Patient-Doctor Dialogue
Bridging the Communication Gap

Caregivers: How and Why to Donate Plasma

Rituximab: The Newest Autoimmune Disease Therapy

Understanding and Treating MS
Vaccine Guidelines for Immunodeficient Patients
GAMUNEX®-C
Immune Globulin Injection (Human) 10% Caprylate/Chromatography Purified

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

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

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

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

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

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

WARNINGS AND PRECAUTIONS
- IgA deficient patients with antibodies against IgA are at greater risk of developing severe hypersensitivity and anaphylactic reactions. Have epinephrine available immediately to treat any acute severe hypersensitivity reactions.
- Monitor renal function, including blood urea nitrogen, serum creatinine, and urine output in patients at risk of developing acute renal failure.
- GAMUNEX-C is not approved for subcutaneous use in ITP patients. Due to a potential risk of hematoma formation, do not administer GAMUNEX-C subcutaneously in patients with ITP.
- Hyperproteinememia, 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.
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 polynuropathy; PI=Primary immunodeficiency; ITP=Idiopathic thrombocytopenic purpura.


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

Please see adjacent page for brief summary of GAMUNEX-C full Prescribing Information.
About IG Living

IG Living is the only magazine dedicated to bringing comprehensive healthcare information, immune globulin information, community and reimbursement news, and resources for successful living directly to immune globulin consumers and their healthcare providers.

IG Living, ISSN 1949-4548, published bimonthly, is a community service provided by FFF Enterprises, 41093 County Center Drive, Temecula, CA 92591, (800) 843-7477 x1362, fax (951) 699-9655.

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SHARON BURTON-YOUNG, RN  
*Founder and CEO, Infusion Care of Delaware*

**Plasma: How and Why You Should Donate**  
“Caregivers are our secret weapons, and plasma donation can be another part of their arsenal.”

AMY EHLERS, BS, PHARMD, BCPS  
*Director of Pharmacy, NuFACTOR Specialty Pharmacy*

**Rituximab: An Autoimmune Disease Therapy**  
“As more autoimmune diseases are diagnosed, the future use of rituximab will be expanded.”

KONNER LIVELY  
*IG Teen Patient*

**Teen Talk: There Is Life After Loneliness**  
“If you are feeling like I did, know that you don’t have to feel alone; there are others out there who understand what you are going through.”

LIN STEARNS  
*Writer and CVID Patient*

**IG Chronicles: Just Us Peas in a Pod**  
“It may feel like CVID is just us peas in a pod, but we can promote a better quality of life through attitude, knowledge and simply sticking together!”

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**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!  
*IG Living’s* Facebook page has hundreds of fans who respond to our questions that are posted each Monday through Friday. Together, these fans share their life stories and thoughts. What’s more, they are making a connection with one another that otherwise wouldn’t be possible.  
So, be a part of it now at [www.igliving.com/blogengine](http://www.igliving.com/blogengine) and [www.facebook.com/IGLivingMagazine](http://www.facebook.com/IGLivingMagazine).

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**Connect with Other IG Living Readers through Monthly Teleconferences!**

IGL’s Readers Group Teleconferences 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 teleconferences.
- *IG Living* will send you invitations to let you know when the monthly, hosted, toll-free teleconferences will be held, as well as what topic relevant to the IG community will be discussed.
- The moderated, hour-long calls will be filled on a first-come, first-served basis and will be limited.

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

Sign up for the Teleconferences now by emailing akazemi@IGLiving.com or calling (800) 843-7477, ext. 1366.
The Autoimmune Epidemic

America is experiencing an epidemic: Growing numbers of autoimmune disorders are being diagnosed at unprecedented rates. Based on a Mayo Clinic study conducted in 2011, it is estimated that one in 12 women and one in 20 men in the U.S. will develop some sort of autoimmune disease in their lifetime. The most common of the more than 150 autoimmune disorders are rheumatoid arthritis, systemic lupus erythematosus, psoriatic arthritis, polymyalgia rheumatica, giant cell arteritis, ankylosing spondylitis and Sjogren’s syndrome. Why this is happening is unclear, but it is believed that both genetic and environmental factors play a role.

Immune-deficient patients seem to be especially prone to autoimmune disorders. In fact, defects within certain components of the immune system carry a high risk for the development of autoimmune disease. For instance, between 20 percent and 37 percent of patients with common variable immune deficiency, the most prevalent immune deficiency, also will develop one or more autoimmune disorders.

