Effects of Chronic Illness
Treating the Whole Family

Peripheral Neuropathy & Its Underlying Cause
Understanding & Treating Lupus

Medicare Reimbursement for IG
Boosting Kids’ Immune Systems with Vitamins
GAMUNEX®-C
Immune Globulin Injection (Human) 10% Caprylate/Chromatography Purified

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

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

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

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

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

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

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

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

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

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

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

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

Talecris Biotherapeutics, Inc.
Research Triangle Park, NC 27709 USA
U.S. License No. 1716

08939771/08939782-BS
Revised: October 2010
Important Safety Information for GAMUNEX-C

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

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

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

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

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

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

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

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

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

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

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

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

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


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

Please see adjacent page for brief summary of GAMUNEX-C full Prescribing Information.
A bout IG Livin g is the only m agazine dedicated to bringing com prehensive healthcare inform ation, immun e globulin inform ation, com m unity and reim bursem ent new s, and resources for successful living directly to imm une globulin consum ers and their healthcare providers.

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IG Livin g 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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TO DD LEVINE, M D
Phoenix Neurological Associates; Founder and Director, Samaritan ALS Clinic; Co-Director of the Neurophysiology Department, Banner Good Samaritan Medical Center; Clinical Assistant Professor, University of Arizona

Diagnosing and Treating Peripheral Neuropathy
“It is important to properly evaluate the symptoms of weakness, numbness and pain to identify what is causing the problem.”

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

Parenting: Packed with Essential Vitamins and Minerals
“Whether taken as a tablet or the old-fashioned way — in the form of numerous colorful fruits and vegetables — vitamins have an important effect on the body’s immune system.”

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

Diagnosing Specific Antibody Deficiency: The Effects of the Conjugated-Pneumococcal Vaccine, Part 1
“There has been a long-held belief that children do not respond to polysaccharide antigen vaccines, such as the 23-valent pneumococcal vaccine, until after 2 years of age. This is a dogma that is not true.”

ERIKA LAWRENCE, PHD
Associate Professor, Department of Psychology, University of Iowa

The Impact of Chronic Illness on the Family
“Family responses to chronic illness vary based on the age and developmental stage the ill individual, the strength and coping mechanisms of the family, and the family lifecycle stage.”

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

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

Connect with Other IG Living Readers through Monthly 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 rrhodes@IGLiving.com or calling (800) 843-7477, ext. 1362.
Editorial

The Rising Prevalence of Autoimmune Disease

When I was in my mid-20s, I started experiencing periodic numbness in my fingers. My fingertips would actually freeze and turn stark white from the knuckles down. Then, as they would warm up, they would turn a bright purple color. I did some research, and I concluded on my own that I had Raynaud’s phenomenon, an autoimmune disease that causes poor circulation in the fingers and toes. While I have never been formally diagnosed, now 25 years later, I still experience Raynaud’s symptoms on a daily basis, and I have learned to manage the disease myself. I know that what triggers it are changing climate conditions, such as walking from outdoors into the grocery store, or when my body starts to cool down after exercising. During these times, I avoid activities that require me to use my fingers until I can warm them by placing them under my armpits or under warm water when possible.

Some 5 percent to 8 percent of the population is affected by autoimmune diseases. There are an identified 80 clinically distinct autoimmune diseases, ranging from mild like mine to debilitating. What’s more, many people who are affected by autoimmune disease often have multiple conditions. For instance, in a report by the Autoimmune Diseases Coordinating Committee of the National Institutes of Health (NIH), one patient is profiled as having been diagnosed with Raynaud’s, antiphospholipid antibody syndrome, vasculitis, purpura and debilitating neuropathies. In fact, in some instances, many other diseases such as neuropathies are believed to be caused by autoimmune diseases.

Despite the rising prevalence of autoimmune diseases, which disproportionately affect women and are among the 10 leading causes of death for women in every age group up to 64 years of age, so little is still understood and known about them — including how to prevent, effectively treat and, ultimately, cure them.

In this issue, we take a look at lupus, one of the more serious autoimmune diseases that affects some 1.5 million Americans and can be fatal. What causes it, how it’s diagnosed and treated and what the future outlook is are all important for understanding this disease.

We also feature an article about peripheral neuropathy, written by Dr. Todd Levine, who specializes in neuromuscular diseases. Peripheral neuropathy affects 20 million Americans today. Yet, to properly diagnose and treat it, the underlying cause must first be identified. Dr. Levine explains the varying causes of the different types of peripheral neuropathy, including the immune-mediated neuropathies.

The impact of autoimmune diseases can’t be overstated. To put it in perspective, cancer affects 9 million people each year, heart disease affects 22 million people, and autoimmune disease affects 14 million to 22 million. The burdens imposed by these chronic, debilitating diseases include poor quality of life, high healthcare costs and substantial loss of productivity. In 2000, the NIH put in place a strategic plan called the NIH Initiative on Autoimmune Diseases. So far, research has enhanced our knowledge about the underlying causes of autoimmune diseases, it has helped to develop more effective therapies, and it has resulted in strategies for preventing their onset. However, much more needs to be done to stop the rising prevalence of these chronic disorders.
Cancer Risk for PIID Patients Corrected

I recently reviewed the latest issue of *IG Living* and had a major concern regarding one of the articles: The Link Between PIID and Skin Cancer [February-March 2012, p.26]. While I think this is a valid issue to address, some statements of “facts” are simply incorrect and unsupported by data or medical literature. Specifically, I have concerns about statements made regarding CVID, as this is my area of clinical expertise.

The article states: “In addition to skin cancer, CVID patients also are more susceptible to breast, prostate, gastrointestinal tract and lymphoid system cancers.” No references are provided to support this statement; this is because there is no scientific evidence to support most of this statement. Large studies of CVID patients have shown an increased risk of lymphoma and GI malignancy, but to my knowledge there is no compelling data to show an increase in skin cancer and absolutely no data linking breast and prostate cancer to CVID.

In my view, this issue should be addressed in publication, either by providing the scientific references to support these statements about CVID or with a retraction. You have a large readership and they accept the medical validity of what is written in the publication. This article is misinforming CVID patients about their condition, which leads to a whole slew of problems.

I greatly appreciate your efforts in publishing *IG Living*. I believe it’s an excellent and much-appreciated resource for patients. However, we need to maintain the scientific integrity of the medical information provided. This article unfortunately doesn’t meet that goal.

— Marc Riedl, MD, MS, associate professor of medicine
Section Head, Clinical Immunology and Allergy
UCLA David Geffen School of Medicine

The Editor replies:

Thanks so much for your feedback and concerns regarding the article on PIID and skin cancer. This article’s author, Jennifer Kester, used two sources regarding the link between PIID and different forms of cancer.

The first source was taken from the U.S. National Library of Medicine at the National Institutes of Health website, which states: “Individuals with CVID are susceptible to malignancy, particularly lymphoma. In their series of 248 individuals, Cunningham-Rundles & Bodian [1999] found that nearly eight percent developed non-Hodgkins lymphoma (NHL); another one to two percent had Hodgkins lymphoma; and other individuals had 24 different cancers, including breast cancer, prostate cancer, squamous cell carcinoma, melanoma, and basal cell carcinoma” (http://www.ncbi.nlm.nih.gov/books/NBK1299).

The second source was the National Foundation to Support Cell Transplant Research website, which states: “CVID, common variable immune deficiency, is characterized by onset between 24 months of age and young adulthood with increased susceptibility to infection and diminished response to vaccines. Individuals may experience meningitis or other systemic bacterial infections, recurrent eye or skin infections and gastrointestinal symptoms including chronic diarrhea and bloating. They have increased susceptibility to malignancies, especially lymphomas, and may also develop breast cancer, prostate cancer and skin cancer” (http://www.nfctr.org/rar_childhood_disorders.php).

You are correct that the article should not have stated that CVID patients “are more susceptible” to skin, breast, prostate, gastrointestinal tract and lymphoid system cancers. This literature states that they are more susceptible to lymphomas only, yet they also may develop these other forms of cancers. Unfortunately, the writer misinterpreted the information she read, and we erroneously published it. Of course, we very much want to clear this up for our readers, so I’m glad you brought it to our attention.

Your feedback, opinions, suggestions and anything else you’d like to share are important! Email us at editor@IGLiving.com or visit www.IGLiving.com and go to the Be Heard/Feedback page to send your comments.
UpFront

Faces of IG Living

IG Living

On the first day of Christmas, my true love gave to me:
1 IV drip of IG.

Mary McDole, Beckah Prairie York, Natalie Romanoff, Dale Manning Cook, Julia Kegg, Jennifer Van Grinsven Crone, I.G. Living!, Dale Manning Cook, Jennifer Van Grinsven Crone, Tammie Spivey Allegro, Debbie Spencer

On the 12th day of Christmas, my true love gave to me:
12 very sweet nurses, 11 nice cold ice packs, 10 doctor visits, 9 squares of gauze, 8 vials for blood testing, 7 insurance calls, 6 tubes for sub q, 5 grams of Gammagard, 4 vital checks, 3 needle pokes, 2 helper drugs and 1 IV drip of IG.

IG Living

Hundreds of medical professionals nationwide in recent years have refused to care for patients unless they sign anti-defamation contracts. In the contracts, patients must agree not to post public comments about the provider, or risk being sued. What do you think of medical gag orders? Would you sign a contract that forbids you to post comments about a doctor’s quality of care?

Chad Shaffer

Under most circumstances, I think gag orders are wrong. Medical care and privacy issues are not quite the same thing, however. Whether patients realize it or not, [you] have already “signed” [your] own gag law contract that prohibits physicians from commenting on your care. It’s the HIPAA rules, and I am sure most of you sign something periodically at the doctor’s office. If you have one, then why shouldn’t the physician have similar protection? There are other ways you can express an untoward outcome or experience with a physician that maintains the confidential patient-doctor relationship. A patient can report issues to the state board for review and assessment. To post comments about your care in a public forum without the other person having the ability to respond just doesn’t make sense. If you are serious about a complaint, take it up with your local state board and let a fair investigation begin. What do you think?

Christine Kucelin

I think it’s wrong to be asked to sign it; however, I am sure there have been instances [in which] doctors have treated an insane or unreasonable person that may have posted comments that are not true. I think the gag order should specify that a patient will not post false things or [they will] be sued. I think we all have the right to post actual things. I would have to question the confidence of a doctor [who] would ask me to sign such a thing. Either [they] are doing [their] job, or [they’re] not.

Jerrie Wright

No. I have the right to refuse to sign anything that I feel will impact my quality of care. If [an] MD [is] offering the paper to sign, I would ask them what they have to gain from it. An MD who took their oath for what it meant would not, in my opinion, have to ask [me] to sign something like this. It is a shame that in the U.S., we have MDs who are there for the monies. Our child has juvenile dermatomyositis, and we have so far been blessed to have wonderful MDs to provide her care. They are here for her and our family as we go through the changes this disease has [brought about]. We do brag about them and the students they have working with them. Quality of life should always be the No. 1 reason [for being] an MD.

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.
THE IMMUNE SYSTEM is designed to recognize parts of proteins from pathogenic organisms as foreign antigens. Proteins are polymers constructed of some 20 different amino acids as the building blocks that have structural and functional roles in all organisms. While many proteins are very similar among all organisms — from viruses to human beings — there are some differences, and the immune system is capable of recognizing these differences in order to generate an immune response. These differences in the proteins’ amino acid sequences and structures are known as “epitopes.” The ultimate result of the immune response is the formation of specific antibodies that recognize the specific epitopes present on the proteins from the pathogenic organisms.

For reasons that are not totally clear, a newborn has a good capacity to respond to protein antigens, but not so much to polysaccharide antigens. This is likely due in large part to the fact that the T lymphocytes of the immune system do not directly recognize polysaccharide antigens in order to respond to them. The ultimate formation of anti-polysaccharide antibodies may be somewhat of a “bystander effect” as the immune system is becoming activated against a protein antigen. In other words, if there is a B lymphocyte, which can respond to a specific polysaccharide antigen, in the vicinity of a T lymphocyte-B lymphocyte pair that are responding to a specific protein antigen, the former B lymphocyte may get caught up in the milieu of activating cytokines and be induced to produce anti-polysaccharide antibodies. This process may be somewhat happenstance, therefore taking a longer period of time for generation of a large repertoire of useful anti-polysaccharide antibodies to occur — say by 2 years of age.

Based on these issues, there has been a long-held belief that children do not respond to polysaccharide antigen vaccines, such as the 23-valent pneumococcal vaccine, until after 2 years of age. This is a dogma that is not true. Studies performed by the World Health Organization (WHO) on infants in Gambia and the Maori of New Zealand indicate that by 3 months to 6 months of age, infants can generate significant responses to the non-protein-conjugated 23-valent pneumococcal vaccine; although the responses may not be as vigorous as those from a non-immunodeficient adult, they can be sufficient to provide protection against disease.

Due to the way the immune system responds and matures, one of the paradoxes of the immune system development arises. Previously, it was noted that severe infections arising from pneumococcal bacteria peak in number by approximately 1 year of age, and decrease thereafter. This may in part be due to the description above, where there is exposure to the pneumococcal bacteria in a non-disease-inducing status, whereby infections do not occur, but antibodies can be produced. Under normal circumstances, these anti-pneumococcal antibodies subsequently lessen the chance and severity of the infections due to pneumococcus, but only after 1 year of age. Therefore, a period of time exists in which infants are at significant risk for bacterial infections due to the lack of anti-polysaccharide antibodies. This is the time from which their maternal antibodies disappear to when they have generated sufficient antibodies for protection against infection.

Taking these concepts into consideration, it is possible to induce an earlier significant anti-polysaccharide antibody response. An example of this is some 30 years ago when an initial polysaccharide antigen was conjugated with a protein antigen. The first one of significance was to the polysaccharide antigen of Haemophilus influenzae type B (Hib) conjugated to the diphtheria toxoid protein. The goal was and remains: As the immune system responds normally to the diphtheria toxoid protein, the bystander effect elicits an immune response of antibody production to the Hib polysaccharide antigen. Prior to 30 years ago, severe infections to Hib were a leading cause of morbidity and mortality in infants and young children, with as many as 100 per 100,000 developing severe disease. Since the conjugated Hib vaccine introduction in the 1980s, this has dropped to approximately one per 100,000 — a truly dramatic effect.

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

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

**Medicare and IG**

Does Medicare pay for immune globulin (IG) therapy? This question ranks among the most frequently asked of IG Living by both patients and doctors. The easy answer is yes. Medicare does pay for IG when medically necessary for all FDA-approved indications, as well as a number of off-label indications. Unfortunately, how Medicare pays and under what circumstances is not nearly that simple.

