn’s or ulcerative colitis, are chronic autoimmune diseases that often require the use of prescription medications to achieve and maintain disease remission. Aminosalicylates, such as sulfasalazine (Azulfidine), mesalamine (Asacol, Pentasa), olsalazine (Dipentum) and balsalazide (Colazal), are drugs that interfere with the body’s ability to control inflammation. Corticosteroids can help with flare-ups, providing rapid relief and a decrease of symptoms on a short-term basis. Patients who do not respond to traditional therapies or whose disease is more progressive may require the use of immune modulation drugs, such as methotrexate, Imuran, cyclosporine, Humira and Cimzia.

Whether a patient’s disease is well-controlled or they have occasional flare-ups, acute awareness to nutritional needs is the key to a healthier life. Some patients may require nutritional support via high-calorie, high-nutrition supplement drinks, such as Boost or Ensure. But, these drinks are not meant to displace a well-balanced meal, and a doctor should be consulted before using any supplements.

**A Serious Issue**

Symptoms of IBS and IBD are not to be taken lightly. Gone unchecked, symptoms can lead to malnutrition and a decreased quality of life. Openly discussing symptoms and treatments with a doctor is the best way to get relief and support.

**Sources**


**KRIS MCFALLS** is the full-time patient advocate for IG Living magazine.
Nutrition Supplements

**Align**
Align is a digestive care probiotic supplement made with Bifantis. Bifantis replenishes the digestive tract with beneficial bacteria, helping to build and maintain a healthy digestive system — providing ongoing protection against occasional digestive upsets. Align is intended to be taken once a day and comes in capsule form. It is available in local pharmacies.

[www.aligngi.com](http://www.aligngi.com)

**Boost**
Boost nutritional energy drinks contain 26 vitamins and minerals, antioxidants and protein. It comes in six forms: Boost Drink for extra energy, Kid Essentials, Boost Plus Drink containing supplemental nutrients for weight gain, Boost High Protein Drink, Boost High Protein Powder and Boost Glucosamine Control (a snack for people with diabetes). It is available at most drugstores.

[www.boost.com](http://www.boost.com)

**Culturelle**
Culturelle comes in five different products. Natural Health and Wellness, Culturelle for Kids, Digestive Health (dairy- and gluten-free), Dairy Free Natural Health and Wellness (which contains inulin, is encapsulated in a vegetable capsule and is dairy- and gluten-free) and Digestive Relief (for occasional bouts of diarrhea and upset stomach). All are available in most major health food stores.

[www.culturelle.com](http://www.culturelle.com)

**Ensure**
Ensure shakes contain fiber, omega-3s and 24 essential vitamins and minerals, and come in four different forms. Ensure Plus is intended to help individuals gain or maintain a healthy weight and contains 350 calories. Ensure High Protein contains 12 grams of high-quality protein to help build muscle. And, Ensure High Calcium has added calcium. All products are available in drugstores.

[www.ensure.com](http://www.ensure.com)

Support Groups

**Crohn’s and Colitis Foundation of America**
This nonprofit, volunteer-driven organization is dedicated to finding the cure for Crohn’s disease and ulcerative colitis. The foundation funds cutting-edge studies at major medical institutions, nurtures investigators at the early stages of their careers, and finances underdeveloped areas of research. It hosts educational workshops and symposiums, and publishes a scientific journal, *Inflammatory Bowel Diseases*, created to enable medical professionals to keep pace with this rapidly growing field.

[www.ccfa.org](http://www.ccfa.org)

**Crohn’s & Me**
Crohn’s & Me is a website created to be a community for people affected by the disease. The site explains the disease, provides online tracking tools, allows individuals to share their stories, provides lifestyle tips and lists treatment options. Individuals can become a member of the site free of charge.

[www.crohnsandme.com](http://www.crohnsandme.com)

**IBS Self Help and Support Group**
The Irritable Bowel Syndrome (IBS) Self Help and Support Group, established in 1987, is an award-winning patient advocate group in support of self-management for those who suffer from IBS, those who are looking for support for someone who has IBS, and medical professionals who want to learn more about IBS. In addition to forums, the website provides a list of helpful links, a very comprehensive booklist, research studies, brochures, medical tests, diagnostic criteria and diet, treatment and medications about, and for, the disease. The community has more than 37,000 active members.