As the number of these disorders continues to rise, getting the correct diagnosis is becoming even more problematic, often compounded by a lack of clear communication between patients and their doctors. According to our article “Bridging the Patient-Doctor Communication Gap,” the No. 1 complaint of patients is that their doctors don’t listen, whereas the No. 1 complaint of practitioners is the “Google-stack”: the reams of information patients collect on the Internet to self-diagnose their symptoms. But, as explained in our article, it is possible to overcome these communication hurdles by understanding that communication is a two-way street, and good relationships between doctors and patients are built upon mutual trust and respect.

Fortunately, treatment with intravenous immune globulin (IVIG) may ameliorate many autoimmune disorders. This is true for multiple sclerosis (MS), a chronic, unpredictable disease that attacks the central nervous system. How MS manifests, is diagnosed and treated is discussed in “Understanding and Treating Multiple Sclerosis.” While not one of the most common among immune-deficient patients, it is a highly prevalent autoimmune disorder that is sometimes treated with IVIG, even though this therapy remains controversial.

One therapy that has shown particular promise for autoimmune disorders is rituximab, an antibody therapy that doesn’t suppress the immune system. Rheumatoid arthritis is the only autoimmune disease that is approved by the FDA to be treated with rituximab; however, hundreds of smaller studies have successfully treated other autoimmune diseases with rituximab, which is why it is often used when other first-line therapies fail. And, as we explore in “Rituximab: An Autoimmune Disease Therapy,” its use is predicted to expand as more autoimmune diseases are diagnosed.

I hope you find these articles useful, as well as the many others in this issue that are presented to inform and empower our community.

Ronale Tucker Rhodes, MS, Editor
“Port”: Not a Four-Letter Word

What was the author’s main point in writing the article “Port”: Not Just a Four-Letter Word that was published in the April-May issue of IG Living? I have a PowerPort, as strongly encouraged and ordered by my doctors. The overall tone of the article makes it sound as if ports are frowned upon and that physicians don’t really believe patients need them and laugh behind the port-implanted patients’ backs. I must say this offended me — especially as “port” was paralleled to bad four-letter words. If I didn’t have my port, I would not be able to have necessary biweekly lab draws and daily IV hydration therapy and potassium infusions that, in my medical situation, are life-sustaining. I also have myasthenia gravis, so I have received intravenous immune globulin infusions. I know the author was probably intending to be humorous, but I just thought I’d share a different angle from a patient who was ordered by both her doctors to have a port implanted. I have no peripheral venous access left for typical IVs to be inserted or for venipunctures to be performed. I’m respectfully submitting these comments; I’m not angry, I just wanted to share another patient’s perspective.

— Reader

The author replies:

Please allow me to stand with you and other IG Living readers who rely on their ports for venous access; without them, we’d risk undeserved scars from failed IV attempts, exasperated NICU, critical care and ER nurses and, from recent personal experience, a very impatient anesthesiologist who blew his top right along with four precious peripheral veins. Finally, but certainly not the least, it hurts like nobody’s business to have a well-trained, highly educated, compassionate medical professional dig for a vein that disappears like Houdini, is as flat as Richard Simmons’ abs and flips like Kathy Rigby! My article was simply my vain attempt at poking fun at those who do not share our enthusiasm for our precious pain (and vein!) preservers. If it wasn’t for my daughter’s very patient and savvy pediatric nurse who suggested a port, we’d still be poking at her highly coveted, uber-guarded and sole-veneinous access in her right foot! We love ports so much, my father got one, then my son Caleb and, now, yours truly. I enthusiastically concur: Without my port, my arms would be one hole short of a championship golf course, and I’d be the potty-mouthed patient no one in their right mind would want to access. If my words offended rather than entertained, I’ll schedule an intimate date with a bar of Dial.

— Cheryl Haggard

Revised PANDAS/PANS Diagnostic Criteria

Thank you very much for your article on PANDAS and PANS in the December-January issue of IG Living. I did want to bring to your attention that the diagnostic criteria for PANDAS were revised in 2012. They are different than what you have listed. Readers can view them on the National Institute of Mental Health website at intramural.nimh.nih.gov/pdn/web.htm.

— Vickie Blavat

PANDAS Network

Information-Sharing: A Two-Way Street

The recent article about subcutaneous immune globulin (SCIG) in the April-May issue helped me so much! After seven-plus years on intravenous IG, I had two really bad reactions and had to switch to SCIG. I had home health help for three infusions, and I am now on my own — six or so infusions now. There were little details about the setup and prep that no one had explained, and your article cleared up several things for me.