**Alphabet Soup**

Medicare is commonly referred to as alphabet soup because the different plans are named as letters (i.e., Part A, Part B, Part C, etc.). Which letter covers IG and how much it covers depends primarily on the subscriber's plan choice, where the treatment takes place and what disease the patient has. For a list of disease states covered by Medicare, see the website address at the end of this article.

**Part B Plus Medigap**

Both intravenous IG (IVIG) and subcutaneous IG (SCIG) therapy are covered under Medicare Part B. This plan covers medically necessary services such as doctor appointments, durable medical equipment, preventive services and outpatient treatments. Payment and coverage decisions for IG under Medicare Part B is administered by one of the four regional Durable Medical Equipment Medicare Administrative Contractors (DME MACs), which are named regions I, II, III and IV. Part B covers 80 percent of the contracted rate for IVIG and SCIG.

In order to get more than 80 percent coverage, patients must have additional coverage such as a Medigap plan (also known as a supplemental plan). Medigap plans are administered by private insurers. In general, they cover the 20 percent gap that Medicare Part B does not cover. Unfortunately, for the disabled under age 65, federal law does not currently require insurance companies to sell Medigap plans to them. That requirement is left up to the states. As of this writing, 27 states mandate that a Medigap plan be made available to the disabled under age 65. To see a complete list of participating states, go to www.medicare.gov/Publications/Pubs/pdf/02110.pdf.

In lieu of a Part B plus Medigap plan, Medicare recipients may choose a Medicare Advantage plan (also known as Medicare Part C) or, if available, they can be privately insured through a working spouse or retirement plan.

**Part C/Advantage Plans**

Medicare Advantage plans are administered by private insurance companies. They are an inclusive plan that must cover traditional Medicare benefits (Parts A and B), with the exception of hospice, which falls back to traditional Medicare. Medicare recipients choosing an Advantage plan cannot have a Medigap plan. Medicare Advantage/Part C plans must cover at least the same as traditional Medicare. This means that Advantage plans must cover at least 80 percent of the contracted rate of IG under the medical part of the policy. Often, however, several plans cover more than 80 percent. Additionally, like traditional insurance, these plans typically have a maximum out-of-pocket after which coverage is 100 percent.

Keep in mind that even though Advantage plans must cover at least what traditional Medicare covers, no two plans are the same. Those considering Advantage plans need to clarify all the details before agreeing to...
purchase the plan. Last, many Advantage plans include drug coverage. Therefore, subscribers may not need to purchase an additional drug coverage plan.

Medicare Advantage plans may be an attractive alternative for those under age 65 who qualify for Medicare coverage due to a disability if they live in one of the states that do not offer Medigap plans.

Plan D/Prescription Coverage

Medicare Part D is for prescription drug coverage. Having a Part D plan in addition to Medicare Part B is known as a stand-alone drug plan. In general, patients with a disease other than primary immunodeficiency disease (PIDD) may receive IVIG under the Part D benefit. However, only the drug is covered; nursing and supplies are not generally covered under stand-alone Part D plans. Additionally, if receiving IVIG under Part D, there is a coverage gap, not so affectionately known as the doughnut hole, in which the patient is financially responsible for 100 percent of the charges until they reach their out-of-pocket maximum.

Drug Coverage Under Advantage Plans

While an Advantage plan also may include a drug benefit, it is not considered a stand-alone plan. As a result, the plan may differ from traditional Medicare Part D plans. Consequently, PIDD patients using SCIG may be able to access their medication using the prescription benefit of an Advantage plan. Patients doing this, however, could be subject to higher out-of-pocket costs than traditional Medicare plus Medigap. Advantage plan members should clarify with their IG provider as to which benefit they are accessing to pay for services before agreeing to receive the medication.

Where Treatments Take Place Matters

Whether IG is covered under Part B, Part C or Part D, where the treatment occurs makes a difference. Additionally, the amount that is reimbursed can and does vary depending on the setting. All infusion providers and specialty pharmacies are required to use a specific code that tells Medicare where the IG is infused. If the place of service code does not match Medicare policies, the entire claim could be denied.

Home. Currently, Medicare will cover immune globulin in the home under certain circumstances:

1. If the patient is certified home-bound and the diagnosis is one covered under Medicare, nursing and the medication are covered under Medicare Part B.
2. If the patient has a PIDD and uses IVIG, only the drug is covered in the home setting. Nursing and supplies are the responsibility of the patient.
3. If the patient uses an IG product that is FDA-approved to be infused subcutaneously, the medication and supplies are covered. However, nurse training in the home setting is not covered.
4. If the patient has a disease that is not PIDD, such as chronic inflammatory demyelinating polyneuropathy (CIDP) or polymyositis, for example, coverage for the drug is available only under Part D. Again, nursing and supplies are the financial responsibility of the patient. Because of the aforementioned doughnut hole, this may not be an attractive option for patients.

Doctor’s office/infusion clinic. Many patients find comfort in knowing that a doctor or a well-trained nurse is close by during their infusion. Medicare will cover IG infusions in a clinical setting as long as the provider is a contracted Medicare provider. The types of clinics that may provide IG therapy include cancer centers, rheumatology clinics and neurology clinics. Reimbursement for IG in this setting should fall under Part B for traditional Medicare or the medical portion of an Advantage plan policy.

Outpatient hospital. Medicare will cover infusions under Part B in an outpatient hospital setting as long as the hospital is a contracted Medicare provider. For Advantage plans, care should fall under the medical portion of the plan. All services and supplies needed for the infusion should be included in the coverage.

Confusion Prevails

Even though Medicare does cover IG therapy, the confusion over how it is covered still worries patients and providers. And, because medicine is in a constant state of evolution, the rules will always be subject to change. Patients and providers, therefore, are wise to gather as much information as possible to help them make the most informed decisions.

Source: http://www.medicare.gov
Medicare Coverage Database. www.cms.gov/medicare-coverage-database/details/article-details.aspx?articleId=50875&ver=9&ConIdr=238&ConVr=2&CntrtrSelected=238*2&Cov erageSelection=Both&ArticleType=All&PolicvType=Final&s=All&KeyWord=IVIG&KeyWordLook Up=Title&KeyWordSearchType=And&clickon=search&bc=gAAAAABAAAAA&
Researchers at St. Michael’s Hospital in Toronto, Ont., Canada, have identified a potential new cause for unexplained miscarriages in mice. In addition, they have identified two possible treatments to prevent these miscarriages, which have broader implications for the development of new drugs to treat heart attacks and strokes.

The researchers found that the same kind of blood clotting in coronary arteries or blood vessels in the brain that causes heart attacks and strokes also happens in the placenta. The massive clotting can destroy the placenta, block blood flow to the fetus and cause miscarriages. This condition is known as fetal and neonatal immune thrombocytopenia (FNIT), a bleeding disorder that affects between one in 800 and one in 1,500 live births (most commonly among Caucasians) in which mothers generate antibodies that attack and destroy platelets in their fetuses and newborns. In severe cases, FNIT may lead to bleeding in the brains of the fetuses and newborns and cause neurological impairment or even death.

Seventy-five percent to 95 percent of FNIT cases are caused by maternal antibodies to one specific platelet antigen, HPA-1. However, these researchers discovered that another antigen, HPA-2, causes a type of FNIT never described before that can lead to miscarriages in more than 83 percent of mice. They also discovered that the HPA-2 antibodies sometimes not only destroy platelets, but activate them and cause massive clotting in the placentas. Because only six to eight reported live human births in the world with FNIT caused by HPA-2 have been reported, this research suggests that the reason these cases are so rare is that most of the affected fetuses died through miscarriages, before doctors examined them.

In mice, these miscarriages can be prevented using at least two therapies. One is the transfusion of immune globulin (IVIG). The other is the transfusion of an antibody known as anti-FcRn, which blocks the attacking maternal antibodies from crossing the placenta.

The research was reported on in the November issue of the Journal of Clinical Investigation. It is hoped that the findings will be important in the development of safer anti-thrombotic drugs, which are under development by several companies.

Researchers at the Laboratory of Allergic Diseases at the U.S. National Institute of Allergy and Infectious Diseases (NIAID) have identified a new immune disease, which they have named PLCG2-associated antibody deficiency and immune dysregulation (PLAID). Symptoms of the disorder include immune deficiency, inflammatory skin disorders and cold-induced hives (cold urticaria). The team identified the genetic mutation that causes PLAID in 27 members of three unrelated families who suffered from cold urticaria.

“This is one of the few examples in which the allergy symptom directed us to a genetic syndrome,” says study leader Dr. Joshua Milner. “In trying to understand the link between this group of conditions — autoimmunity, chronic infections and cold urticaria — we not only identified a disease-causing mutation, but uncovered a unique and fascinating genetic mechanism at the crux of allergy, immune defense and self-tolerance.” The research was published online in the January 11 edition of the New England Journal of Medicine.
**Vaccines**

**Vaccine May Halt Autoimmune Disease**

A synthetic vaccine based on nanotechnology may halt autoimmune diseases such as Crohn’s and rheumatoid arthritis. The vaccine works by tricking the immune system into producing antibodies that target an enzyme that causes autoimmune diseases.

With autoimmune diseases, some members of the enzyme family, especially the enzyme matrix metalloproteinases (MMP), get out of control. MMPs are normally held in check naturally by inhibitor molecules called TIMPs, and previous attempts to mimic TIMPs with artificial drugs have produced serious side effects. Rather than target the MMPs directly, researchers at the Weizmann Institute in Rehovot, Israel, created tiny metallic vaccine molecules that fool the immune system into manufacturing its own MMP-suppressing antibodies. When tested on mice with a rodent version of Crohn’s, the vaccine significantly reduced their symptoms. Untreated mice suffered severe damage to their colons, while those injected with the vaccine experienced only “limited” inflammation.

More research is needed before experts can be sure the therapy is safe for humans. The research was published in the journal *Nature Medicine*.

**Healthcare**

**U.S. Sets New Goals for a Healthier Nation**

In October, the U.S. Department of Health and Human Services (HHS) released a list of critical health priorities for the coming decade designed to serve as a blueprint to help reach the Healthy People 2020 objective of improving the health of all Americans. The goals are designed to help policymakers at the federal, state and community level make priorities for the coming decade.

According to HHS Assistant Secretary for Health Howard Koh, MD, MPH, the top priorities are expanding access to medical care and increasing the number of Americans with their own primary care provider. Other goals include increasing the percentage of eligible Americans who are screened for colorectal cancer from the current 54 percent to 70 percent and the percentage of eligible women who have mammograms from 70 percent to 77 percent; increasing the percentage of people with high blood pressure and diabetes whose conditions are adequately controlled with medication; increasing the percentage of young teens who receive booster doses of the tetanus-diphtheria-acellular pertussis vaccine from 47 percent to 80 percent and increasing the vaccination rate with two doses of the varicella vaccine in this age group from 37 percent to 90 percent; and increasing the number of Americans who see a dentist regularly to around 49 percent, from a current rate of about 44 percent. For the first time, the goals include a section identifying social factors that help determine health. For instance, a major goal is to increase the percentage of students who graduate from high school with a regular diploma in four years from around 75 percent to 82 percent, a move made in recognition of the fact that higher education is closely linked to better health.

Koh, who presented the list of priorities at the annual meeting of the American Public Health Association, noted that over the previous decade, the average life expectancy of Americans has increased from 77 years to 78 years. And, three out of four health objectives identified by health officials to be met by 2010 were either met or substantial progress was made toward meeting them.
CSL Behring has received a 2012 EURORDIS (European Organization for Rare Diseases) Award for its pioneering work in developing and manufacturing therapies used to treat rare and serious medical conditions. In 2011, the company received the National Organization for Rare Disorders Corporate Award for new treatments for rare diseases brought to market in the U.S.

Medicines

Grifols Offers Gamunex CoPay Card to CIDP Patients

The Gamunex CoPay Card Program by Grifols is a coupon program that helps patients with chronic inflammatory demyelinating polynuropathy (CIDP) cover the copay costs for Gamunex-C. The program provides up to $2,500 per patient per 12-month period, and it is open to CIDP patients who are not using any state or federally funded healthcare program such as Medicare, Medicaid, Medicare Advantage and Tricare; who are not residing or receiving treatment in the state of Massachusetts; and who are not getting treatments in hospitals or hospital-associated outpatient clinics. For additional information, go to gamunexcard.com.

Research

People with Job’s Syndrome Have Impaired Immune Memory

A research team at the National Institute of Allergy and Infectious Diseases (NIAID) has found that people with Job’s syndrome (also known as autosomal-dominant hyper-immunoglobulin E syndrome) have a lower number of immune memory cells, which makes them more susceptible to viral reactivation. The findings, which appear in the Nov. 23, 2011, issue of Immunity, provide a potential treatment strategy for people with Job’s syndrome, as well as offer clues about how immune cells in healthy people control chronic viral infections.

The NIAID team examined patients with Job’s syndrome to better understand how immune memory develops. Job’s syndrome is a condition caused by a mutation in a gene for the protein STAT3, which aids in the development and specialization of specific types of immune memory T cells. The team observed that, when compared with healthy people, patients with Job’s syndrome lack a major population of circulating memory T cells, which are thought to be a source for long-term T cell memory. These low numbers of central memory T cells are closely associated with these patients’ increased susceptibility to varicella-zoster virus (VZV) reactivation, causing them to have a significantly higher chance of developing shingles at a young age (less than 50 years old) and of experiencing repeated episodes of shingles compared with healthy people. What’s unique about this finding is that people with Job’s syndrome do not typically experience severe chicken pox or have difficulty clearing the initial infection. This means that Job’s syndrome is one of the few diseases that predispose patients to developing shingles, but it does not affect their response to chicken pox.

Based on these findings, the researchers concluded that measuring circulating central memory T cells, or STAT3 function, could be a way to identify someone who is at greater risk for developing shingles or could benefit from the shingles vaccine. In addition, new therapeutics that boost the activity of STAT3 also could help protect people from VZV reactivation. Further study is needed, they said, to determine if young, otherwise healthy people who experience episodes of shingles have impaired memory T cells. In addition, more research also is needed to better understand what level of immune memory is needed to protect people who have received the chicken pox vaccine from developing shingles.
Did You Know?

**Research**

**Positive Results for Phase II Clinical Trial for ITP Patients**

Symphogen has presented final Phase II data demonstrating that its recombinant polyclonal antibody drug candidate, rozrolimupab, exhibited a favorable safety profile and induced a rapid increase in blood platelets in patients with immune thrombocytopenia purpura (ITP). The Phase II study was an open-label, multicenter clinical trial evaluating the efficacy, safety and tolerability of rozrolimupab (SYM001) in adult RhD positive, nonsplenectomized ITP patients. Results demonstrated that at 300 ug/kg, eight of 13 (62 percent) of patients responded at day seven. The median time to respond was 59 hours, and the median duration of response was 14 days. The most common adverse events, mostly mild to moderate, were headache (18 percent), pyrexia (13 percent), chills (8 percent) and fatigue (8 percent). Four serious adverse events considered related to the drug were reported: decreased hemoglobin, extravascular hemolysis/dizziness and two cases of transient rise in D-dimer values without clinical symptoms.