[www.ibsgroup.org](http://www.ibsgroup.org)
Sources

Celiac Disease: A Hidden Epidemic
Authors: Peter Green, MD, and Rory Jones
Publisher: Collins Living, www.harpercollins.com

Celiac Disease: A Hidden Epidemic is an authoritative guide to this serious autoimmune disease. As co-author Dr. Peter Green explains, “Celiac disease has been dubbed the ‘great pretender.’ Its symptoms can easily masquerade as a number of other illnesses … and it is those conditions that are often diagnosed instead of celiac disease. The one thing most patients have in common is a long road to diagnosis.”

The book is intended to help patients “know what to … ask their physicians and how to understand the answers.” It covers diagnosis, management, the latest research, complications, related disorders, coping with psychological aspects and adjusting to the gluten-free diet. It is available in hardcover or as an e-book.

Th17 Cells: Role in Inflammation and Autoimmune Disease
A Patient and Her Doctor Negotiate a Life with Chronic Illness
Author: Alida Brill
Publisher: Schaffner Press, www.schaffnerpress.com

This book is intended for scientific libraries; researchers and clinicians from immunology, inflammation research, rheumatology, dermatology; and the pharmaceutical and biotech industry. It describes the function of the Th17 cell subset and its prevalent role in many inflammatory and autoimmune diseases. The mechanism by which this T cell subset is generated and maintained shows specific differences between mice and man, which are reviewed. One characteristic of Th17 cells is to secrete specific and unique cytokines such as IL-17A, IL-17F and IL-22, and their functions are described in detail. These cytokines have acquired a prevalent role in many pathological conditions in animal models and in humans. Targeting these cytokines, as well as the generation of Th17 cells, may open new ways to treat many autoimmune diseases. The role of Th17 in infectious conditions is also addressed.

Overcoming Diabetes, Lupus, Arthritis, Sarcoidosis, Prednisone, Obesity
Author: David Dobson
Publisher: Xlibris, www.Xlibris.com

Author David Dobson isn’t a medical doctor, but he has gone through enough illnesses in his lifetime to qualify him as a quasi-medical expert on the subject. A former Army officer, Dobson’s worst battle has been the one for his health. Besieged by a multitude of autoimmune diseases, Dobson’s strict discipline and positive outlook helped him come out a victor. He shares the strategies that helped him keep his health in this well-researched part-autobiographical, part-medical resource book.

For most of his young life, Dobson had the flu, chronic cough and a sinus infection before being diagnosed with sarcoidosis, an inflammatory disease caused by an overacting immune system, resulting in damage to the body’s own tissues. The treatment involved taking prednisone, a powerful anti-inflammatory drug that caused weight gain, peptic ulcer, facial swelling and high blood pressure. Before long, Dobson found out that he was also suffering from diabetes. After attending a seminar on how to reverse diabetes, he was soon on his way to a healthier and more enjoyable life.

The journey was not without its bumps, however. Stress and anxiety soon found him back in worse conditions and with more illnesses. But, by adhering to a strict diet and exercise program, Dobson conquered some of the most challenging ailments. His account, discoveries, treatments, regimens and recipes are summed up in Overcoming Diabetes, Lupus, Arthritis, Sarcoidosis, Prednisone, Obesity.
General Resources

Other Organization Websites
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
- Alliance for Plasma Therapies (fair access to plasma therapies): www.plasmaalliance.org
- American Autoimmune Related Diseases Association (AARDA): www.aarda.org
- American Chronic Pain Association (ACPA): www.theacpa.org
- Band-Aides and Blackboards: www.lehman.cuny.edu/faculty/jfleitas/bandaides
- Cleveland Clinic: www.clevelandclinic.org/health
- 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
- Platelet Disorder Support Association: www.pdsa.org
- The nonprofit Patient Services Incorporated, www.uneedpsi.org, specializes in health insurance premium, pharmacy co-payment and co-payment waiver assistance for people with chronic illnesses. (800) 366-7741
- WebMD (medical reference): www.webmd.com