— Nancy Wagner Johnstone

I enjoyed the article titled Coping with Chronic Illness in the August-September 2012 issue, and found it to be timely and informative. I don’t think you can ever exhaust that particular topic, and every little insight can help patients and caregivers. We all have people in our lives who just don’t get it, and the more they can understand, the better our relationships will be.

— Karen Dawn Wheat

Your feedback, opinions, suggestions and anything else you’d like to share are important! Email us at editor@IGLiving.com or visit www.IGLiving.com and go to the Be Heard/Feedback page to send your comments.
The Neuropathy Action Foundation (NAF) and the GBS/CIDP Foundation International have launched a joint nationwide campaign to raise awareness of multifocal motor neuropathy (MMN), a rare and incurable neurological condition in which multiple motor nerves are attacked by the immune system. The national campaign will include an educational MMN-specific brochure, as well as a public service announcement (PSA) to help patients and medical professionals identify, treat and manage the progressive condition.

“Multifocal motor neuropathy is a serious but treatable condition where early and accurate diagnosis is critical to preserving the livelihood of those touched by the disease. The ability to control the progression of MMN is directly related to how quickly the disease is correctly diagnosed,” said NAF Founder Dominick Spatafora. “I was originally diagnosed with ALS and told that I had only three to five years to live. It took more than a year before I was correctly diagnosed with MMN and began receiving the life-sustaining IVIG [intravenous immune globulin] treatments that continue to help me 10 years later.”

Although MMN is a rare disease — likely affecting no more than one to two in 100,000 people — it can cause serious disability if not correctly diagnosed. Most MMN patients are originally misdiagnosed multiple times before correctly being diagnosed with MMN, and the correct diagnosis may take years. To view the PSA, go to www.youtube.com/watch?v=0j0g_YtwhcE&feature=plcp&nomobile=1.

The Jeffrey Modell Foundation (JMF), in collaboration with physician experts and patient groups from 32 countries, launched World Primary Immunodeficiency Week (WPIW), which began April 22. Weeklong events brought together patients and their families, healthcare professionals, scientists and others from the industry to raise awareness for earliest possible diagnosis and appropriate treatment of primary immunodeficiency (PIDD). One of the highlights of the week was a simultaneous launch of thousands of specially designed balloons at Jeffrey Modell Diagnostic and Research Centers throughout the U.S. and around the world.

“To affirm our dedication, passion and commitment to our mutual mission, Jeffrey Modell diagnostic, research and referral centers throughout the U.S. and abroad will be participating in this event by launching balloons on the same day. This will signify how, since inception, WPIW has ‘ballooned’ into a global movement,” said Vicki Modell, co-founder of JMF. “We need to be sure that each child and adult has the best chance of being diagnosed early and effectively treated. Awareness is critical, and World Primary Immunodeficiency Week is an excellent opportunity to drive this global movement to educate the medical community and the public about primary immunodeficiencies.”

JMF also sponsored a global competition for the most innovative, creative, unique and cutting-edge idea to create awareness of PIDDs during WPIW. Three winners were awarded a prize of $5,000 each to fund their events during WPIW.

Vicki and Fred Modell established JMF 26 years ago in memory of their son, Jeffrey, who lost his battle with PIDD at age 15. JMF is a global organization dedicated to research, physician education, patient support, public awareness, advocacy and newborn screening. There are 196 Jeffrey Modell Centers in 68 countries spanning six continents.
Wednesday is my FREEDOM60® infusion day.

When I switched to HlgH-Flo Subcutaneous Safety Needles™
I noticed better looking infusion sites on Thursday!

HlgH-Flo doesn’t hurt going in or on removal - and cut 15 minutes off my infusion time. They come with authentic 3M Tegaderm™ so it doesn’t irritate my skin the way other dressings can.

I use only FREEDOM60 Precision Flow Rate Tubing™. By adding HlgH-Flo needles, I’m able to achieve the best infusion from my FREEDOM60® pump.

Changing to HlgH-Flo needles was the easiest switch ever!

Joanna T.
hypogammaglobulinemia patient, NY

connections matter.

Only genuine FREEDOM60 Precision Flow Rate Tubing™ and HlgH-Flo Subcutaneous Safety Needles™ were designed to provide the most accurate and unobstructed infusion with your FREEDOM60® pump – ensuring a complete and unified system.