**Research**

**DNA Sequence Causes Most Severe Lupus**

Researchers at the Catholic University of Sacred Heart in Rome found that a genetic accelerator known as HS1.2 is responsible for the most severe cases of lupus. HS1.2 leads to enhanced activation of the “transcription factor NF-KB” (a molecule that reads the genes to make them work), which in turn dramatically increases the aggressiveness of the inflammatory processes underlying the disease. The researchers likened HS1.2 to the accelerator of a car because it boosts the pathological immune response typical of the disease by enhancing the production of antibodies that attack the patient’s body instead of defending it. They found that 30 percent of lupus patients have the HS1.2 enhancer in their DNA. The discovery could lead to more targeted and effective therapies against lupus. The results of their study were published in the *Annals of Rheumatic Diseases*.

**Research**

**IDF Seeks Women Participants For New PIDD Survey**

The Immune Deficiency Foundation (IDF) is seeking female participants for a survey focusing on women’s reproductive/pregnancy issues as they relate to primary immunodeficiency disease (PIDD). The survey is made possible by an unrestricted educational grant to study the impact of PIDD on family planning, conception, fertility and pregnancy.

Study results will be used for research purposes, and to better inform physicians, patients and the parents of patients impacted by PIDD. It is hoped that the data obtained from this survey will help improve patient health outcomes and better inform patients and parents of those affected by PIDD on the management of these diseases before, during and after a pregnancy.

The survey is intended for women over the age of 18 who have a PIDD, or the mothers of patients with a PIDD. It is a web-based survey, and an invitation to participate will be sent to interested participants by email. If an invitation is not received, the IDF may not have an email to reach the participant, so participants are asked to contact IDF at (800) 296-4433.

**Did You Know?**

In May, The Texas Department of State Health Services added severe combined immunodeficiency (SCID) to the list of diseases that all newborns in Texas are screened for at birth.
Baxter has begun a Phase I prospective, open-label study that will assess the safety, tolerability and pharmacokinetics of its lead investigational candidate, BAX 855, a longer-acting (PEGylated) form of a full-length recombinant factor VIII (rFVIII) protein, in previously treated patients aged 12 years or older with severe hemophilia A. When used for prophylaxis, Baxter’s Advate requires patients to infuse every two to three days to reduce the occurrence of bleeding episodes. This Phase I trial is the first step in assessing whether BAX 855 can be infused less frequently.

Baxter has begun a second Phase III clinical trial of its Gammagard Liquid 10% Immune Globulin Infusion (Human), which is marketed as Kiovig outside the U.S. and Canada, for the treatment of mild to moderate Alzheimer’s disease. This second Phase III trial is identical in design to the first Phase III trial, and it will assess the safety and effectiveness of Gammagard Liquid as a potential treatment for signs and symptoms associated with Alzheimer’s disease.

Neovacs has announced the full results for its TNF-K-003 clinical trial with TNF-Kinoid in rheumatoid arthritis. According to the company, the results confirm the very good safety profile of the TNF-Kinoid. No patient withdrew from the study because of an adverse event, and no Kinoid-related serious adverse events were reported.

Trubion Pharmaceuticals has started a clinical trial of a drug for lupus. SBI-087 is made to hit a target called CD20, which is currently blocked by Genentech and Biogen Idec’s rituximab (Rituxan) for patients with a different form of autoimmune disease, rheumatoid arthritis. The Trubion drug, which is also being tested for rheumatoid arthritis, is being developed in partnership with Madison, N.J.-based Wyeth.

In March, CSL Behring enrolled its first patient in the PATH study, an international clinical trial designed to evaluate the efficacy, safety and tolerability of two different doses of subcutaneous immunoglobulin (SCIG), compared with placebo, in maintenance treatment of chronic inflammatory demyelinating polyneuropathy (CIDP).
Researchers at the University of Oklahoma are collaborating on a new, multisite, placebo-controlled study testing the effectiveness of intravenous immunoglobulin (IVIG) for reducing obsessive compulsive disorder (OCD) symptoms in children with pediatric autoimmune neuropsychiatric disorder associated with streptococcus (PANDAS).

Previous human and animal research suggested mechanisms by which strep-triggered antibodies mistakenly attack specific brain circuitry, resulting in obsessional thoughts and compulsive behaviors. This occurs when the immune system’s antibodies react not only to strep, but also to mimicked molecules. These “cross-reactive anti-brain antibodies can cause OCD, tics and the other neuropsychiatric symptoms of PANDAS,” according to lead researcher Susan Swedo, MD, of the National Institutes of Health’s (NIH) National Institute of Mental Health, who first characterized PANDAS two decades ago. According to Swedo, researchers “predict that IVIG will have striking benefits for OCD and other psychiatric symptoms, and will prove most effective for children who show high levels of anti-brain antibodies when they enter the study.”

Prospective study participants are first screened by phone by investigators at the NIH or the Yale Child Study Center, where the study is occurring. Those who meet eligibility requirements are then randomized to receive either IVIG or a placebo during a brief inpatient stay at the NIH Clinical Center. The researchers remain blind to which children receive IVIG; after six weeks of placebo control, they give any children whose symptoms fail to improve the option to receive open-label active treatment with IVIG.

In addition to assaying for antibodies that attack brain cells, the researchers use magnetic resonance imaging to see if the treatment reduces inflammation in an area of the brain known as the basal ganglia, which is thought to be the target of the errant antibodies. They also analyze levels of immune system chemical messengers (cytokines) in cerebrospinal fluid and blood — with an eye to identifying biomarkers of disease activity and potential predictors of treatment response.

Acute onset OCD in children with PANDAS is known as pediatric acute-onset neuropsychiatric syndrome (PANS). PANS expands on PANDAS, which is limited to a subset of cases traceable to an autoimmune process triggered by a strep infection. Currently, criteria for a broadened diagnosis of PANS have been proposed, including: 1) abrupt, dramatic onset of OCD or anorexia, 2) concurrent presence of at least two additional neuropsychiatric symptoms with similarly severe and acute onset (including anxiety; mood swings and depression; aggression, irritability and oppositional behaviors; developmental regression; sudden deterioration in school performance or learning abilities; sensory and motor abnormalities; and somatic signs and symptoms), and 3) symptoms are unexplainable by a known neurologic or medical disorder.

The PANS criteria grew out of a PANDAS workshop convened at NIH in July 2010 at which clinicians reported that evaluations of more than 400 youth diagnosed with PANDAS confirmed that affected boys outnumbered girls two to one with psychiatric symptoms, always including OCD, usually beginning before 8 years old.
Research

Chinese Herb Remedy May Treat Autoimmune Diseases

Researchers at the Harvard School of Dental Medicine have discovered the molecular secrets behind the Chinese herbal remedy known as chang shan. The researchers investigated halofuginone (HF), a compound derived from this extract’s bioactive ingredient, and discovered that HF triggers a stress-response pathway that blocks Th17 cells, which means it may be used to treat autoimmune disorders.

In 2006, Th17 immune cells were discovered to be “bad actors” implicated in many autoimmune diseases such as inflammatory bowel disease, rheumatoid arthritis, multiple sclerosis and psoriasis. In this new study, which was published online in the Feb. 12 issue of *Nature Chemical Biology*, researchers investigated how HF activates the amino acid response pathway (AAR). By looking at the most basic process that cells use to translate a gene’s DNA code into the amino acid chain that makes up a protein, the researchers were able to home in on a single amino acid, called proline, and discovered that HF targeted and inhibited a particular enzyme (tRNA synthetase EPRS) responsible for incorporating proline into proteins that normally contain it. When this occurred, the AAR response kicked in and produced the therapeutic effects of HF treatment. Providing supplemental proline reversed the effects of HF on Th17 cell differentiation, while adding back other amino acids did not, establishing the specificity of HF for proline incorporation. Added proline also reversed other therapeutic effects of HF, inhibiting its effectiveness against the malaria parasite, as well as certain cellular processes linked to tissue scarring. Researchers do not yet fully understand the role that amino acid limitation plays in disease response or why restricting proline inhibits Th17 cell products.

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Many different coping strategies can help family members deal with the stresses of chronic illness. But when coping strategies aren’t enough, help should be sought.
A family develops a kind of homeostasis — a normal dynamic and routine — that is disrupted when a member of that family develops a chronic illness. Indeed, chronic illness changes family members’ roles, responsibilities and boundaries. It disrupts their self-images and self-esteem. It results in uncertain and unpredictable futures. And it triggers distressing emotions — anxiety, depression, resentments, feelings of helplessness, as well as illness-related factors such as permanent changes in physical appearance or bodily functioning.

How a family responds to chronic illness varies based on the age and developmental stage of the ill individual, the strength and coping mechanisms of the family, and the family life-cycle stage. There are many different ways that a chronic illness can affect a family. The person who is chronically ill may feel guilty about the demands his or her illness makes on the family. He or she may resent the change in roles and responsibilities caused by the limitations imposed by the illness, and he or she must deal with the threat to his or her autonomy and the need to depend on healthy family members.

The spouse or partner of a person with a chronic illness can be faced with dual challenges: 1) as the primary provider of support to the ill partner and 2) as a family member who needs support in coping with the illness-related stresses he or she is experiencing. The burdens of being the primary caregiver may take their toll. The spouse may feel trapped while trying to balance dependence and autonomy of the patient with his or her own needs. He or she may feel tired or emotionally drained by the long duration of the illness or the extent of the caregiver workload. The spouse may struggle with feeling powerless when his or her partner is in pain, or by the pressure to be emotionally strong. There may be concerns about the consequences of the illness for the spouse, his or her partner and, if relevant, the children. And, he or she may have to restructure family roles and responsibilities as the disease progresses or presents new challenges.

Having a child with a chronic illness affects the parents in unique ways as well. Some studies suggest that having a child with a chronic illness has a negative impact on the relationship, including lack of time with the spouse, communication problems, higher divorce rates, increased relationship conflict, increased role strain, and decreased relationship satisfaction. Yet, other studies indicate that there are no effects, and still others have found positive effects including increased closeness, greater cohesion and increased support. Indeed, rates of divorce are lower among couples who have a child with a chronic illness. However, the impact that having a child with a chronic illness has on the adult relationship depends on the severity, course and prognosis of the illness, as well as on the quality of the relationship before the child became ill.

Given all of these adjustment demands, one might expect that the presence of a chronic illness would inevitably result in significant emotional difficulties and breakdown in family functioning. But, despite the presence of conditions and situations that are clearly traumatic and disruptive, a substantial proportion of families make satisfactory if not magnificent adjustments.

The person who is chronically ill may feel guilty about the demands his or her illness makes on the family.

Effective Ways for Couples to Cope

Most people talk about coping as problem-focused (taking care of what needs to get done) or emotion-focused (trying to reduce emotional distress). However, there is a third type of coping that is critical for couples or families faced with a chronic illness: “relationship-focused coping.” Relationship-focused coping means focusing on maintaining the quality of the relationship as part of the coping process. When faced with a stressful situation, each partner may attend to the other’s emotional needs in order to maintain the integrity of the relationship. Partners endeavor to manage their own distress without creating upset or problems for the other partner. Relationship-focused coping involves a balance between self and other, with the goal of maintaining the integrity of the relationship above either spouse’s needs. Effective strategies include negotiating or compromising, considering the other person’s perspective and being empathic. Specific strategies include:

View the illness as a couple or family problem: If both partners take a relationship perspective, they see the illness as a problem for the relationship, rather than just a problem
for one individual. They talk about the relationship as a way to cope and maintain the relationship. Couples who become aware of and discuss the relationship implications of a partner’s illness can anticipate how their relationship may change and prepare for the difficulties they may face. Couples who are resilient when faced with a chronic illness believe that they are in it together and serve as each other’s confidante, advisor and sounding board. Therefore, their attention should focus on the relationship as its own entity. For example, if the caregiver is thinking about how difficult it must be for his or her partner to be ill, then the focus is on the partner within the relationship but not on the relationship itself. With “relationship awareness,” the caregiver focuses on the relationship by telling his or her partner that the difficulties posed by the illness are “their” difficulties, thereby taking a relationship perspective in dealing with the illness.

Couples’ coping strategies can be effective if they are similar or complementary.

Use similar or complementary coping strategies: The goal of couple coping is to maximize the fit between partners’ coping styles in order to most effectively cope as a couple. Strategies that work in direct opposition or cancel out each other lead to poorer family dynamics. Couples’ coping strategies can be effective if they are similar or complementary, though. If partners use similar coping strategies, it might be easier to contend with stress. Coping efforts are coordinated and mutually reinforcing — that is, one partner’s efforts do not impede the other’s efforts. Complementary coping styles can be effective when they work together to reach a desired goal, e.g., by filling a coping “gap.” In fact, complementary strategies may be more effective than similar strategies because the couple, as a unit, has a broader coping repertoire.

What if each partner has very different coping strategies? Partners need to be aware of and talk about their own and their partner’s coping styles. The goal is to understand and respect each other’s ways of coping. Also, given that each has different coping styles, it is especially important to compromise, communicate about feelings to each other, give each other time alone, and reassure each other of their love and concern.

One relationship-focused coping strategy is protective buffering, which involves “hiding concerns, denying worries and yielding to the partner to avoid disagreements.” Although protective buffering is ostensibly used to avoid disagreements and “protect” the relationship, it can negatively affect the person using it because the partner may feel constrained in expressing negative emotions or worries. However, protective buffering doesn’t appear to harm the partner being “protected.” In general, it is necessary to balance taking care of oneself and the partner — for both the individuals and for the relationship.

Effective Ways for the Whole Family to Cope

Communicate with each other: Family members should communicate constructively about the illness and treatment. They should use active and empathic listening skills and consider other family members’ perspectives. When sharing something sensitive, they should be mindful of what is being communicated, how it is being said, and when it is communicated in relation to the others’ level of reception. They need to talk openly about the chronic illness, but not allow talk of the illness to dominate the family members’ daily lives.

Support each other: Effective support in a family involves more than just “being supportive.” First, different people want different types of support: Some people want practical help, others want to be listened to, and still others want to know that the other family members think they are strong enough or capable enough to handle things. More support is not necessarily better. What kind of support the family member wants needs to be understood and then provided. Second, family members need to learn to ask for the kind of help or support that is wanted. Each member of the family wants some kind of support. Others tend to provide the kind of support they would want, but they may not know what the others want. It shouldn’t be assumed that family members can read each other’s minds.