IG Manufacturer Websites
- Baxter: www.baxter.com
- CSL Behring: www.cslbehring.com
- Grifols: www.grifolsusa.com
- Octapharma: www.octapharma.com
- Talecris: www.talecris.com

IG Manufacturer Websites
- Baxter: www.baxter.com
- CSL Behring: www.cslbehring.com
- Grifols: www.grifolsusa.com
- Octapharma: www.octapharma.com
- Talecris: www.talecris.com

Disease-State Resources

Ataxia Telangiectasia (A-T)

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

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

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

Evans Syndrome

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

Guillain-Barré Syndrome (GBS)

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

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

Idiopathic Thrombocytopenic Purpura (ITP)

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

Kawasaki Disease

Websites
- American Heart Association (how the disease affects the heart): www.americanheart.org/presenter.jhtml?identifier=4634
- Kawasaki Disease Foundation: www.kdfoundation.org

For a more comprehensive list of resources, visit the Resources page at www.IGLiving.com.
**Sources**

- **KidsHealth**: http://kidshealth.org/parent/medical/heart/kawasaki.html

**Mitochondrial Disease**

**Websites**
- United Mitochondrial Disease Foundation:  www.umdf.org

**Multifocal Motor Neuropathy (MMN)**

**Websites**
- The Neuromuscular Center at Washington University:  www.neuro.wustl.edu/neuromuscular
- The Neuropathy Association:  www.neuropathy.org

**Multiple Sclerosis (MS)**

**Websites**
- All About Multiple Sclerosis:  www.mult-sclerosis.org/index.html
- Multiple Sclerosis Association of America:  www.msaa.com
- Multiple Sclerosis Foundation:  www.msfacts.org
- 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**
- Autoimmune Information Network Inc.:  www.aininc.org

**Myositis**

**Websites**
- International Myositis Assessment and Clinical Studies Group:  https://dir-apps.niehs.nih.gov/imacs/index.cfm?action=home.main

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

**Online Peer Support**
- Myositis Association Community Forum:  www.myositis.org
- Myositis Support Group:  www.myositisupportgroup.org
- Myositis Support Group – UK:  www.myositis.org.uk

**Pemphigus and Pemphigoid**

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

**Peripheral Neuropathy (PN)**

**Websites**
- 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, advocate the need for early intervention and support research into the causes and treatment of neuropathies. (212) 692-0662

- Neuphathy Action Foundation:  www.neuropathyaction.org

**Online Peer Support**
- Calgary Neuropathy Support Group:  www.calgarypners.org

**Primary Immune Deficiency Disease (PIDD)**

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

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

- The National Institute of Child Health and Human Development (NICHD), www.nichd.nih.gov, is part of the National Institutes of Health. Go to the “Health Information and Media” tab on the website and do a search under

- American Academy of Allergy, Asthma & Immunology:  www.aaaai.org
- International Patient Organization for Primary Immunodeficiencies (IPOPI):  www.ipopi.org
- Michigan Immunodeficiency Foundation:  www.midf.org
• National Institute of Child Health and Human Development (NICHD) (Click on “Health Information and Media” tab and search for “primary immunodeficiency”: www.nichd.nih.gov
• New England Primary Immunodeficiency Network: www.nepin.org
• Rainbow Allergy-Immunology: www.uhhospitals.org/tabid/132/Default.aspx
• Team Hope (for families and patients in New England): www.teamhope.info

Online Peer Support
• IDF Common Ground: www.idfcommonground.org
• IDF Discussion Forum: my.primaryimmune.org/forum
• IDF Friends: my.primaryimmune.org
• Jeffrey Modell Foundation Message Board: www.info4pi.org
• Rhode Island peer group: http://health.groups.yahoo.com/group/RhodeIslandPIDD

Scleroderma
Websites
Scleroderma Center: http://scleroderma.jhmi.edu
• Scleroderma Foundation: www.scleroderma.org
• Scleroderma Research Foundation: www.srfcure.org

Online Peer Support
• Cure2Zone.com: curezone.com/forums/f.asp?f=404
• International Scleroderma Network: www.sclero.org/support/targets/a-to-z.html

Stiff-Person Syndrome (SPS)
Websites
• American Autoimmune Related Diseases Association Inc.: www.aarda.org
• Autoimmune Information Network Inc.: www.aininc.org
• Living with Stiff Person Syndrome (personal account): www.livingwithsp.com