Ask for them by name.
Research

Institute to Study Neuro Immune Conditions

In March, Nova Southeastern University’s College of Osteopathic Medicine opened an institute to treat patients with neuro immune conditions such as chronic fatigue syndrome. The institute, which plans to treat about 1,300 patients, will also be the first in the nation to study neuro-inflammatory and neurodegenerative disorders such as Parkinson’s disease and multiple sclerosis using the newest genomic techniques.

The researchers will measure which genes turn on or off in patients, as well as the causes of relapse and illness persistence. Experts say this research will help scientists develop new medications to treat these illnesses.

Research

Short Course of IVIG Slows Early Alzheimer’s

Results from a new study in patients with early Alzheimer’s disease showed a short course of intravenous immune globulin (IVIG) slows the disease’s progression. Conducted at the Sutter Neuroscience Institute in Sacramento, Calif., the study hopes to replicate results of an earlier Phase II trial of IVIG in Alzheimer’s disease patients, but with a lower total dose of IVIG. In that trial, mild to moderate Alzheimer’s patients who received IVIG infusions every two to four weeks, beginning with a six-month randomized phase and continuing with a one-year open-label extension, experienced reduced brain atrophy and a near halt in their cognitive decline.

In this new study, patients with early Alzheimer’s disease who received five doses of IVIG showed significantly less brain atrophy after one year than a placebo group. One-year results from the first 28 patients in a planned two-year, placebo-controlled trial showed a 5.7 percent reduction from baseline in MRI-measured ventricular brain volume among the 14 receiving IVIG, compared with an 8.76 percent decline in the 14 assigned to placebo infusions. A total of 52 patients with mild cognitive impairment attributed to Alzheimer’s disease were enrolled in the study, although two dropped out before completing the eight-week course of IVIG therapy. Of the remaining 50 patients, the last to enroll had just recently finished dosing.

Because IVIG is derived from human donor plasma and is extremely scarce, production capabilities for all current suppliers could not begin to provide enough for all patients with mild to moderate Alzheimer’s disease — let alone the even larger population with mild cognitive impairment — should the treatment prove to be effective. Therefore, if a short-term dosing regimen is as effective as continuing therapy, it would stretch the existing supplies to cover a greater number of patients. The eight-week, five-dose schedule used in the new study is the same as that already used in several other neurological applications and could reasonably be expected to be beneficial, said Shawn Kile, MD, of Sutter Neuroscience Institute.
A 7-YEAR-OLD girl with chronic sinusitis and a history of other problems presented for evaluation of possible immunodeficiency. She was reported to have been essentially well during infancy, but her parents thought that she may have had a milk protein allergy. She had not been previously evaluated for allergies. Around 3 years of age, she began having problems with recurrent ear infections and bouts of sinusitis, which required multiple courses of antibiotics. When she presented for her evaluation, she had had symptoms of sinusitis for greater than four months that had not responded to treatment with oral antibiotics.

The girl’s family history, as best as could be accounted, did raise some suspicion for antibody deficiency. There had been both paternal and maternal relatives with recurrent infections such as sinusitis, though no one was known to have had a formal evaluation for an antibody deficiency. Of course, this is not surprising due to the difficulty and known delay in obtaining a primary immunodeficiency diagnosis.

The typical expected course of events in childhood is for the infant to be relatively well through the first 3 months to 6 months of life. Protective maternal antibodies that pass through the placenta into the fetus begin to wane at birth and are generally thought to drop below protective levels for most by 3 months to 6 months of life. Consequently, children must be making their own antibodies to the infectious organisms to which they are exposed beginning at birth. Children will have, on average, approximately six respiratory infections a year as their immune system becomes “educated” against the pathogenic organisms to which they are exposed in the normal environment. In general, by 5 years or 6 years of age, most children have gained sufficient immune education to remain mostly free from infections. This further improves so that by the age of puberty, most children are reported to have outgrown tendencies to become commonly ill with infections. Further, the current practice of receiving immunizations against pneumococcal bacteria and Haemophilus influenzae type B early in life enhances this earlier progression toward an adult-type immune status, and greatly reduces the morbidity and mortality due to infections with these organisms that previously plagued infants and children. Typically (except possibly in some large day-care settings), by 3 years of age, the infection rate has decreased and a more normal adult-appearing immune system has begun to be established.