Increase and lean on social support outside of the family. It is well-known that having a social support network outside of the family benefits all members of the family, both physically and psychologically. Sometimes, just knowing they are available if needed — even if they are not turned to — can be helpful.

Integrate tasks of illness into the family’s daily routine. This will help the family develop a coordinated, cooperative
WARNING: ACUTE RENAL DYSFUNCTION AND ACUTE RENAL FAILURE

- Renal dysfunction, acute renal failure, osmotic nephrosis, and death may occur with immune globulin intravenous (IGIV) products in predisposed patients. Patients predisposed to renal dysfunction include those with any degree of pre-existing renal insufficiency, diabetes mellitus, age greater than 65, volume depletion, sepsis, paraproteinemia, or patients receiving known nephrotoxic drugs.

- Renal dysfunction and acute renal failure occur more commonly in patients receiving IGIV products containing sucrose. [1] GAMMAKED does not contain sucrose.

- For patients at risk of renal dysfunction or failure, administer GAMMAKED at the minimum concentration available and the minimum infusion rate practicable. (see Warnings and Precautions)
Brief Summary
See full Prescribing Information for complete information

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Indications
GAMMAKED consists of immune globulin injection (human) 10% liquid that is used:
- As replacement therapy of Primary Humoral Immunodeficiency (PI).
- To treat patients with Idiopathic Thrombocytopenic Purpura (ITP) to raise platelet counts to prevent bleeding or to allow a patient with ITP to undergo surgery.
- To treat Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) to improve neuromuscular disability and impairment and for maintenance therapy to prevent relapse.

Contraindications
- GAMMAKED is contraindicated in patients who have had an anaphylactic or severe systemic reaction to the administration of human immune globulin.
- GAMMAKED is contraindicated in IgA deficient patients with antibodies against IgA and history of hypersensitivity.

Warnings and Precautions
- Severe hypersensitivity reactions may occur with IGIV products, including GAMMAKED. In this case, discontinue GAMMAKED infusion immediately and institute appropriate treatment. Medications such as epinephrine should be available for immediate treatment of acute hypersensitivity reaction.
- Ensure that patients are not volume depleted prior to the initiation of the infusion of GAMMAKED. Periodic monitoring of renal function and urine output is particularly important in patients judged to have a potential increased risk for developing acute renal failure. Assess renal function, including measurement of blood urea nitrogen (BUN)/serum creatinine, prior to the initial infusion of GAMMAKED and at appropriate intervals. If renal function deteriorates, consider discontinuation of GAMMAKED. For patients judged to be at risk for developing renal dysfunction (e.g., any degree of pre-existing renal insufficiency, diabetes mellitus, age greater than 65, volume depletion, sepsis, paraproteinemia, or patients receiving known nephrotoxic drugs) administer GAMMAKED at the minimum infusion rate practicable.
- Do not administer GAMMAKED subcutaneously in patients with ITP because of the risk of hematoma formation.
- Hyperproteinemia, increased serum viscosity and hypotension may occur in patients receiving IGIV treatment, including GAMMAKED. It is clinically critical to distinguish true hypotension from a pseudohypotension that is associated with concomitant decreased calculated serum osmolality or elevated osmol gap.
- Thrombotic events have been reported following IGIV treatment and may occur in patients receiving IGIV treatment, including GAMMAKED. Patients at risk may include those with a history of atherosclerosis, multiple cardiovascular risk factors, advanced age, impaired cardiac output, coagulation disorders, prolonged periods of immobilization and/or known or suspected hyperviscosity. Consider baseline assessment of blood viscosity in patients at risk for hyperviscosity. For these patients, administer GAMMAKED at the minimum rate of infusion practicable.
- Aseptic Meningitis Syndrome (AMS) may occur infrequently with IGIV treatment, including GAMMAKED. Discontinuation of IGIV treatment has resulted in remission of AMS within several days without sequelae. The syndrome usually begins within several hours to two days following IGIV treatment. AMS is characterized by the following symptoms and signs: severe headache, nuchal rigidity, drowsiness, fever, photophobia, painful eye movements, nausea and vomiting. Cerebrospinal fluid (CSF) studies are frequently positive with pleocytosis up to several thousand cells per cu mm, predominantly from the granulocytic series, and with elevated protein levels up to several hundred mg/dL, but negative culture results. Conduct a thorough neurological examination on patients exhibiting such symptoms and signs including CSF studies, to rule out other causes of meningitis. AMS may occur more frequently in association with high doses (2 g/kg) and/or rapid infusion of IGIV.
- IGIV products, including GAMMAKED, may contain blood group antibodies which may act as hemolysins and induce in vivo coating of red blood cells (RBCs) with immunoglobulin, causing a positive direct antiglobulin reaction and, rarely, hemolysis. Delayed hemolytic anemia can develop subsequent to IGIV therapy due to enhanced RBC sequestration, and acute hemolysis consistent with intravascular hemolysis, has been reported. Monitor patients for clinical signs and symptoms of hemolysis. If signs and/or symptoms of hemolysis are present after GAMMAKED infusion, perform appropriate confirmatory laboratory testing.
- Noncardiogenic pulmonary edema may occur in patients following treatment with IGIV products, including GAMMAKED. Transfusion-related Acute Lung Injury is characterized by severe respiratory distress, pulmonary edema, hypoxemia, normal left ventricular function, and fever. Symptoms typically occur within 1 to 6 hours after treatment. Monitor patients for pulmonary adverse reactions. If TRALI is suspected, perform appropriate tests for the presence of anti-neutrophil and anti-HLA antibodies in both the product and patient serum. TRALI may be managed using oxygen therapy with adequate ventilatory support.
- The high dose regimen (1g/kg x 1-2 days) is not recommended for individuals with expanded fluid volumes or where fluid volume may be a concern.
- Because GAMMAKED is made from human blood, it may carry a risk of transmitting infectious agents. No cases have ever been identified for GAMMAKED. ALL infections suspected by a physician possibly to have been transmitted by this product should be reported by the physician or other healthcare provider to Talecris Biotherapeutics, Inc. [1-800-520-2807]
- After infusion of IgG, the transitory rise of the various passively transferred antibodies in the patient’s blood may yield positive serological testing results, with the potential for misleading interpretation. Passive transmission of antibodies to erythrocyte antigens may cause a positive direct or indirect antiglobulin (Coombs’) test.

Adverse Reactions
Clinical Trials
- PI - The most common adverse reactions (≥5%) with intravenous use of GAMMAKED were headache, cough, injection site reaction, nausea, inflammation of the throat, and hives. Vomiting was reported more frequently in pediatric patients. The most common adverse reactions (≥5%) with subcutaneous use of GAMMAKED were infusion site reactions, headache, fatigue, joint pain, and fever.
- ITP - The most common adverse reactions during clinical trials (reported in ≥5% of subjects) were headache, vomiting, fever, nausea, back pain and rash.
- CIDP - The most common adverse reactions during clinical trials (reported in ≥5% of subjects) were headache, fever, chills, hypertension, rash, nausea and weakness.

Postmarketing Experience
- Hemolytic anemia and aseptic meningitis have been identified and reported during the post marketing use of GAMMAKED.
- The following adverse reactions have been reported during the overall post marketing use of IGIV products:
  - Respiratory: Apnea, Acute Respiratory Distress Syndrome (ARDS), TRALI, cyanosis, hypoxemia, pulmonary edema, dyspnea, bronchospasm
  - Cardiovascular: Cardiac arrest, thromboembolism, vascular collapse, hypotension
  - Neurological: Coma, loss of consciousness, seizures/convulsions, tremor
  - Integumentary: Stevens-Johnson syndrome, epidermolysis, erythema multiforme, bullous dermatitis
  - Hematologic: Pancytopenia, leukopenia, hemolysis, positive direct antiglobulin (Coombs test)
  - General/Body as a Whole: Pyrexia, rigors
  - Musculoskeletal: Back pain
  - Gastrointestinal: Hepatic dysfunction, abdominal pain

Drug Interactions
- Passive transfer of antibodies may transiently interfere with the response to live viral vaccines, such as measles, mumps and rubella. This may confound serologic testing. Inform the immunizing physician of recent therapy with GAMMAKED so that appropriate measures may be taken.

Use in Specific Populations
- Pregnancy Category C. There is no human or animal data. It should only be given to a pregnant woman if clearly needed.
- Geriatric: In patients over 65 years of age, do not exceed the recommended dose, and administer GAMMAKED at the minimum infusion rate practicable.

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Research Triangle Park, NC 27709
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approach to dealing with the multiple demands of the illness and its treatment.

Take care of family members’ physical and psychological health. This is important for every family member. They need to learn relaxation or mediation techniques, exercise regularly, take some time alone, take time to get away from it all as a family, take walks and plan fun activities.

Find the new normal: The goal is to move from crisis management to incorporating the illness into the family’s daily lives. The fact is that life will never be exactly the same as it was before. Therefore, family members should challenge themselves to define what a “normal” family life is now. They should struggle to find ways to understand and make meaning of the experience.

Family therapy is particularly helpful early on.

When Should Help Be Sought?

If a significant amount of time has passed (a year or more), and “yes” can be said to one of the following, it may be time for a family to seek help:

• When the chronic illness colors every aspect of a family’s interactions.

• When partners have different coping strategies and cannot find common ground regarding the many demands of the illness.

• When one or more family members routinely withdraws into silence. This may or may not be helpful for the person, but it will not be helpful for the couple’s relationship or for the family.

• When one or more family members routinely takes on a reactive, anxiety-driven, tell-all communicative style of coping.

• When a family is still stuck in the “crisis phase” and not the ongoing process of adapting to a “new normal.”

There are many different kinds of help out there. Family therapy is particularly helpful early on. It has been shown to promote positive adjustment for families. It can help educate families about the person’s specific chronic illness and teach family members effective coping skills.

Child or adolescent therapy can be used to educate a child about his or her illness, and to teach stress-management techniques to promote healthy coping skills and create a buffer against stress. Child therapy also is recommended to help children or adolescents express and learn to cope with their emotions (fear, anger, sadness) when a parent is ill.

Group therapy or support groups can help caregivers, patients or parents of children with a chronic illness by reducing stress.

Certain types of couple therapy have been shown to be very effective at improving couples’ coping skills, at helping couples learn to engage in relationship-focused coping strategies, and at improving communication and support skills. Couple therapy has been shown to improve patients’, caregivers’ and couple health and functioning.

Make sure to find someone who is specifically trained in the type of therapy that is being sought — not someone who “does it all.” Also, ask if the therapist is specifically qualified to work with families in which one member is chronically ill. You have unique challenges to face and need an expert.

Coping Is Possible

Chronic illness can be extremely disruptive to family life. But, it can be possible to maintain a homeostasis by using the many coping strategies available, as well as seeking help when necessary.

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References


So little is known about this autoimmune disease, which affects more than 1.5 million Americans. But, improved testing and increased research are resulting in better treatment for lupus patients.

By Ronale Tucker Rhodes, MS

In 2006, Diane Sawyer aired a segment on “Good Morning America” to spread awareness about lupus. In the segment, she interviewed actress and model Kelly Jean Drury, who eight years earlier, at age 23, was diagnosed with the disease. Kelly is one of the 1.5 million Americans who have lupus. And, while this disease is widespread, occurring in one in 2,000 people, very few are even aware of it, and those who are rarely understand what it is.

Chances are, most people know someone who has lupus. In fact, there is a 20 percent chance of having a parent or sibling with lupus. Ninety percent of those with lupus are women, and the disease is two to three times more prevalent among people of color. While diagnosis typically occurs between the ages of 15 and 45, the disease can strike at any age, and many go undiagnosed. Those who do get diagnosed typically wait, on average, four years and have to see three different doctors to get diagnosed.
What Is Lupus?

Lupus is a chronic autoimmune disease that can damage any part of the body — skin, joints and/or organs. Lupus occurs when the immune system cannot tell the difference between viruses, bacteria and germs (foreign invaders) and the body's healthy tissues, and it creates autoantibodies that attack and destroy the healthy tissues. Those autoantibodies cause inflammation, pain and damage in various parts of the body. The disease can range from mild to life-threatening. It is not contagious, and it is not like or related to cancer or HIV.

There are five types of lupus. The most common, and the one most people are referring to when they say lupus, is systemic lupus erythematosus (SLE). This type affects many different organ systems, and is marked by chronic inflammation, especially of the kidneys, joints and skin. The cardiovascular and nervous systems also can be affected.

When lupus is limited to the skin, it is called chronic cutaneous lupus erythematosus (CCLE, also known as discoid lupus erythematosus [DLE]), subacute cutaneous lupus erythematosus (SCLE) or tumid lupus. While this type of lupus can exist in people who do not have systemic lupus, 5 percent or more of people with this type of lupus may develop SLE later in life.

Drug-induced lupus erythematosus is caused by certain drugs that cause lupus-like symptoms in people who do not have SLE. This form of lupus is temporary, and it usually subsides within months after the medication is stopped. Medications known to cause this type of lupus include blood pressure medications hydralazine and methyl-dopa; a heart medication called procainamide; a drug called D-penicillamine used to treat metal poisoning; minocycline, used to treat acne; and anti-TNF, used to treat rheumatoid arthritis. Many physicians are choosing not to prescribe these medications.

Neonatal lupus erythematosus may affect the babies of women who possess the autoantibodies anti-Ro, anti-La and anti-RNP. Only 40 percent of women bearing children with neonatal lupus actually have lupus, but lupus or Sjogren's syndrome may occur later in life. While neonatal lupus typically involves only the baby's skin and subsides on its own without treatment, 1 percent to 2 percent of these infants experience congenital heart block, which can be treated with the implantation of a pacemaker.

Childhood lupus affects the body in the same manner as adult lupus, yet boys are more likely to get childhood lupus than men are likely to get adult lupus. Childhood lupus also affects certain organs such as the kidneys to a greater degree, and the incidence of kidney disease is two times greater. It also requires more aggressive treatment than adult lupus.

Symptoms of Lupus

On the “Good Morning America” segment, Dr. Susan Manzi, co-director of the Lupus Center of Excellence at the University of Pittsburgh Medical Center, said: “Lupus patients are like snowflakes. No two present the same; no two look the same.” In fact, signs and symptoms may come on suddenly or develop slowly, they may be mild or severe, and they may be temporary or permanent. Most patients have episodes, called flares, when signs and symptoms get worse for a while and then improve or even disappear completely for some time.

Chances are, most people know someone who has lupus.