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.
• DisabilityInfo.gov: www.disabilityinfo.gov
  U.S. Federal government’s disability-related information and resources.
• Individuals with Disabilities Education Improvement Act of 2004:  http://idea.ed.gov/explore/home
• National Disabilities Rights Network: www.ndr.org
  This website offers a search tool to find resources in your state to assist with school rights and advocacy.
• Social Security: www.ssa.gov/disability

Product Information
• Influenza and the influenza vaccine: www.cdc.gov/flu or call (800) CDC-INFO: (800) 232-4636
• IVIG Carimune NF: www.carimune.com
• IVIG Flebobagam: www.grifolsusa.com/pdfs/flebo_14Jun05.pdf
• IVIG Gammagard Liquid: www.gammagardliquid.com
• IVIG Gammagard S/D: www.immunedisease.com
• IVIG Gamunex: www.gamunex.com
• IVIG Octagam: www.octapharma.com
• IVIG Privigen: www.privigen.com
• SCIG (subcutaneous immune globulin) Vivaglobin: www.vivaglobin.com

Pump and Infusion Sets Websites
• EMED Corporation: www.safetymedicalproducts.com
• Graseby Marcal Medical: www.marcalmedical.com
• Intra Pump Infusion Systems: www.intrapump.com
• Micrel Medical Devices: www.micrelmed.com
• Norfolk Medical: www.norfolkmedical.com
• Repro Med Systems, Inc: 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.
Hizentra is contraindicated in patients with hyperprolinemia because it contains the stabilizer such as polysorbate 80.

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

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

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

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

5 Warnings and Precautions

5.1 Hypersensitivity

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

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

5.2 Reactions Reported to Occur With IGIV Treatment

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

Renal Dysfunction/Failure

Renal dysfunction/failure, osmotic nephropathy, and death may occur with use of human immune globulin products. Ensure that patients are not volume depleted and assess renal function, including measurement of blood urea nitrogen (BUN) and serum creatinine, before the initial infusion of Hizentra and at appropriate intervals thereafter.

Periodic monitoring of renal function and urine output is particularly important in patients judged to have a potential increased risk of developing acute renal failure. If renal function deteriorates, consider discontinuing Hizentra. For patients judged to be at risk of developing renal dysfunction/failure because of pre-existing renal insufficiency or predisposition to acute renal failure (such as those with diabetes mellitus or hypovolemia, those who are overweight or use concomitant nephrotoxic medicinal products, or those who are over 65 years of age), administer Hizentra at the minimum rate practicable.

Thrombotic Events

Thrombotic events may occur with use of human immune globulin products. Patients at increased risk may include those with a history of atherosclerosis, multiple cardiovascular risk factors, advanced age, impaired cardiac output, hypercoagulable disorders, prolonged periods of immobilization, and/or known or suspected hyperviscosity. Because of the potentially increased risk of thrombosis, consider baseline assessment of blood viscosity in patients at risk for hyperviscosity, including those with cryoglobulins, fasting chylomicronemia/markedly high triglycerides (triglycerides), or monoclonal gammapathies. For patients judged to be at risk of developing thrombotic events, administer Hizentra at the minimum rate practicable.

Aseptic Meningitis Syndrome (AMS)

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

Conduct a thorough neurological examination, including CSF studies, to rule out other causes of meningitis in patients exhibiting signs and symptoms of AMS. Discontinuation of IGIV treatment has resulted in remission of AMS within several days without sequelae.

Hemolysis

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

Transfusion-Related Acute Lung Injury (TRALI)

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

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

5.3 Transmissible Infectious Agents

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

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

5.4 Laboratory Tests

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

6 Adverse Reactions

The most common adverse reactions (ARs), observed in ≥25% of study subjects receiving Hizentra, were local reactions (i.e., swelling, redness, heat, pain, and itching at the injection site), headache, vomiting, pain, and fatigue.

6.1 Clinical Trials Experience

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

The safety of Hizentra was evaluated in a clinical study for 15 months in subjects with PI who had been treated previously with IGIV every 3 or 4 weeks. The safety analyses included 49 subjects in the intention-to-treat (ITT) population. The ITT population consisted of all subjects who received at least one dose of Hizentra (see Clinical Studies [14]).