This girl was having an opposite course of events. She was initially well, and then around 3 years of age began having recurrent sinopulmonary disease that worsened into recalcitrant sinusitis by 7 years of age. This type of presentation is suggestive of the development of some form of antibody deficiency such as common variable immunodeficiency. And, since some forms of antibody deficiencies have features that can be inherited, it seemed this girl may have an antibody deficiency.

However, it’s always possible that contributory environmental and other genetic factors can mimic the possibility of an antibody deficiency, or they can cause children with physiologically delayed maturation of immunity to experience worse symptoms. But, these factors also could make symptoms worse in someone who actually has an antibody deficiency. Therefore, such factors as exposure to cigarette smoke or other pollutants and airborne irritants, gastroesophageal reflux (stomach acid getting into the nose, sinuses and Eustachian tubes), excess exposure to respiratory viral infections (which is possible in large day-care settings), allergies, and a consideration of such diseases as cystic fibrosis must be evaluated and aggressively treated to be completely successful in the overall treatment.

We will continue with this case next issue, exploring the physical examination and some interesting test results.

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Disclaimer: This case report is intended as an illustration for education purposes only. It does not necessarily represent an individual or precise information from patient files.
Did You Know

Routine Vaccine Guidelines for Immunodeficient Patients

VACCINES SAVE THE lives of more than three million people worldwide each year and prevent millions of others from suffering from diseases and permanent disabilities. But, vaccines aren’t always safe, nor are they always effective, for everyone. This can be especially true for individuals who are immunocompromised. Following are some guidelines about which vaccines should and should not be administered to patients with primary and secondary immune deficiencies and their family members and caregivers.

What Are the Recommended Vaccines?

The Centers for Disease Control and Prevention (CDC) recommends routine vaccinations for individuals at different ages. For newborns and up to age 1, the recommended vaccines include hepatitis B, rotavirus oral, DTaP (diphtheria, tetanus, pertussis), Hib (H. influenzae), pneumococcal conjugate, poliovirus inactivated and seasonal influenza. Vaccines recommended for children ages 1 through 7 include MMR (measles, mumps, rubella), varicella, hepatitis A, boosters for earlier vaccines (except rotavirus) and seasonal influenza. For children ages 7 through 18, the recommended vaccines include Tdap (tetanus, diphtheria, pertussis), meningococcal conjugate, papilloma virus vaccine, boosters for incomplete vaccines, and seasonal influenza. Tdap is a booster immunization that offers continued protection from those diseases for adolescents and adults. Adults ages 18 through 59 are recommended to receive tetanus, adult diphtheria and acellular pertussis every 10 years, meningococcal vaccine (for college students), complete MMR, hepatitis A, hepatitis B, varicella and seasonal influenza. At age 60 and older, the recommended vaccines are pneumococcal polysaccharide, pneumococcal conjugate, Tdap, zoster (shingles) and seasonal influenza. There also are some additional vaccines for high-risk adults or for adults who have high-risk family members.

Which Routine Vaccines Should Immunodeficient Patients Not Receive?

Live virus vaccines pose the greatest risk for individuals with immunodeficiencies. Those with severe antibody deficiencies (e.g., X-linked agammaglobulinemia and common variable immunodeficiency) should not receive the oral polio (OPV, although it is no longer available in the U.S.), smallpox, live Bacillus Calmette–Guérin (BCG), live oral typhoid (Ty21a), rotavirus or yellow fever vaccines, nor should they receive the live attenuated influenza nasal vaccine (LAIV). Individuals with less severe antibody deficiencies (e.g., selective IgA deficiency and IgG subclass deficiency) should not receive OPV, live BCG, rotavirus or yellow fever vaccines. However, other live vaccines appear to be safe.

All live vaccines should be avoided by those with T lymphocyte (cell-mediated and humoral) disorders, including those with complete defects (e.g., severe combined immunodeficiency disease and complete DiGeorge syndrome) and partial defects (e.g., most patients with DiGeorge syndrome, Wiskott-Aldrich syndrome and ataxia-telangiectasia). Patients with phagocytic function deficiencies (e.g., chronic granulomatous disease, leukocyte adhesion defect and myeloperoxidase deficiency) should avoid all live bacterial viruses.

There are no contraindicated vaccines for patients with complement deficiencies (e.g., persistent complement, properdin or factor B deficiency).

Which Routine Vaccines Should Immunodeficient Patients Receive?