The symptoms of lupus depend upon what body organs are affected and how seriously they are affected. Those with SLE typically are extremely tired and have skin rashes or joint pain. In fact, nearly all people with lupus have mild to extreme fatigue, and increased fatigue is a sign that a flare is about to occur. Most people with lupus have joint pain at some time, and about 70 percent of lupus patients report that joint and muscle pain was their first sign of the disease. Most lupus patients also develop skin rashes, which are often an important clue to the diagnosis. Typically, a butterfly rash appears over the cheeks and bridge of the nose. Other common skin symptoms include skin sores or flaky red spots on the arms, hands, face, neck or back; mouth or lip sores; and a scaly, red or purple raised rash on the face, neck, scalp, ears, arms and chest.

Other symptoms of lupus include sensitivity to light, nervous system symptoms (most commonly headaches, but also difficulty with memory or concentration, or numbness or weakness of the arms or legs), heart problems, lung problems, mental health problems (anxiety and depression), low-grade fever, changes in weight, swollen glands, inflammation of blood vessels in the skin, swelling of the hands and feet, and anemia. Some people with
lupus also have Raynaud's phenomenon, which affects the small vessels that supply blood to the skin and soft tissues under the skin of the fingers and toes, causing them to turn white and/or blue or red. With Raynaud's, the skin affected will feel numb, tingly and cold to the touch.5

There also are other conditions that cause symptoms similar to lupus. These conditions include rheumatoid arthritis, glomerulonephritis (inflammation of the kidneys), scleroderma (a disease of the body's connective tissue), Sjogren's syndrome (a condition that causes dry mouth and dry eyes), chronic fatigue syndrome and vasculitis (inflammation of the blood vessels).6

Because no two people present with the same signs and symptoms, lupus can be difficult to diagnose.

Causes of Lupus

Dr. Manzi says that lupus patients often think they may have somehow “caught” the disease. But, lupus isn’t contagious. Nor is there a single moment that may have triggered the onset of the disease. In fact, doctors and researchers are unsure what exactly causes lupus, but they do believe that it is a combination of elements. The first is genetics. While there is no scientific proof, it is believed that heredity is at least one factor in determining an individual's propensity for developing lupus. However, if a person has a family history of the disease, he or she may be more susceptible to it, but it doesn’t mean he or she will get it.

The second element is the environment. It’s possible that certain environmental factors may trigger lupus. Those factors include exposure to ultraviolet light, smoking and stress, and exposure to toxins such as trichloroethylene in well water and silica dust. In the past, it was believed that certain hair products and topicals caused lupus, but that’s no longer true.

While research concerning the link between lupus and hormones is in its infancy, the findings are still nebulous, with some studies finding a link and others failing to find a link. However, abnormal estrogen metabolism is considered a risk factor. Viruses and bacteria also are considered risk factors. Individuals with viruses like cytomegalovirus, parvovirus and hepatitis C may develop lupus, but no causal link has been established. It is known that the Epstein-Barr virus in children is linked to childhood lupus. Last, long-term use of some medicines can trigger lupus and lupus flares.7

Diagnosing Lupus

Because no two people present with the same signs and symptoms, lupus can be difficult to diagnose. Plus, there is no one test that can diagnose lupus. The disease is typically diagnosed through a combination of laboratory tests, signs and symptoms, and physical examination findings.

Laboratory tests include both urine and blood tests. A complete blood count to measure the number of red and white blood cells and platelets, as well as the amount of hemoglobin (a protein in red blood cells), could show anemia, which commonly occurs in lupus. A low white blood cell or platelet count also may occur in lupus. A blood test to determine the erythrocyte sedimentation rate (the rate at which red blood cells settle to the bottom of a tube in an hour) can be conducted to see if there is a faster than normal rate, which may be a sign of a systemic disease such as lupus. Since lupus can affect organs, a blood test can assess how well an individual's kidneys and liver are functioning. And, a urinalysis may show an increased protein level or red blood cells in the urine, which may occur if lupus has affected the kidneys. Another lab test, the antinuclear antibody (ANA) test, looks for the presence of these antibodies, which may indicate a stimulated immune system. However, while most people with lupus have a positive ANA test, most people with a positive ANA test do not have lupus. So, more specific antibody testing may be needed.

Imaging tests also may be needed if it is suspected that lupus is affecting the lungs or heart. A chest X-ray can reveal abnormal shadows that suggest fluid or inflammation in the lungs. And, an echocardiogram, which uses sound waves to produce real-time images of the heart, can check for problems with valves and other portions of the heart. Because lupus can harm the kidneys in different ways and treatments can vary, in some cases, it may be necessary to take a biopsy of kidney tissue. The biopsy is obtained with a needle or through a small incision.8

Beyond lab tests, there are criteria developed for diagnosis of lupus. In 1997, the American College of Rheumatology (ACR) updated its criteria for the third time. According to the ACR, a patient may have lupus if four or

Treatment for Lupus

How lupus is treated also depends upon the disease signs and symptoms. Fortunately, treatments have increased dramatically in recent decades. Which medications are used to treat the disease is based on the patient's age, sex, health, symptoms and lifestyle. And, as signs and symptoms flare and subside, these medications and their dosages may need to be changed.9

Medications most commonly used to control lupus include nonsteroidal anti-inflammatory drugs (NSAIDs — either over-the-counter or those available by prescription), antimalarial drugs (such as hydroxychloroquine [Plaquenil]), corticosteroids (such as prednisone) and immune suppressants (such as cyclophosphamide [Cytoxan], azathioprine [Imuran, Azasan], mycophenolate [Cellcept], leflunomide [Arava], and methotrexate [Trexall]).10

Which medications are used to treat the disease is based on the patient’s age, sex, health, symptoms and lifestyle.

NSAIDs decrease inflammation, and they may be used alone or in combination with other types of drugs to control pain, swelling and fever. Antimalarials were originally used to treat malaria, but clinical studies have found that continuous treatment with antimalarials may prevent flares of lupus from recurring. Corticosteroids, closely related to a natural anti-inflammatory hormone, work by rapidly suppressing inflammation, and because they are so potent, doctors seek the lowest dose with the greatest benefit. Immunosuppressives restrain the overactive immune system by blocking the production of immune cells.

A few other therapies also are available. Belimumab (Benlysta), a B-lymphocyte stimulator (BLys) protein inhibitor, was approved by the U.S. Food and Drug Administration in March 2011 for patients with lupus who are receiving other standard therapies. It is given by infusion to reduce the number of abnormal B cells thought to be a problem in lupus patients. However, studies to date have shown that African-American patients and patients of African heritage do not respond to belimumab. Methotrexate (Folex, Mexate, Rheumatrex), a disease-modifying antirheumatic drug, may help control the disease in some patients. And, dehydroepiandrosterone (DHEA) and intravenous immunoglobulin (IVIG) may be used for controlling lupus when other treatments haven’t worked.9

Managing Day to Day

Lupus can significantly impact an individual’s quality of life. A study on work loss associated with lupus estimated that almost three-quarters of the study’s 982 participants would stop working before the usual age of retirement, and half of those who had jobs when they were diagnosed (during their mid-30s on average) would no longer be working by age 50.9

Most patients, like Kelly Jean Drury, live their lives day by day, never knowing what the future holds. Yet, lupus patients can maintain a high-quality lifestyle. First, it’s important that they learn the warning signs of a flare and take steps to reduce its intensity. Periodic increases in disease activity can usually be managed by varying medications. And, during flares, lupus patients should avoid ultraviolet light.11
They also must receive regular healthcare, rather than seeking help only when symptoms worsen. Women should receive regular preventive healthcare, such as gynecological and breast examinations. Men should have the prostate-specific antigen test. And, both men and women should have their blood pressure and cholesterol checked on a regular basis. If taking corticosteroids or antimalarial medications, an eye exam should be performed at least yearly. And, patients should be aware of their increased risk of premature cardiovascular disease.9

Most people with lupus will live a normal life span due to improved diagnosis and treatment.

Women with lupus are considered high risk. They have an increased risk of miscarriages and can have flares during pregnancy. Therefore, they require close observation during pregnancy, delivery and the postpartum period, including fetal monitoring by the obstetrician during later pregnancy. Those who are at risk for miscarriages can be identified with a test for the presence of phospholipid antibodies in the blood. If these antibodies are present in a woman with lupus, blood-thinning medications and IVIG for selected people can be used.11

Future Outlook

Researchers are working hard to answer several questions about lupus: Why are women more likely than men to have the disease? Why are there more cases of lupus in some racial and ethnic groups, and why are cases in these groups often more severe? What goes wrong in the immune system and why? How can immune system function be corrected once something goes wrong? What treatment approaches will work best to lessen lupus symptoms? And, how can lupus be cured?

The National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), a component of the U.S. Department of Health and Human Services’ National Institutes of Health, is funding researchers across the U.S. who are developing new and better ways to study the disease. These include laboratory studies that compare aspects of the immune systems of people with lupus with those of others both with and without lupus, as well as using mice with disorders resembling lupus to better understand the abnormalities of the immune system that occur in lupus and to identify possible new therapies.9

The Lupus Foundation of America is also on the forefront of fighting this disease. But, as Dr. Manzi explains in the “Good Morning America” segment, the foundation needs a Michael J. Fox for lupus; it has no spokesperson to be its advocate. “This is a very devastating disease,” says Dr. Manzi. “It can be fatal. Kelly’s very lucky that her disease is under control, but many people die of this disease. So, we need more research dollars; we need more help.”

The overall outlook for people with lupus is improving every decade. It is unknown whether the number of deaths attributed to the disease has been on the rise in the last 20 years due to the actual increase in mortality or just better identification and reporting of the disease. But, it is known that most people with lupus will live a normal life span due to improved diagnosis and treatment.7

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

References
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More than 20 million Americans have peripheral neuropathy. People who suffer from peripheral neuropathy are experiencing a malfunction in the way in which the nerves of the peripheral nervous system send signals from the brain to the arms and legs and vice versa. Their symptoms can present in many ways, from numbness to painful sensations, balance difficulties and weakness.

Localizing a Peripheral Neuropathy

Identifying the cause of peripheral neuropathy can be difficult. This is because the symptoms of peripheral neuropathy can be due to dysfunction in the parts of the peripheral nervous system or even diseases that affect organs other than peripheral nerves. A good analogy for patients experiencing symptoms would be to imagine they are placing a call from New York (their brain) to Los Angeles (their foot). During the call, the patients hear static, or in other words, they feel numbness or pain. But that static (the pain and numbness) is in fact abnormal nerve sensations. Unfortunately, these patients can’t tell if the problem is originating in New York, Los Angeles or in between. Identifying where the pain and numbness are originating is the first issue that needs to be addressed. Is there a chance the symptoms are coming from a problem with the brain, the spine, blood circulation, or even a problem in the bones or ligaments? Diseases in any of these parts of the body can mimic peripheral neuropathy.

The process of localization can involve several different tests. Typically, a neurologist will take a history of patients’ symptoms, perform a neurologic exam, and then order electromyography (EMG) and nerve conduction studies. These steps are crucial to understanding if the peripheral
nerves are involved. It also identifies several other important issues: how badly the nerves are damaged, which nerves are damaged and which parts of the nerves are damaged. In this way, localization goes from identifying the peripheral nerve as the problem to understanding how the disease process is affecting the nerves. One of the most important things to understand from this stage of the evaluation is how quickly the disease has damaged the nerves; for instance, has it developed over days, weeks or years?

The inner workings of a nerve can be broken down into three parts. The first part is the cell body, which sits in or near the spinal cord and is the machinery that keeps the cells working. If nerves of the peripheral nervous system are damaged, they can regrow as long as the cell body is alive. Once the cell body dies, the nerve can never regenerate. The second part is the axon, which is the long part of the nerve that extends from the spinal cord to the skin and muscles that the nerve supplies. Unlike all other cells in the body that are microscopic in all three dimensions, the peripheral nerve cells can be three feet long. These axons convey all the signals to move and to feel back and forth from the brain. The third part is the insulation around the nerves (myelin), which allows the nerves to quickly conduct the electrical signals. The more myelin wrapped around each axon, the bigger the nerve is and the faster the nerve conducts electricity — in some cases as fast as 350 miles per hour. By localizing the neuropathy, a neurologist can tell whether the damage is to the cell body, the axon or the myelin. And this is crucial because there are vastly different diseases that affect each part.

**Diagnosing and Treating the Types of Peripheral Neuropathy Diseases**

Once a neurologist understands the what (neuropathy) and the where (the cell body, the axon or the myelin), the next step is to figure out the why. This is the part that is still somewhat discouraging. Despite an ever-growing number of blood tests, spinal taps, nerve biopsies, genetic tests, etc., at least 50 percent of neuropathies remain undiagnosed. These types of neuropathies are called idiopathic (meaning, we are too idiotic to figure it out). One group of diseases that commonly affect peripheral nerves are: disorders of glucose metabolism such as diabetes, vitamin B12 deficiency, alcohol use and drugs such as chemotherapies. These diseases comprise about 30 percent of the other types of neuropathies, and tend to damage the axon. Approximately 10 percent to 15 percent of neuropathies may be inherited. These patients often have multiple family members with neuropathy and, unfortunately, inherited neuropathies are not treatable at the present time. The final group of diseases that affect the nerves are related to problems with patients’ immune systems. The immune system, which is supposed to fight bacteria and viruses, can start to work incorrectly and damage the nerves. These immune-mediated neuropathies are the most treatable.

**Despite an ever-growing number of blood tests, spinal taps, nerve biopsies, genetic tests, etc., at least 50 percent of neuropathies remain undiagnosed.**

Immune-mediated neuropathies can be subdivided into different groups based on the type of nerve that is damaged: motor, sensory, motor and sensory, or autonomic. They are diagnosed based on characteristic changes on nerve conduction studies, or abnormalities in blood tests, spinal taps or nerve biopsies. At times, it can be difficult to be certain that a neuropathy is caused by the immune system. In these cases, doctors may opt for a short trial of a therapy designed to treat immune neuropathies. If patients respond, then it is likely their neuropathy is immune-mediated. Should patients fail to respond to the first trial of therapy, it is still possible they have an immune-mediated neuropathy. However, repeated failures may indicate the neuropathy is untreatable. One final point is that even if patients have diabetes, they may have a superimposed immune-mediated neuropathy that is treatable. So for diabetics, if a neuropathy starts to rapidly get worse or to involve a lot of motor weakness, a more aggressive evaluation should be undertaken; the neuropathy should not simply be blamed on the diabetes.