Subjects were treated with Hizentra at weekly doses ranging from 66 to 331 mg/kg body weight during the wash-in/wash-out period and from 72 to 379 mg/kg during the efficacy period. The 49 subjects received a total of 2264 weekly infusions of Hizentra.

No deaths or serious ARs occurred during the study. Two subjects withdrew from the study due to ARs. One subject experienced a severe injection-site reaction one day after the third weekly infusion, and the other subject experienced moderate myositis. Both reactions were judged to be “at least possibly related” to the administration of Hizentra.

Table 2 summarizes the most frequent adverse events (AEs) (experienced by at least 4 subjects), irrespective of causality. Included are all AEs and those considered temporally associated with the Hizentra infusion, i.e., occurring during or within 72 hours after the end of an infusion. Local reactions were the most frequent AEs observed, with injection-site reactions (i.e., swelling, redness, heat, pain, and itching at the site of injection) comprising 98% of local reactions.

Table 2: Incidence of Subjects With Adverse Events (AEs)* (Experienced by 4 or More Subjects) and Rate per Infusion, Irrespective of Causality (ITT Population)

<table>
<thead>
<tr>
<th>AE (≥4 Subjects)</th>
<th>All AEs*</th>
<th>AEs* Occurring During or Within 72 Hours of Infusion</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Number (%) of Subjects (n=49)</td>
<td>Number (Rate) of AEs (n=2264 Infusions)</td>
</tr>
<tr>
<td>Local reactions†</td>
<td>9 (100)</td>
<td>1340 (0.592)</td>
</tr>
</tbody>
</table>
Table 2: Incidence of Subjects With Adverse Reactions (Experienced by 2 or More Subjects) to Hizentra and Rate per Infusion (ITT Population)

<table>
<thead>
<tr>
<th>Adverse Reaction (≥2 Subjects)</th>
<th>Number (% of Subjects (n=49)</th>
<th>Number (Rate) of Adverse Reactions (n=2264 Infusions)</th>
<th>Number (% of Subjects (n=49)</th>
<th>Number (Rate) of Adverse Reactions (n=2264 Infusions)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Other AEs:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>13 (26.5)</td>
<td>40 (0.018)</td>
<td>12 (24.5)</td>
<td>32 (0.014)</td>
</tr>
<tr>
<td>Cough</td>
<td>8 (16.3)</td>
<td>9 (0.004)</td>
<td>5 (10.2)</td>
<td>6 (0.003)</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>7 (14.3)</td>
<td>8 (0.004)</td>
<td>5 (10.2)</td>
<td>6 (0.003)</td>
</tr>
<tr>
<td>Fatigue</td>
<td>6 (12.2)</td>
<td>6 (0.003)</td>
<td>4 (8.2)</td>
<td>5 (0.002)</td>
</tr>
<tr>
<td>Back pain</td>
<td>5 (10.2)</td>
<td>11 (0.005)</td>
<td>4 (8.2)</td>
<td>5 (0.002)</td>
</tr>
<tr>
<td>Nausea</td>
<td>5 (10.2)</td>
<td>5 (0.002)</td>
<td>4 (8.2)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Abdominal pain, upper</td>
<td>5 (10.2)</td>
<td>5 (0.002)</td>
<td>3 (6.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Rash</td>
<td>5 (10.2)</td>
<td>7 (0.003)</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Pain in extremity</td>
<td>4 (8.2)</td>
<td>7 (0.003)</td>
<td>4 (8.2)</td>
<td>6 (0.003)</td>
</tr>
<tr>
<td>Migraine</td>
<td>4 (8.2)</td>
<td>5 (0.002)</td>
<td>3 (6.1)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Pain</td>
<td>4 (8.2)</td>
<td>5 (0.002)</td>
<td>3 (6.1)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Epistaxis</td>
<td>4 (8.2)</td>
<td>6 (0.003)</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Pharyngolaryngeal pain</td>
<td>4 (8.2)</td>
<td>6 (0.003)</td>
<td>2 (4.1)</td>
<td>1 (&lt;0.001)</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>4 (8.2)</td>
<td>5 (0.002)</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
</tr>
</tbody>
</table>

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

The ratio of infusions with temporally associated AEs, including local reactions, to all infusions was 1338 to 2264 (59.1%; upper 95% confidence limit of 62.4%). Excluding local reactions, the corresponding ratio was 173 to 2264 (7.6%; upper 95% confidence limit of 8.9%).