Inactivated vaccines do not represent a danger to immunocompromised individuals and generally should be administered as recommended for healthy persons. Certain vaccines are recommended or encouraged specifically because immunosuppression is a risk factor for complications...

By Ronale Tucker Rhodes, MS

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from vaccine-preventable diseases. However, because a relatively functional immune system is required to develop an immune response to a vaccine, an immunocompromised individual may not be protected even if the vaccine has been given. And, those who are receiving immune globulin (IG) also may not receive any benefit from the vaccines because the antibodies in the IG already provide the protective effect. Even so, there are some vaccines that are specifically recommended that immunodeficient patients receive regardless of whether they may or may not be effective.

All individuals with primary and secondary immune deficiencies should receive the pneumococcal vaccine. Those with severe antibody deficiencies should also consider the measles and varicella vaccines. However, the effectiveness of any vaccine for these patients is uncertain and depends on the humoral response. In addition, IG interferes with the immune response to the measles vaccine and possibly to the varicella vaccine. So, in general, immunodeficient patients who make no antibodies don’t need routine vaccines. In those with less severe antibody deficiencies, all vaccines are likely effective; however, the immune response might be weakened.

Vaccines are likely ineffective in individuals with complete T lymphocyte defects. However, the effectiveness of a vaccine in those with partial T lymphocyte defects depends on the degree of immune suppression. And, it is recommended that individuals with partial defects also receive the meningococcal vaccine and Hib vaccine (if not already received as an infant).

For individuals with complement deficiencies, all routine vaccines are likely effective, but it is specifically recommended that they also receive the meningococcal vaccine. All live viral vaccines and inactivated vaccines are safe and effective for those with phagocytic function deficiencies.

It’s very important that all individuals, regardless of their type of immune deficiency, receive the annual seasonal influenza vaccine. This is especially true because the flu virus mutates from year to year, and immunodeficient patients are at increased risk of complications from the flu. In fact, the influenza vaccine may actually stimulate T cell immunity to provide patients with better recovery should they contract the flu.

Vaccines and Immune Globulin

Immunodeficient patients often wonder why they need to receive recommended routine vaccines when also receiving immune globulin (IG), since IG is a sterilized solution obtained from pooled human blood plasma that contains the immunoglobulins (or antibodies) to protect against the infectious agents that cause various diseases. It’s true that patients receiving IG don’t necessarily need or don’t respond to vaccines. However, those vaccines that are safe for immunodeficient patients are still recommended. The reason is that people receiving IG are using other people’s antibodies to help fight off or prevent an illness from occurring. But, this protection is temporary and should not be confused with getting an immunization, which provides longer-term protection.

Vaccines for Family Members and Caregivers

Family members and caregivers of immunodeficient patients also need to be concerned about which vaccines they receive. In general, all routine vaccines for children and adults in the household need to be kept up to date. Specific recommendations for household contacts include vaccination against influenza, Tdap, pneumococcal and MMR. The MMR vaccine, although consisting of attenuated, live viruses, is not contraindicated in household contacts of immunocompromised persons because transmission of vaccine viruses does not occur. Individuals 60 and older should ensure they receive the pneumococcal polysaccharide vaccine and the shingles vaccine.

Better Safe than Sorry

Routine vaccines may not provide the same protective benefits for immunodeficient patients as they do for those with healthy immune systems. However, for those vaccines that are safe for patients to receive, the benefits far outweigh the risks. Therefore, patients — even those being treated with IG — should be sure to receive those routine vaccines that are recommended. And, it is essential that family members and caregivers of immunodeficient patients keep up to date on their vaccines and that they avoid vaccines that are contraindicated for loved ones and patients, unless noted.

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

References are available upon request by emailing editor@IGLiving.com.
Bridging the Patient-Doctor Communication Gap

Communication breakdowns create friction in every type of relationship. But, when miscommunication occurs between physicians and patients, the result can be a prescription for frustration.

By Trudie Mitschang
After suffering through two years of chronic pain, Tammie Allegro, a marketing professional in California, was desperate for answers. The busy mother of two made an appointment to see her primary care doctor, and after describing her symptoms, was surprised by his dismissive response. “He told me: ‘Your pain is caused by your weight. Just lose a little weight, and the pain should get better. Oh yeah, you should really be exercising, too!’” Humiliated, Allegro accepted the doctor’s judgmental assessment and attempted to follow his advice. “I started working out again, and I tried to lose weight, but moving made everything worse. This created such a sense of failure for me, I began to think I would always be in pain and no one would ever understand.”