Immune-mediated motor neuropathies are relatively rare. The most common is multifocal motor neuropathy. These patients have weakness, typically beginning in their arms, and have no numbness or pain. They respond very well to intravenous immune globulin (IVIG) and often can fully recover. The IVIG usually has to be continued indefinitely.
Although not classically a neuropathy, one disease that presents as pure weakness (no numbness or pain) is myasthenia gravis (MG). MG is caused by specific autoantibodies that inhibit a part of the muscle fiber to react to the nerve stimuli to move. As the disease worsens, patients experience weakness that can be in their arms or legs, in their eyes (which can cause double vision or droopy eyes), or even in their muscles that allow them to swallow, talk or breathe. Untreated, this can be a fatal disease. MG can be diagnosed by nerve conduction studies, but in almost all cases, an abnormal antibody can be identified in the blood of these patients. When the symptoms are severe, resulting in talking, swallowing and breathing problems, patients are typically treated acutely with either IVIG or plasmapheresis. Over the long run, most patients can be transitioned to oral drugs such as corticosteroids or one of several organ transplant rejection drugs to lower the level of the antibodies.

Patients must remember that even in the best hands, 50 percent of neuropathies will have no identifiable cause.

The immune-mediated pure sensory neuropathies are very rare and present as numbness and pain without weakness. In some cases, only the smallest nerves (those nerves with no myelin) are damaged. In these cases, the nerve conduction studies and exam may be normal. However, these neuropathies can be diagnosed with a skin biopsy that allows the pathologist to examine the small nerves to see if they are healthy. If these so-called small-fiber neuropathies, or pure sensory neuropathies, are related to an immune disorder such as Sjögren’s disease, they can respond to drugs that modulate the immune system such as IVIG, steroids or plasmapheresis. Tying the pure sensory neuropathies to an immune system disorder can be very challenging, and physicians often rely on abnormal blood tests and a strong clinical suspicion. In the right setting, a short course of IVIG (three months or so) to see if the symptoms can improve is very warranted and has little long-term risk.

There are many different types of motor and sensory neuropathies caused by the immune system. If they present very rapidly, causing weakness and sensory loss over a two-week period or so, it is called acute inflammatory demyelinating polyneuropathy, or Guillain-Barré syndrome. These patients may have severe weakness and may even require a ventilator to breathe. They are treated with one dose of IVIG or plasmapheresis, which allows their nerves to slowly recover and, in most cases, fully recover. If the disease presents more slowly (over more than six weeks), it is called chronic inflammatory demyelinating polyneuropathy (CIDP). These patients can be subdivided further in some cases based on the identification of certain specific blood tests or patterns on the nerve conduction studies. There are many treatment options available for CIDP patients, including IVIG, corticosteroids, chemotherapy drugs, drugs to prevent organ transplant rejection, etc. The key is to find drugs that the patients respond to and that they can tolerate. In most cases, patients will require the therapy indefinitely or at least for several to many years. Therefore, tolerability, expense and convenience all need to be considered by the patients and the neurologist.

The Importance of Proper Diagnosis

It is important to properly evaluate the symptoms of weakness, numbness and pain to identify what is causing the problem. Once patients have been definitively diagnosed with peripheral neuropathy, a neurologist familiar with nerve diseases should tailor the evaluation to the type of neuropathy and see if they can uncover the cause. If the cause is believed to be the immune system, there are several treatment options, depending on the type of neuropathy. Finally, and perhaps most importantly, patients need to be their own advocates. They should ask their doctor what type of neuropathy they have and what he or she believes is causing it. If their neuropathy is rapidly getting worse and the neurologist is not conducting additional tests to uncover the reason, then they should get another opinion. However, while this is unfortunate, patients must remember that even in the best hands, 50 percent of neuropathies will have no identifiable cause.

TODD LEVINE, MD, is a member of Phoenix Neurological Associates, Phoenix, Ariz., with a subspecialty practice in neuromuscular diseases. He is the founder and director of the Samaritan ALS clinic, co-director of the neurophysiology department at Banner Good Samaritan Medical Center, and a clinical assistant professor in neurology at the University of Arizona.
Ask the Experts

**Patient:** Is it true that I will be penalized if I don’t sign up for Medicare when I become eligible? I have a retirement plan, so I do not think I need Medicare Part B.

**IG Living:** We asked Leslie Vaughan, RPh, to answer your question.

**Leslie:** In short, yes. With one exception, you must sign up for Medicare during your initial period of eligibility or face penalties for every 12-month period you delay. There is a seven-month period to sign up for Medicare. That time period includes the three months before your birthday month during the year you turn 65, your birthday month, and three months after your birthday month. For instance, if your birthday is July 4, you are eligible to sign up for Medicare between April and October. If you sign up for coverage in the first three months (April through June), your Medicare start date will be the first day of your birthday month (July), unless your birthday is the first day of July, in which case coverage starts the first day of the previous month (June). If you enroll during your birthday month or in one of the three months after your birthday month (July through October), coverage will begin on the first day of the following month.

If you fail to sign up for Medicare during the seven-month period of your initial eligibility, you could be penalized 10 percent of your monthly premium for every year you fail to sign up. For Medicare Part A, you could be penalized for twice the number of years you fail to sign up. In other words, if you fail to sign up for two years, you will be penalized for four years. For Medicare Part B, you could be penalized 10 percent for every year you delay for the rest of your life. This means that if you fail to sign up for Medicare Part B for two years, your monthly premiums will be 20 percent higher for the remainder of the time you are covered by Medicare. Additionally, there may be penalties for those who do not elect Medicare Part D plans when first eligible.

However, there is an exception. If you or your spouse is employed and you are covered by an employment-sponsored group plan, you can delay signing up for Medicare until your coverage ends. Once your coverage ends, you will qualify for a special enrollment period, and as long as you present a certificate of credible coverage, you will not be penalized for delaying enrollment. It is important to note, though, that retirement plans and COBRA do not count as employment-sponsored coverage. If you have a retirement plan or COBRA plan, you still must enroll in Medicare when eligible or face penalties.

More information about this can be found at [www.medicare.gov/Publications/Pubs/pdf/11219.pdf](http://www.medicare.gov/Publications/Pubs/pdf/11219.pdf).

**Sajal:** What natural fruits are available in India to help me gain strength in my quadriceps muscles?

**IG Living:** Eating a healthy, well-balanced diet is especially important for people with a chronic illness. We asked **IG Living** contributor Jill Weisenberger, MS, RD, CDE, to address your question.

**Jill:** There are no foods that specifically target leg muscles. In fact, it’s not possible to gain strength from food alone. To build muscle, one must perform strength-training exercises, allow the muscles a break from strenuous exercise to rest and repair, and consume a balanced diet with adequate protein. Fruits are an important part of any diet. While they are low in protein, they are rich in nutrients and usually low in calories. A good post-workout snack could be a piece of fruit along with a good protein source such as dairy foods, eggs or meats. For example, try Greek yogurt mixed with fresh berries.

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

**JILL WEISENBERGER,** MS, RD, CDE, is a registered dietitian and has a website All That’s Nutrition at [www.allthatsnutrition.com](http://www.allthatsnutrition.com).

Have a question?  
**Email us at** [editor@IGLiving.com](mailto:editor@IGLiving.com).  
Your information will remain confidential unless permission is given.
Let’s Talk!

By Trudie Mitschang

At an age when most 10-year-olds are learning about integers, factors and U.S. history, Tyler Carlsen was learning about the workings of his immune system. Sickly since birth, Tyler had gone undiagnosed until the fifth grade, when doctors finally determined he was suffering from common variable immune deficiency (CVID), a disorder characterized by low levels of serum immunoglobulins (antibodies) and an increased susceptibility to infections. Tyler came to our attention when we saw a Facebook post he made looking for patients willing to be interviewed for a documentary he is making. Now, at only 17 years of age, Tyler is already defying the odds and making a difference in the lives of those diagnosed with chronic disease.

Trudie: You were diagnosed with CVID in the fifth grade. What was your life like prior to diagnosis?

Tyler: I remember feeling like I was having a normal childhood. But I also knew I was sick much more frequently than any other kids. I’m not sure if I was repeatedly misdiagnosed. I was young; my parents were taking me from one doctor to another, and they were not looking for a bigger reason as to why I was always sick. They would just treat the problem I presented with, and that was it. Looking back on my childhood, and having my mother tell me about my sicknesses, it makes me wonder how they could have missed this.

Trudie: Tell us how you cope as a teen living with chronic illness.

Tyler: I have an amazing support system of friends and family. I get lost in music and movies; I love distractions. I have a girlfriend now, and she is great at keeping me positive and happy. I do miss a lot of school, so I am on what’s called a 504 Plan. This plan allows me time to make up missed assignments, as well as get the support of tutors if I ever fall too far behind. This plan also makes it possible to keep all my doctor’s appointments [and] infusion dates and [take] sick time without getting penalized.

Trudie: What is the most challenging thing about living with CVID?

Tyler: I am beginning to understand I set my own limitations. I have been living with CVID for 17 years, and now I realize that it’s not just me. Many people have limitations; you have to learn how to live your life in spite of your condition.

Trudie: How has intravenous immune globulin (IVIG) helped?

Tyler: IVIG has allowed me to meet so many people from all over the world, especially through social media, and the support we lend one another is uplifting. The experiences I have had with these people have inspired me to make a documentary, which I hope will shed some light on CVID.

Trudie: Tell us a little about your documentary.

Tyler: My project’s name is called Swimming Against the Tide: Living with CVID. I have found that even my closest friends and family don’t really understand what CVID is and how deeply it affects me. I want to get the word out there for all to see and hear, and to be the voice of those who don’t know how to explain it. I find that most CVID patients have the same advice offered to them over and over again, as well as always being told that we don’t look sick. This documentary is not meant to make others feel sorry for CVID patients; it is about helping people understand.
Trudie: I read that you got sponsorship from Children’s Hospital Boston and Grandview Productions. As a 17-year-old, how did you accomplish that?

Tyler: My immunologist at Children’s Hospital Boston, Dr. Luigi Notarangelo, was instrumental in his role working with the Immune Deficiency Foundation (IDF). He pointed me in the right direction to get started. Grandview Productions is a film company that I have worked with on two films already; they are an inspiring group of young filmmakers from Cape Cod. They have moved out to California, but come back here to film. I approached them with a written proposal, and they were very supportive. They are allowing me the use of their equipment and, more importantly, their experience.

Trudie: What has chronic illness taught you?

Tyler: I have learned that I am stronger and braver than I thought I was. I’m also learning where I draw my strength and comfort from.

Trudie: What motivates or inspires you?

Tyler: My family motivates me, as well as inspires me. People I have met who have overcome insurmountable odds and are living their lives to the fullest inspire me.

Trudie: What do you do for fun?

Tyler: I have fun writing and making movies. I enjoy being with my family, seeing movies and being with my friends and my girlfriend. I am lucky to have such good people around me all the time — this I know.

Trudie Mitschang is a staff writer for IG Living magazine.

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

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“MOM, WHAT’S FOR dinner?” my kids demand! Three ravenous, ferocious “feedlings” follow each other into the kitchen one after another, ripping the refrigerator apart on a hunt for anything that is both savory and will satisfy after-school starvation.

I watch in horror as celery and green apples fly through the air with the greatest of ease; the cream cheese somehow manages to land intact on the kitchen island.

“Cream cheese? Ewwie-icky!” the youngest whines. “I want peanut butter! Where is the peanut butter? Mom, what did you do with the peanut butter? I’m starving!”

Then the eldest: “I don’t want celery, I want cereal! Mom, do we have any Chocolate Frosted Sugar Bombs?” I step away from the pantry as the 14-year-old scales to the top shelf mumbling angrily under his breath: “I don’t know what Mom is thinking. I haven’t eaten anything green since Molly was born. I’m a teenager and I need all things sugar!” Shaking the box of Sugar Bombs, Calvin quickly realizes his brother, Caleb, has devoured the majority of the cereal. (Caleb has had an insatiable appetite since starting prednisone; the steroid seems to be helping his immune system fight an aggressive sinus and lung infection.) Calvin throws the empty box of Sugar Bombs on the floor and starts toward his brother, who is sucking down instant Ramen noodles, his bare feet perched on the family room coffee table. Our dogs, Jax and Woody, deploy from under the kitchen table — it’s the perfect spot to nab a mealtime treat or catch taco night tidbits. Heck, when Nana comes for dinner, Jax and Woody are hand-fed a banquet! — and begin sopping up Sugar Bombs dregs that blanket the hardwood floor.

I fumble through the pantry, desperate to fill Molly’s request, when I realize that Calvin is on his way to strangle his brother with a Ramen noodle. I’m not worried about bloodshed because Caleb is more of a physical threat. Since starting steroids, our seventh-grader has grown a beard and is speaking like Barry White.

“CREAMY peanut butter! Hallelujah!” I announce, wiping a drop of sweat from my brow. I turn to give Molly the jar, and I find her licking the floor with Jax and Woody.

Ah, well. I smile. Another after-school snack session is on the books. OK, I might be exaggerating a little bit about these sessions, but I’m not totally off the mark. Raising three kids with an immune deficiency, two of whom receive intravenous immune globulin (IVI G), means a little insanity is a common side effect. In fact, it’s when things are “normal” that I get a little freaked out!

So, when I really think about it, there is nothing normal about primary immune disease! In all seriousness, the illnesses we fight are as rare as are the treatments that heal them, specifically IG (or gamma globulin).

We were first introduced to IG when Caleb was on the antibiotic roller coaster. Every few rounds, he’d get off just to jump right back on again. The infections were never-ending, and my husband, Mark, and I began to think Caleb might not have the strength to fight another day. Enter IG, and the surgeries, PICC-lines and constant hospitalizations came to a standstill. Caleb got his life back, and for the first time in our lives as parents, we got to watch him grow up instead of watching his life slip away.

Sound familiar? We are kin to a
worldwide family that understands the lifesaving power of IG, and my gut tells me science has only scratched the surface of its healing omnipotence. For example, our kids’ immunologist shared with us that IG has been used to help a young patient recover from the effects of an intestinal parasite. (Let’s not go “there” on how it was administered. I’m sure you can figure that out!) And it wasn’t until a recent Haggard family tradition that we realized all that IG can do.

It’s Tuesday night and nobody asks me “What’s for dinner?” Cumin, season salt, chili powder and garlic, along with a host of other secret ingredients married with ground sirloin danced in my five-generation-old taco pan. I knew this particular “Tuesday Taco Night” was special because Caleb finally got the green light from his doctor to consume crunchier cuisine. He had been looking forward to taco night for two weeks after recovering from a sinus reconstruction and tonsillectomy. Nonetheless, I was quite clear Dr. Fanning didn’t want spicy taco meat in his reconstructed nasal passages.

“Do something!” Caleb demanded. His stress quickly changed to panic as he rubbed his cheeks, being careful not to undo Dr. Fanning’s handiwork (or, frankly, Caleb’s million-dollar maxillaries!). “Get it out! Get it out!” Caleb cried.