Table 3 summarizes the most frequent AEs (i.e., those AEs considered by the investigators to be “at least possibly related” to Hizentra administration) experienced by at least 2 subjects.

Table 3: Incidence of Subjects With Adverse Reactions (Experienced by 2 or More Subjects) to Hizentra and Rate per Infusion (ITT Population)

<table>
<thead>
<tr>
<th>Adverse Reaction (≥2 Subjects)</th>
<th>Number (% of Subjects (n=49)</th>
<th>Number (Rate) of Adverse Reactions (n=2264 Infusions)</th>
<th>Number (% of Subjects (n=49)</th>
<th>Number (Rate) of Adverse Reactions (n=2264 Infusions)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Local reactions</td>
<td>49 (100)</td>
<td>1338 (0.591)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other AEs:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>12 (24.5)</td>
<td>36 (0.016)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vomiting</td>
<td>3 (6.1)</td>
<td>3 (0.001)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pain</td>
<td>3 (6.1)</td>
<td>4 (0.002)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fatigue</td>
<td>3 (6.1)</td>
<td>3 (0.001)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Contusion</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Back pain</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Migraine</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Diarrhea</td>
<td>2 (4.1)</td>
<td>2 (&lt;0.001)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Abdominal pain, upper</td>
<td>2 (4.1)</td>
<td>2 (&lt;0.001)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nausea</td>
<td>2 (4.1)</td>
<td>2 (&lt;0.001)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rash</td>
<td>2 (4.1)</td>
<td>2 (&lt;0.001)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Arthralgia</td>
<td>2 (4.1)</td>
<td>2 (&lt;0.001)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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

8.4 Pediatric Use
Hizentra was evaluated in 10 pediatric subjects (3 children and 7 adolescents) with PI. No pediatric-specific dose requirements were necessary to achieve the desired serum IgG levels. Hizentra was not evaluated in neonates or infants.

8.5 Geriatric Use
Of the 49 subjects evaluated in the clinical study of Hizentra, 6 subjects were 65 years of age or older. No overall differences in safety or efficacy were observed between these subjects and younger subjects.

15 REFERENCES
**Vivaglobin®**

**Immune Globulin Subcutaneous (Human)**

**BRIEF SUMMARY OF PRESCRIBING INFORMATION**

**Vivaglobin®** Immune Globulin Subcutaneous (Human), is indicated for the treatment of patients with primary immune deficiency (PID).

**CONTRAINDICATIONS**

- Animal reproduction studies have not been conducted with Vivaglobin® Immune Globulin Subcutaneous (Human). It is also not known whether Vivaglobin® can cause fetal harm when administered to a pregnant woman, or can affect labor, delivery, or breastfeeding.

**WARNINGS**

- Administration of immune globulin preparations can cause disease. Because Vivaglobin® is made from human blood, it may carry a risk of transmitting infectious agents, e.g., viruses, and theoretically, the CJD agent. The risk that such plasma-derived products will transmit an infectious agent has been reduced by screening plasma donors for prior exposure to certain viruses, by testing for the presence of certain current viruses in the blood, and by inactivating and/or removing certain viruses during manufacture (see DESCRIPTION section for virus reduction measures).

**DESCRIPTION**

Vivaglobin® is made from human plasma. Products made from human plasma may contain infectious agents, such as viruses, that can cause disease. Because Vivaglobin® is made from human blood, it may carry a risk of transmitting infectious agents, e.g., viruses, and theoretically, the CJD agent. The risk that such plasma-derived products will transmit an infectious agent has been reduced by screening plasma donors for prior exposure to certain viruses, by testing for the presence of certain current viruses in the blood, and by inactivating and/or removing certain viruses during manufacture (see DESCRIPTION section for virus reduction measures).