Unfortunately, Allegro’s experience is not unusual. Dominick Frosch, PhD, an associate investigator at the Palo Alto Medical Foundation’s Research Institute and associate professor at the University of California, Los Angeles, explored this issue during focus groups in San Francisco to examine how patients discuss healthcare issues with their physicians. He found that even well-educated patients feel intimidated in the physician’s office. “In the context of a medical consultation, people feel uniquely vulnerable,” Frosch said. “Asserting their views might require disagreeing. Patients fear that will lead to negative consequences that might impact their care in the future.”

The Link Between Miscommunication and Misdiagnosis

Nearly 200,000 Americans die from medical error every year, with the majority of cases attributed to misdiagnosis. While there are many factors contributing to this disturbing number, at its root is a consistent lack of clear communication between doctors and the patients they treat. The problem is a complex one; limited time for appointments leads to rushed interactions that increase the likelihood of miscommunication; an imbalance of power defines the patient-doctor relationship, with the doctor clearly in a position of control; a pronounced language barrier creates ample opportunity for confusion, with patients often speaking descriptively and emotionally, while physicians tend to communicate in a more objective, detached manner, often using medical terminology and jargon that can come across as cold and uncaring. Patients are often the most vocal when it comes to complaining about these issues, but clearly frustration exists on both sides.

Studies show the No. 1 complaint patients have about their doctors is the perception that they simply don’t listen; at the top of the list for practitioners, however, is what many call the “Google stack”: the reams of paper patients bring in containing research they’ve done about their symptoms, with the expectation that doctors should simply confirm the patients’ self-diagnosis. While patients assume this research is helpful, in reality, the behavior is viewed as counterproductive. Of course, like it or not, the trend toward patients researching symptoms online is here to stay, and some patient advocates insist physicians need to not only accept, but embrace this new normal. “Over 75 percent of the people in the U.S. have access to the Internet. Patients reading about diseases — and even reading the latest journal articles — is a reality,” says Rick Labuda, founder of the Conquer Chiari Foundation, an organization dedicated to improving the experiences and outcomes of Chiari and syringomyelia patients. “Trying to stop it is like trying to stop a tsunami. It would benefit both parties for doctors to accept, and encourage, that patients want and need to be informed.”

Moving Away from “Cookbook” Medicine

In the book When Doctors Don’t Listen: How to Avoid Misdiagnoses and Unnecessary Tests, author Dr. Leana Wen, an emergency physician at Brigham & Women’s and Massachusetts General and a clinical fellow at Harvard Medical School, reveals what patients have long suspected: Doctors often tune out a patient’s story during consultations, focusing instead on specific symptoms, which may lead to overtesting and overtreating. Wen says this style of “cookbook medicine” is both outdated and dangerous, and she advocates for patients to speak up for themselves during routine exams. “I encourage patients to ‘tell their story and speak up if they are interrupted,’” says Wen.
Studies show the No. 1 complaint patients have about their doctors is the perception that they simply don’t listen.

Wen says the communication hurdle does not end with a diagnosis, and she encourages patients to ask plenty of questions, even if the doctor seems reluctant to provide answers. “Talk through your diagnosis with your doctor, and make sure you understand its predicted course,” she says. “What treatment options do you have, and what risks and benefits do they carry? If your working diagnosis turns out to be wrong, what warning signs should you be on the lookout for? It’s important to be proactive in your own healthcare.”

The Importance of Empathy

In an April 2013 survey in The Wall Street Journal titled “The Experts: How to Improve Patient-Doctor Communication,” Rita Redberg, professor of medicine and a cardiologist at the University of California San Francisco (UCSF) Medical Center, says a tendency to look for the “quick fix” has resulted in multiple problems within doctor-patient relationships. “Doctor-patient communication has changed in many ways since I graduated medical school more than 30 years ago. The recent introduction of electronic health records in the office, for example, requires many doctors to spend much
of a patient exam looking at a computer screen instead of the patient in order to record information," she explains. “This kind of distraction means it is more important than ever to listen carefully for what ails the patient. We have a tendency for a ‘quick fix,’ which often means ordering a test or writing a prescription. We need to be sure we are treating the symptom or problem that brought the patient in.” ³