We couldn’t use regular saline because Dr. Fanning didn’t want us to irritate the airways (not that taco meat wasn’t irritating enough!). Then it hit me! If immunologists are using IG for one end of the human body, then why couldn’t I use it for the other end? “Mark!” I yelled down the hallway. “Grab Molly’s sub-q stuff, and we’ll use that instead!”

A few tense moments later, I held a syringe in my hands and very, very slowly I pushed precious “liquid gold” into Caleb’s right nostril.

Not … too … fast, I coached myself. Easy does it.

Caleb’s face twitched and his eyes started to water. “I think I’m gonna blow!” Caleb cringed, trying to speak with his teeth clenched. “Here it comes!”

It took only about seven seconds of steady-as-she-goes pressure depositing our dear beloved IG into Caleb’s besieged beak. I turned my attention toward Mark and cried (read in slow motion): “Hand … me… the… towel!”

Now I’ve seen a lot of gross stuff over the years. A couple of gallons of stomach flu had nothing on Caleb’s present moment post-sneeze discharge. I stretched out the towel and inspected its contents. I took special care to make sure Caleb’s brains didn’t come along with the rest of his taco dinner.

“Totally cool!” Calvin squealed with delight.

“Eww! Is that blood, Daddy?” Molly piped in.

“No, darling, that’s taco-flavored gamma globulin,” Mark concluded.

You’ll be happy to hear Dr. Fanning’s inspection of Caleb’s taco meat infestation came out OK. And, he said flushing Caleb with IG was a smart move, as it probably kept another infection from rearing its ugly head.

“Yeah, the lab reported normal flora, but…” Dr. Fanning paused.

“There is some bad news.”

“Oh?” I panicked.

“Yes,” replied Dr. Fanning. “The lab wants you to know your secret taco meat recipe isn’t a secret anymore.”

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.

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Sometimes I feel like a human onion (a funny paradox considering I am highly allergic to onions). The more I keep peeling back my auto-immune layers, the more I keep finding. It appears autoimmune diseases like companionship. I was hoping my multifocal motor neuropathy (MMN) enjoyed leading a solitary life. But recently, I also was diagnosed with lupus, Hashimoto’s disease and Sjogren’s syndrome; it seems my autoimmune system is quite the social butterfly.

I had gotten used to MMN treatments, I learned all I could about the world of MMN, and I mapped out my entire future with only MMN. Now, these three new autoimmune diseases have joined the party, and I have to learn their names and what they do, and I don’t even get to decide if I want to get to know them. They are here to stay like bad roommates you can never ask to move out.

When the rheumatologist told me about each of my new conditions, I imagined parts of me starting to disintegrate and float away into the air. For the first time, I felt like I was losing parts of myself. How much more can my body take? If I’m not safe in my own body, how can I ever feel safe again?

I am a voracious reader; I have been since I was a child. I believe in the power of words. Ideas can come from an issue of Vogue magazine or a Pulitzer Prize-winning novel and everything in between. A character, a quote, a poem or a play can touch something so deep in my core, I am never the same and am thus transformed.

Feeling like a shell of my former self after all the news about my multiple diagnoses, I high-tailed it to one of my favorite refuges for comfort: the local bookstore. After aimlessly strolling around getting lost among the pretty book covers, I somehow ended up with a book in my hand by playwright and author Eve Ensler titled Insecure at Last. The title definitely seemed to fit how I felt about my life at the moment. I was skimming through the book when I saw it — a hidden treasure of words, buried in this book, discovered on a random page that would change how I felt about my life: “Consider what would happen if security were not the point of our existence. That we find freedom, aliveness, and power not from what contains, locates, or protects us but from what dissolves, reveals, and expands us.”

The power of words once again enlightened and transformed me. I stood a little straighter, and there was even the hint of a smile on my face. It was time to let go and stop trying to control everything happening to me, because I can’t. I am a rheumatologic and neurologic mystery that is unfolding. The same way I am enraptured by an Agatha Christie novel, I should be as engaged and enthralled with my own life. I don’t know what’s going to happen next, but who says it has to be bad?

I know I’m not alone on this crazy autoimmune journey or even the more unpredictable and bigger journey called life. I might lose my luggage along the way, have my reservations canceled and not be able to speak the language, but I’m daring to keep going. My eyes have been opened to a whole new world of learning, discovery and imagination. Time to find my pith helmet; terra incognita awaits, and I can’t wait to find out what hidden treasures I’ll find.

STACY OLIVER was diagnosed in 2008 with multifocal motor neuropathy (MMN). She is the assistant director of the Center for the Writing Arts at Northwestern University, a jewelry and accessory artist (www.dancingstonesjewelry.com), and she is working on her secret super hero identity as Neuropathy Girl.
Parenting:

Packed with Essential Vitamins and Minerals

While vitamins, minerals and supplements are important for boosting children’s immune systems, the best sources of these come from a balanced diet.

By Mark T. Haggard

IF YOU’RE OLDER than 30-something, you might remember those commercials — crammed between “School House Rock” and “H.R. Pufnstuf” — about the latest sugary-sweet cereal from Kellogg’s “packed with essential vitamins and minerals.” How much good do you think those vitamins and minerals were surrounded by 10,000 calories? When we were 10 years old, it didn’t matter; it tasted good, and there was a toy submarine in the bottom of the box. But, for those of us who now have kids — especially those of us whose children have an immune deficiency disease — it matters greatly.

A few years ago, I asked a doctor about the effectiveness of vitamins and supplements, and I was told the tests are inconclusive. Kurt Harlan, PharmD, at St. Joseph’s Home Health Network in Orange, Calif., says that “a physician would not put their entire faith in vitamins and supplements for the treatment of immune deficiencies” because there is nothing “curative” about them; they are only a supplement to people’s ability to fight off infection. Nevertheless, Dr. Harlan adds that “western” doctors are increasingly understanding that there is a role for “eastern” medicine practices, and among those practices are vitamins and minerals.

Vitamins

Whether taken as a tablet or the old-fashioned way — in the form of numerous colorful fruits and vegetables — vitamins have an important effect on the body’s immune system.

Vitamin A is essential to the immune system’s defensive barrier, helping the body’s mucous membranes — nostrils, sinuses, eyes, inner mouth, throat and gut — stay moist and evenly dispersed, and trapping, blocking and eliminating pathogens. Without vitamin A, mucosal tissue cannot properly prevent entry of viruses and bacteria, allowing infections to enter the body’s system. Vitamin A also contributes to the manufacture of enzymes, which search and destroy invaders that
Whether taken as a tablet or the old-fashioned way — in the form of numerous colorful fruits and vegetables — vitamins have an important effect on the body’s immune system.

A vitamin B complex is important for cellular metabolism and energy production. Folic acid, B5, B6 and B12 all contribute to the offensive capabilities of the immune system. Vitamin B6, also called pyridoxine, is necessary for the maturation of the body’s B cells and T cells, which circulate throughout the blood stream in an immature state until they contact a pathogen; they then mature into specialized cells that kill the pathogen. Vitamin B6 also is partially responsible for increasing white blood cells, including “killer T cells” in the bloodstream.

Vitamin C stimulates production and function of white blood cells, particularly lymphocytes (B cells and T cells), neutrophils and phagocytes. Neutrophils, the first responders of the immune system, seek and destroy foreign antibodies. Phagocytes ingest bacteria, dying cells and harmful particles during “phagocytosis.” Vitamin C also increases serum levels in circulating antibodies and contributes to the production of interferon, a protein that coats cell surfaces and prevents intrusion by viruses. Studies have shown that low levels of vitamin C reduce some immune functions and increase risk of infection. Like vitamin A, vitamin C is a powerful antioxidant that protects the body from damage by free radicals. Ample levels can be acquired by eating plenty of citrus fruits, berries, green, yellow and red peppers, sweet potatoes, cauliflower, kale, mangoes, melons, tomatoes, broccoli and brussels sprouts.

Vitamin D is needed to activate “killer T cells” so that they can detect and kill pathogens. A 2010 National Institutes of Health (NIH) study found that vitamin D plays a crucial role in the regulation of B and T cells. Research by Marina Rode von Essen, PhD, shows that killer T cells rely on vitamin D to be activated from their dormant state. T lymphocytes and macrophages (a type of phagocyte) have high concentrations of vitamin D receptors in their cell membranes. Vitamin D can be obtained from fish, eggs, fortified milk and cod liver oil, but it is most commonly synthesized by the body in the presence of sunlight.

Vitamin E is an effective modulator of the immune system. A 1979 study by the NIH suggested that regular vitamin E stimulation increases the activity of T lymphocytes. Vitamin E also produces interleukin-2, a protein that destroys bacteria, viruses and cancer cells. An efficient antioxidant as well, vitamin E is found in dark green leafy vegetables, spinach, asparagus, avocados, broccoli, carrots, red peppers, pumpkins, nuts, seeds (particularly sunflower seeds), eggs, milk, wheat germ and vegetable oils.

Supplements

Zinc determines how many lymphocytes are present in the bloodstream, and it is the key to activating lymphoid cells and enzymes used in immune response. A study by the Technical University of Aachen in Germany published in the Journal of Nutrition (May 2003) states that zinc is essential for all highly proliferating cells in the body, and its depletion in the body leads to decreased activity of immune system cells. Zinc is not stored by the body, so it must be ingested daily. It is found in dairy products, nuts (particularly cashews and almonds), seafood, red meats, oysters, beans and whole grains.

Selenium is a trace mineral that stimulates the immune system in response to foreign antigens. Research by scientists at New York University published in Biological Trace Element Research (February 2000) found that taking selenium daily resulted in significantly higher cell-mediated immune responsiveness, particularly that of cytotoxic lymphocytes that destroy tumor cells. It also is a powerful antioxidant. Good sources of selenium are tuna,
red snapper, lobster, shrimp, beef, grains, Brazil nuts and poultry.

Two other important trace minerals for immune health are magnesium and copper. A study published in the European Journal of Clinical Nutrition (October 2003) shows that magnesium affects the number of immune cells and how they function. Another study published in the British Journal of Nutrition (May 2002) shows that copper stimulates the function of immune cells, including neutrophils, interleukin and lymphocytes.

Two herbs also have an important effect on our immune system. Garlic has proven to be a powerful antioxidant and immune booster, increasing white blood cells, natural killer cells and antibody production in the body, as well as reducing carcinogens. Ginseng, according to a study in The Open Nutrition Journal, improves the activity of neutrophils, which are the first responders in the immune system.

Other supplements that are important include: Omega-3s are fatty acids necessary for a healthy immune system, boosting the production of phagocytes, protecting the body from damage from infection, and reducing inflammation. Good sources of omega-3s include flax seeds and fatty cold-water fish such as salmon, tuna, trout, sardines and mackerel. Carotenoids, which improve immune function in a number of ways, include beta carotene, lutein and lycopene, and are found in red, yellow and orange fruits and vegetables. Beta carotene increases the number of infection-fighting cells, natural killer cells and helper T cells; it also kills excess free radicals and protects against cancer by stimulating the immune cells that kill cancer cells. Bioflavonoids aid the immune system by protecting the cells of the body against environmental pollutants. These natural compounds are found in grapes, berries, citrus, dark chocolate and tea.

Supplements and Children

The optimal way for children to get the best combination of vitamins, minerals and supplements is with a healthy and varied diet. Those parents who believe their children need to take chewables or Gummies need to realize the potential danger of vitamin overdose. According to the University of Texas Southwestern Medical Center, vitamins are the leading cause of poisoning deaths in children under 6 years old.

Supplements and Children

The optimal way for children to get the best combination of vitamins, minerals and supplements is with a healthy and varied diet.

Eating an excess of any vitamin can cause nausea, vomiting, diarrhea, constipation, lethargy, headache, blurred vision or abdominal pain. It also may cause skin irritations such as yellowing, peeling, cracking or rash. Ingesting too many multivitamins will cause a sudden increase in iron, which may cause liver damage and scarring of the stomach or intestines. Other symptoms of vitamin overdose are increased urination, cloudy urine, irregular heartbeat, muscle or bone pain, fainting, convulsions or confusion. If a vitamin overdose is suspected, poison control or 911 should be immediately called.

A Balanced Diet

While it is true that no vitamin, mineral or supplement will recreate that part of the immune system in which some children are deficient, a nutritious eating plan can boost other parts of their immune system and help to bear a heavier load. Studies show that vitamins and minerals, along with adequate sleep, exercise and a balanced diet including plenty of fruits and vegetables, will help

Manufacturers make them taste and look like candy. So, children need to be educated about what they are, and parents need to keep them out of children’s reach.

Eating an excess of any vitamin can cause nausea, vomiting, diarrhea, constipation, lethargy, headache, blurred vision or abdominal pain. It also may cause skin irritations such as yellowing, peeling, cracking or rash. Ingesting too many multivitamins will cause a sudden increase in iron, which may cause liver damage and scarring of the stomach or intestines. Other symptoms of vitamin overdose are increased urination, cloudy urine, children’s immune issues. So to promote the best immune defense, don’t go running for the Chocolate Frosted Sugar Bombs on Saturday morning before “H.R. Pufnstuf.”

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.

Sources

For more on the connection between vitamins and supplements and immune deficiency, see:
www.altMD.com
www.livestrong.com
DECREASED MUSCLE STRENGTH and impaired coordination often accompany chronic illness due to either the disease itself or the infections that lead to long periods of convalescence. And while there may not be a cure for most chronic illnesses, regular exercise has been proved to decrease pain, increase stamina and improve overall health.

For many patients, however, the costs, stress and time associated with long-term physical therapy and/or a gym membership make regular exercise a challenge. Fortunately, there are some low-cost alternatives that can help to improve fitness in the comfort of one’s own home. Of course, patients should never start any new exercise routine without first consulting their doctor and/or physical therapist.

Resistance Exercise

Resistance exercises are designed to build strength, muscle tone and endurance. Using an elastic band is a simple and inexpensive way to complete resistance exercises using multiple extremities. These bands are color-coded for different levels of resistance. Physical therapists and occupational therapists commonly use resistance bands for patient rehabilitation because they are easily adaptable to all levels of fitness and they can be used to work upper and lower extremities.

Another good form of resistance exercise is pedaling. For those who do not have the space or budget for a full-sized stationary bike, a good alternative is a pedal exerciser, which is a stationary bike that can be used with a chair instead of one that comes with the seat attached. The advantage of this type of equipment, beyond monetary, is that it also can be placed on a tabletop or counter and used to strengthen the upper extremities. When purchasing a pedal exerciser, it is important to find a sturdy one that can withstand a good amount of pressure. A lot of inexpensive pedal exercisers end up in thrift stores because they are poorly made.

Core Strength

In addition to strengthening upper- and lower-extremity muscles, it is important for patients to focus on balance, posture and coordination. Strengthening core muscles — those used to stabilize the spine and pelvis and keep them strong — improves balance and coordination, which in turn decreases pain and the risk of accidental falls.