**DIAGNOSTIC AND USAGE**

Vivaglobin® Immune Globulin Subcutaneous (Human) is contraindicated in individuals with a history of anaphylactic or severe systemic response to immune globulin preparations and in persons with selective immunoglobulin (IgA) deficiency (serum IgA < 0.05 g/L) who have known antibody against IgA.

**PRECAUTIONS**

- Administration of Vivaglobin® Immune Globulin Subcutaneous (Human), subcutaneously. Do not administer this product intravenously.

**STORAGE**

Vivaglobin® should not be mixed with other medicinal products.

**PREGNANCY CATEGORY C**

**ADVERSE REACTIONS**

- The most frequent adverse reaction was local reaction at the injection site. Table 7 summarizes the most frequent related adverse events by subject reported in the clinical study, and Table 8 summarizes the most frequent related adverse events by infusion.

- In the non-IND Europe and Brazil clinical study, the safety of Immune Globulin Subcutaneous (Human), Vivaglobin® was evaluated for 10 months in 65 subjects with PID. The adverse events and their rates reported in this study were similar to those reported in the US and Canada study, with two notable exceptions for the related adverse events. These events were 59 episodes of headache (1.6%) and 2 episodes of fever (0.1%) in the US and Canada study and no episodes of headache and 18 episodes of fever (0.8%) in the Europe and Brazil study.

- Local injection site reactions - Local injection site reactions consisting of mostly mild or moderate swelling, redness and itching, have been observed with the use of Vivaglobin®. No serious local site reactions were observed. The majority of injection site reactions resolved within four days. Additionally, the number of subjects reporting local injection site reactions decreased substantially after repeated use (see Figure 1). Only three subjects in the US and Canada study and one subject in the Europe and Brazil study discontinued due to local site reactions.

**Table 7: Most Frequent Related Adverse Events by Subject**

<table>
<thead>
<tr>
<th>Related Adverse Event</th>
<th>No. of Subjects (Number of Infusions: 3656)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-Injection Site Reactions</td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>21 (32%)</td>
</tr>
<tr>
<td>Nausea</td>
<td>7 (11%)</td>
</tr>
<tr>
<td>Rash</td>
<td>4 (6%)</td>
</tr>
<tr>
<td>Asthenia</td>
<td>3 (5%)</td>
</tr>
<tr>
<td>Gastrointestinal disorder</td>
<td>3 (5%)</td>
</tr>
<tr>
<td>Fever</td>
<td>2 (3%)</td>
</tr>
<tr>
<td>Skin disorder</td>
<td>2 (3%)</td>
</tr>
<tr>
<td>Tachycardia</td>
<td>1 (2%)</td>
</tr>
<tr>
<td>Urine abnormality</td>
<td>2 (3%)</td>
</tr>
</tbody>
</table>

**Table 8: Most Frequent Related Adverse Events by Infusion**

<table>
<thead>
<tr>
<th>Related Adverse Event</th>
<th>No. of AEs (Number of Infusions: 3656)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-Injection Site Reactions</td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>59 (1.6%)</td>
</tr>
<tr>
<td>Rash</td>
<td>9 (0.2%)</td>
</tr>
<tr>
<td>Nausea</td>
<td>9 (0.2%)</td>
</tr>
<tr>
<td>Nervousness</td>
<td>4 (0.1%)</td>
</tr>
<tr>
<td>Asthenia</td>
<td>3 (0.1%)</td>
</tr>
<tr>
<td>Gastrointestinal disorder</td>
<td>3 (0.1%)</td>
</tr>
<tr>
<td>Skin disorder</td>
<td>3 (0.1%)</td>
</tr>
<tr>
<td>Urine abnormality</td>
<td>3 (0.1%)</td>
</tr>
<tr>
<td>Fever</td>
<td>2 (0.1%)</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>2 (0.1%)</td>
</tr>
<tr>
<td>Gastrointestinal pain</td>
<td>2 (0.1%)</td>
</tr>
<tr>
<td>Tachycardia</td>
<td>2 (0.1%)</td>
</tr>
</tbody>
</table>

**Table 6: Most Frequent Adverse Events by Infusion**

<table>
<thead>
<tr>
<th>Adverse Events (Number of Infusions: 3656)</th>
<th>No. of Adverse Events (Rate)**</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-Injection Site Reactions</td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>1789 (49%)</td>
</tr>
<tr>
<td>Nausea</td>
<td>1112 (30%)</td>
</tr>
<tr>
<td>Rash</td>
<td>601 (16%)</td>
</tr>
<tr>
<td>Severe</td>
<td>65 (2%)</td>
</tr>
<tr>
<td>Unknown Severity</td>
<td>11 (3%)</td>
</tr>
<tr>
<td>Gastrointestinal disorder</td>
<td>40 (11%)</td>
</tr>
</tbody>
</table>

**Table 5: Most Frequent Adverse Events by Subject**

<table>
<thead>
<tr>
<th>Adverse Events (10% of subjects)</th>
<th>No. of Subjects (10% of subjects)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-Injection Site Reactions</td>
<td>60 (18%)</td>
</tr>
</tbody>
</table>

*Including infections*

*Rate = number of reactions/infusion*
Because Vivaglobin®
The risk that such plasma-derived products will transmit an infectious agent has been reduced by screening
Thus, the risk of transmission of infectious agents cannot be
Any infections thought by a physician to have been possibly transmitted by this product should be reported by the
When initiating therapy with Vivaglobin®, the immunizing physician should be informed of recent therapy with Vivaglobin®
There were no apparent differences in the safety and efficacy profiles as compared to adult subjects.
Reactions similar to those reported with administration of other immune globulin products may also
Rarely, immediate anaphylactoid and hypersensitivity reactions may occur. In exceptional cases, sensitization to
Table 5 summarizes the most frequent adverse events by subject reported in the clinical study, and Table 6 summarizes the most frequent

*Only three subjects in the US and Canada study and one subject in the Europe and Brazil study discontinued
Note: Analysis is confined to 70 infusions.

Important Safety Information
Hizentra and Vivaglobin are indicated for the treatment of patients with primary immunodeficiency (PI).
If you have a history of anaphylactic or severe systemic response to immune globulin preparations or selective immunoglobulin A deficiency, check with your physician as neither Vivaglobin nor Hizentra should be used. If your physician suspects you are having anaphylactic or anaphylactoid reactions, treatment will be discontinued. Because Hizentra contains the stabilizer L-proline, you cannot be treated with Hizentra if you have hyperprolinemia.
Hizentra and Vivaglobin are derived from human plasma. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent, cannot be completely eliminated.
In separate clinical trials for Hizentra and Vivaglobin, the most frequent adverse event was injection-site reaction, consisting of mild or moderate swelling, redness, and itching. With Vivaglobin, no serious local site reactions were observed, and reactions tended to decrease substantially after repeated use. Other adverse events with Vivaglobin, irrespective of causality, included headache, gastrointestinal disorder, fever, nausea, sore throat, and rash.
Adverse reactions to Hizentra, observed in 5% or more of clinical trial subjects, were local injection-site reactions, headache, vomiting, pain, and fatigue.

Hizentra is manufactured by CSL Behring AG and distributed by CSL Behring LLC.
Hizentra is a trademark of CSL Behring AG.
Hizentra is a trademark of CSL Behring AG.

Your physician will monitor for reactions associated with IVIg treatment that might occur with Hizentra, including renal dysfunction/failure, thrombotic events, aseptic meningitis syndrome (AMS), hemolysis, and transfusion-related acute lung injury (TRALI).
Patients receiving Ig therapy for the first time, receiving a new product, or not having received Ig therapy within the preceding eight weeks may be at risk for developing reactions, including fever, chills, nausea, and vomiting. On rare occasions, these reactions may lead to shock.
Ig administration could impair the effect of live virus vaccines, such as measles, mumps and rubella. Before getting any vaccination, inform your doctor that you are being treated with Hizentra or Vivaglobin.
In their clinical studies, Hizentra and Vivaglobin were effective in pediatric patients without specific pediatric dose adjustments. With either treatment, no overall differences in safety or efficacy have been observed in patients over 65 or in pediatric patients.
Please see brief summary of full Prescribing Information for Hizentra and Vivaglobin, including Patient Product Information, on previous pages.
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.
*In a clinical study, the median length of a weekly infusion ranged from 1.6 to 2 hours.
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