A multipurpose piece of equipment that will help strengthen core muscles at all fitness levels is a therapy ball. Also known as an exercise ball, fitness ball or yoga ball, each comes in a variety of sizes and colors. Before starting any kind of program with a therapy ball, patients should meet with a physical therapist or certified trainer to make sure they choose a ball correctly sized for them. A ball that is too large or too small could lead to incorrect posture, loss of balance and muscle strain.

Patients first starting out using a therapy ball can do something as simple as sitting on the ball while watching television or working on a computer. Once they are comfortable with this, exercises can be progressed as tolerated with the guidance of an experienced trainer or physical therapist to ensure proper techniques and posture are maintained.

Fitness Is Possible at Any Level

There is always something that can be done to improve the fitness level of patients, no matter what their current level may be. And, while they should first consult a professional to make sure exercises are not doing more harm than good, the equipment they use need not be fancy or expensive to improve their quality of life.
Thera-Band
Thera-Band resistive exercise bands come in latex and latex-free formulas in 50-foot rolls. They have no scent, no powder and come in the colors of the Thera-Band color progression.

Bodylastics
Bodylastics resistance bands exercise system provides variable resistance for all fitness levels and includes four different elastic bands that can be combined to create 15 levels of resistance. The bands are adjustable to give users more control over their workouts.
(800) 500-1979; www.resistancebands.com

Greater Medical
CycleChiser is designed for people with mobility issues, allowing them to maintain a fitness exercise regimen even while seated. The digital LCD counter tabulates total workout time and calories burned. The adjustable resistance settings can be used for upper- and lower-body workouts. It is made with high-quality construction that includes nonskid pads, and it comes with a one-year warranty.
(888) 657-8436; www.greatermedical.com/med064.html

YogaDirect
Yoga Balance/Fitness Balls are designed to strengthen and firm abs, the back and buttocks. They are made from durable vinyl, and they are designed to support up to 600 pounds of pressure. The balls are easily inflated with a foot or hand pump, and the large 75cm Fitness Balls are best for people up to 6 feet 5 inches tall. Each ball is individually wrapped.
(800) 331-8233; www.yogadirect.com/Fitness-Balls--75cm_p_37.html

Ball Dynamics
The FitBALL exercise ball is designed for flexibility, strength and aerobic training. It comes in four sizes, and has a weight-bearing capacity of 2,250 pounds. It is made of a unique latex-free material designed to deflate slowly if punctured.
(800) 752-2255; www.balldynamics.com

3D Innovations
The MagneTrainer ER Mini Exercise Bike provides smooth, quiet pedal motion at variable resistance levels and speeds. It features a wide, solid base that keeps it firmly on the floor or table, and pedals come standard with ball-bearing and long, heavy-duty Velcro straps.
www.magnettrainer.com

Directory of Exercise Equipment

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

**General Resources**

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

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

**Disease-State Resources**

**Ataxia Telangiectasia (A-T)**

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

**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)**

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

**Evans Syndrome**

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

**Guillain-Barré Syndrome (GBS)**

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

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

**Idiopathic Thrombocytopenic Purpura (ITP)**

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

**Kawasaki Disease**

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

**IG Manufacturer Websites**

- Baxter: www.baxter.com
- CSL Behring: www.cslehring.com
- Grifols: www.grifolsusa.com
- Kedrion: www.kedrionusa.com
- Octapharma: www.octapharma.com

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

Websites
- United Mitochondrial Disease Foundation: www.umdf.org
- Mitochondria: www.mitoaction.org

Multifocal Motor Neuropathy (MMN)

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

Multiple Sclerosis (MS)

Websites
- All About Multiple Sclerosis: www.mult-sclerosis.org/index.html
- Multiple Sclerosis Association of America: www.msaam.com
- National Multiple Sclerosis Society: www.nationalmssociety.org

Online Peer Support
- Friends with MS: www.FriendsWithMS.com
- MSWorld’s Chat and Message Board: www.msworld.org

Myasthenia Gravis (MG)

Websites and Chat Rooms
- Myasthenia Gravis Foundation of America (MGFA): www.myasthenia.org

Online Peer Support
- Genetic Alliance: www.geneticalliance.org

Myositis

Websites
- The Myositis Association, www.myositis.org, is to find a cure for inflammatory and other related myopathies, while serving those affected by these diseases. (703) 299-4850
- The Cure JM Foundation, www.curejm.com, is dedicated to early and precise diagnosis, meaningful treatments and, ultimately, cures for primary immunodeficiency. (760) 487-1079

Online Peer Support
- Myositis Association Community Forum: www.myositis.org
- Myositis Support Group: www.myositissupportgroup.org

Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)

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

Pemphigus and Pemphigoid

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

Peripheral Neuropathy (PN)

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

Primary Immune Deficiency Disease (PIDD)

Websites
- The Immune Deficiency Foundation (IDF), www.primaryimmune.org, is 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.ipop.org
- Michigan Immunodeficiency Foundation: www.facebook.com/groups/10804806284350
Sources

- National Institute of Child Health and Human Development (NICHD) (Click on “Health Information” then “A to Z health & human development topics” and select “P” for “Primary Immunodeficiency”): www.nichd.nih.gov
- New England Primary Immunodeficiency Network: www.nepin.org
- Team Hope (for families and patients in New England): www.teamhope.info

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

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**Scleroderma**

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

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

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**Stiff-Person Syndrome (SPS)**

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

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**Other Resources**

**Education and Disability Resources**
- Provides protection for people with disabilities from certain types of discrimination, and requires employers to provide some accommodations of the disability.
- 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 Disability Rights Network: www.ndrn.org. This website offers a search tool to find resources in your state to assist with school rights and advocacy.
- Social Security: www.ssa.gov/disability
- U.S. Department of Education Website: www.ed.gov. This federal government website offers a parents section titled “My Child’s Special Needs.”
- Spells out your rights under Section 504 of the Rehabilitation Act.

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

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

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

**Pump and Infusion Sets Websites**
- EMEC 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.
Immune Globulin Subcutaneous (Human), 20% Liquid

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

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

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

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

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

5.3 Reactions Reported to Occur With IGIV Treatment
The following reactions have been reported to occur with IGIV treatment and may occur with IGSC treatment.

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

Periodic monitoring of renal function and urine output is particularly important in patients judged to have a potential increased risk of developing acute renal failure.9 If renal function deteriorates, consider discontinuing Hizentra. For patients judged to be at risk of developing renal dysfunction because of pre-existing renal insufficiency or predisposition to acute renal failure (such as those with diabetes mellitus or hypovolemia, those who are overweight or use concomitant nephrotoxic medicinal products, or those who are over 65 years of age), administer Hizentra at the minimum rate practicable.

Thrombotic Events
Thrombotic events may occur with use of human immune globulin products9. 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 triacylglycerols (triglycerides), or monoclonal gammapathies. For patients judged to be at risk of developing thrombotic events, administer Hizentra at the minimum rate practicable.

Hemolysis
Hizentra can contain blood group antibodies that may act as hemolysins and induce in vivo coating of red blood cells (RBCs) with immunoglobulin, causing a positive direct antiglobulin (Coombs’) test result and hemolysis. 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.

5.4 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.

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

6 ADVERSE REACTIONS
The most common adverse reactions (ARs), observed in ≥5% of study subjects receiving Hizentra, were local reactions (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 2,264 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>49 (100)</td>
<td>1340 (0.592)</td>
</tr>
<tr>
<td>Other AEs:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>13 (26.5)</td>
<td>40 (0.18)</td>
</tr>
<tr>
<td>Cough</td>
<td>8 (16.3)</td>
<td>9 (0.04)</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>7 (14.3)</td>
<td>8 (0.04)</td>
</tr>
<tr>
<td>Fatigue</td>
<td>6 (12.2)</td>
<td>6 (0.03)</td>
</tr>
<tr>
<td>Back pain</td>
<td>5 (10.2)</td>
<td>11 (0.05)</td>
</tr>
<tr>
<td>Nausea</td>
<td>5 (10.2)</td>
<td>5 (0.02)</td>
</tr>
<tr>
<td>Abdominal pain, upper</td>
<td>5 (10.2)</td>
<td>5 (0.02)</td>
</tr>
<tr>
<td>Rash</td>
<td>5 (10.2)</td>
<td>7 (0.03)</td>
</tr>
<tr>
<td>Pain in extremity</td>
<td>4 (8.2)</td>
<td>7 (0.03)</td>
</tr>
<tr>
<td>Migraine</td>
<td>4 (8.2)</td>
<td>5 (0.02)</td>
</tr>
<tr>
<td>Pain</td>
<td>4 (8.2)</td>
<td>5 (0.02)</td>
</tr>
<tr>
<td>Epistaxis</td>
<td>4 (8.2)</td>
<td>6 (0.03)</td>
</tr>
<tr>
<td>Pharyngolaryngeal pain</td>
<td>4 (8.2)</td>
<td>6 (0.03)</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>4 (8.2)</td>
<td>5 (0.02)</td>
</tr>
</tbody>
</table>

* Excluding infections.
† Rate of AEs per infusion.
†† Includes injection-site reactions as well as bruising, scabbing, pain, irritation, cysts, eczema, and nodules at the injection site.

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 ARs (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>
</tr>
</thead>
<tbody>
<tr>
<td>Local reactions†</td>
<td>49 (100)</td>
<td>1338 (0.591)</td>
</tr>
<tr>
<td>Other AEs:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>12 (24.5)</td>
<td>36 (0.016)</td>
</tr>
<tr>
<td>Vomiting</td>
<td>3 (6.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Pain</td>
<td>3 (6.1)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Fatigue</td>
<td>3 (6.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Confusion</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Back pain</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Migraine</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>2 (4.1)</td>
<td>2 (&lt;0.001)</td>
</tr>
<tr>
<td>Abdominal pain, upper</td>
<td>2 (4.1)</td>
<td>2 (&lt;0.001)</td>
</tr>
<tr>
<td>Nausea</td>
<td>2 (4.1)</td>
<td>2 (&lt;0.001)</td>
</tr>
<tr>
<td>Rash</td>
<td>2 (4.1)</td>
<td>2 (&lt;0.001)</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>2 (4.1)</td>
<td>2 (&lt;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.

Table 4 summarizes injection-site reactions based on investigator assessments 15 to 45 minutes after the end of the 683 infusions administered during regularly scheduled visits (every 4 weeks).

Table 4: Investigator Assessments* of Injection-Site Reactions by Infusion

<table>
<thead>
<tr>
<th>Injection-Site Reaction</th>
<th>Number† (Rate‡) of Reactions (n=683 Infusions§)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Edema/induration</td>
<td>467 (0.68)</td>
</tr>
<tr>
<td>Erythema</td>
<td>346 (0.50)</td>
</tr>
<tr>
<td>Local heat</td>
<td>108 (0.16)</td>
</tr>
<tr>
<td>Local pain</td>
<td>88 (0.13)</td>
</tr>
<tr>
<td>Itching</td>
<td>64 (0.09)</td>
</tr>
</tbody>
</table>

* 15 to 45 minutes after the end of infusions administered at regularly scheduled visits (every 4 weeks).
† For multiple injection sites, every site was judged, but only the site with the strongest reaction was recorded.
‡ Rate of injection-site reactions per infusion.
§ Number of infusions administered during regularly scheduled visits.
Most local reactions were either mild (93.4%) or moderate (6.3%) in intensity.

6.2 Postmarketing Experience

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

The following adverse reactions have been identified and reported during the postmarketing use of IGIV products:

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

To report SUSPECTED ADVERSE REACTIONS, contact CSL Behring Pharmacovigilance at 1-866-915-6938 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

7 DRUG INTERACTIONS

7.1 Live Virus Vaccines

The passive transfer of antibodies with immunoglobulin administration may interfere with the response to live virus vaccines such as measles, mumps, rubella, and varicella (see Patient Counseling Information [17]).

7.2 Serological Testing

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

8 USE IN SPECIFIC POPULATIONS

8.1 Pregnancy

Pregnancy Category C. Animal reproduction studies have not been conducted with Hizentra. It is not known whether Hizentra can cause fetal harm when administered to a pregnant woman or can affect reproduction capacity. Hizentra should be given to pregnant women only if clearly needed.

8.3 Nursing Mothers

Hizentra has not been evaluated in nursing mothers.

8.4 Pediatric Use

The safety and effectiveness of Hizentra have been established in the pediatric age groups 2 to 16, as supported by evidence from adequate and well-controlled studies. Hizentra was evaluated in 10 pediatric subjects with PI (3 children and 7 adolescents) in a study conducted in the US (see Clinical Studies [14]) and in 23 pediatric subjects with PI (18 children and 5 adolescents) in Europe. There were no differences in the safety and efficacy profiles as compared with adult subjects. No pediatric-specific dose requirements were necessary to achieve the desired serum IgG levels.

Safety and effectiveness of Hizentra in pediatric patients below the age of 2 have not been established.

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.
If you live with primary immunodeficiency disease (PIDD)...

Make the leap to Hizentra

The Sub-Q Ig therapy that fits your life

Hizentra is a subcutaneous immune globulin (Sub-Q Ig) therapy that was deliberately designed to give you freedom and flexibility with your Ig treatment.

- The 20% concentration delivers an Ig dose in half the volume of 10% solutions
- Convenient room temperature storage for up to 30 months
- Always ready for immediate use

*Based on an equivalent dose in grams.

To learn about the benefits of Hizentra, visit www.LearnAboutHizentra.com

Ask your doctor about Hizentra today.

Important Safety Information

Hizentra is indicated for the treatment of various forms of 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 Hizentra should not be used.

Hizentra is to be infused under your skin only; do not inject into a blood vessel.

Hypersensitivity reactions may occur with Hizentra. If your physician suspects you are having a negative reaction or are going into shock, treatment will be discontinued. Because Hizentra contains proline, you cannot be treated with Hizentra if you have hyperprolinemia (a high level of proline in your blood).

Hizentra is 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.

The most common drug-related adverse reactions (seen in 5% or more of subjects in the clinical trial) were local reactions (swelling, redness, heat, pain, and itching at the injection site), headache, vomiting, pain, and fatigue.

Your physician will monitor for potentially serious reactions associated with intravenous immunoglobulin treatment that might also occur with Hizentra, including renal dysfunction/failure, osmotic nephropathy, thrombotic events, aseptic meningitis syndrome (AMS), hemolysis, and transfusion-related acute lung injury (TRALI).

Ig administration may impair the effect of virus vaccines, such as measles, mumps and rubella. Before getting any vaccination, inform your doctor that you are using Hizentra.

Please see brief summary of full prescribing information for Hizentra 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.
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