Fred Hassan, chairman of Bausch & Lomb, agrees, adding that empathy is an essential, but often missing component of today’s average patient/doctor interaction. “The single biggest thing is to have empathy and to actively listen and communicate. Doctors are not taught the importance of this skill very well in school,” says Hassan. “More recently, the reimbursement pressures and frequency of patients per hour are creating new ‘justifications’ for some doctors to not connect with their patients at a deeper level. Brochures and iPads help with communications — but empathy makes the decisive difference.” ³

Chronic Illness Compounds Miscommunication

Patients living with chronic disease face several communication challenges not experienced by their healthier counterparts. For one thing, they often see multiple physicians, requiring them to “tell their story” repeatedly. These various specialists who comprise the patient’s medical team may or may not communicate clearly with one another, leading to more frustration on the part of the patient. Artist Rebecca Zook, who was diagnosed with common variable immunodeficiency in 2008, has had numerous interactions with specialists over the years, some good, some frustrating. “I am fortunate that both my current primary care physician and hematologist are good listeners who take time with me and take my concerns seriously precisely because of my rare illnesses,” she says. “Along the way to a diagnosis, though, I met a few doctors [whom] I didn’t click with in the least. It seemed like they felt they knew everything and wouldn’t tolerate questions and concerns from me, or they didn’t have an answer and were afraid or embarrassed to admit it. I don’t expect doctors to know everything, and I recognize that there may be missteps along the way, especially if you have an uncommon illness. But I also know my body and know when something isn’t quite right and will push to make sure I am heard and not dismissed; some doctors don’t like that.”

Still, Zook is reticent to place all blame on doctors, acknowledging that communication is a two-way street. “Sometimes patients need to follow the instructions they were given. The listening goes both ways,” says Zook. “Remember, the doctors can get just as frustrated with us as we can with them. A partnership built on mutual trust and respect is the goal.”

Patients living with chronic disease face several communication challenges not experienced by their healthier counterparts.

For Allegro, the solution to her frustration came in the form of a second opinion. When attempts to lose weight failed and her pain became unbearable, her husband urged her to consult a different doctor. Allegro says this new practitioner modeled what good doctor-patient communication should include: extensive questioning, empathetic listening and a commitment to resolve the problem through accurate diagnosis and treatment. Small gestures, like eye contact, also made a huge difference. “For the first time in years, I felt like there was hope for me,” says Allegro. “He healed the hurt that my previous doctor caused. This man restored my faith in doctors and in me. I didn’t feel crazy anymore. He could see from the look in my eyes that I wasn’t making anything up, and I could see from the look in his eyes that he really wanted to help me.”

TRUDIE MITSCANG is a staff writer for IG Living magazine

References

It is still unknown what causes this nerve disorder, but research is uncovering some interesting information that could lead to prevention and better treatment.

By Annaben Kazemi
In 2004, Mabelle, a successful young chef working in the Chicago area, woke up with an excruciating headache that went from the back of her neck up to the top of her head. The pain was so intense, she felt nauseous. She made it in to see her primary care physician, who immediately sent her for a CT scan. The doctor delivered the stunning news that Mabelle, an otherwise healthy and active 32-year-old, was suffering from either a possible stroke or multiple sclerosis (MS). Mabelle took some time off from work, and after numerous tests, she saw a neurologist who conducted a spinal tap and said it was most likely MS.

As her symptoms progressed, Mabelle moved to Washington, D.C., where she could be closer to family. Then, in 2005, she experienced numbness in her right leg and had trouble walking. After more testing and another neurologist referral, she was finally diagnosed late in 2006 with MS. “Managing MS is an ongoing process, beginning with the very first symptoms and then continuing,” explains Mabelle. “I don’t think my story is unique; the journey to diagnosis is typically long and complicated.”

Having a support network of professionals, family and friends helped during the most trying times of her journey. But, “there were times when it was a struggle, and I felt very frustrated,” says Mabelle. The hardest time was when high-dose corticosteroid treatment failed, and her symptoms progressed. She spent two weeks in the hospital while her doctors tried short-term high doses of corticosteroids (methylprednisolone and then dexamethasone) given intravenously. She then began both physical therapy and counseling to adjust to all the changes her body was going through.

Nonetheless, she says, “Having MS doesn’t mean going from a great future to no future. There’s a way to balance your health concerns with your goals. My life was going in one direction, then this thing [MS] came along and made me reassess what’s really important. Time is much more precious. I don’t want to waste time doing things that don’t fulfill and serve my purpose.”

More than 2.1 million people are affected by MS worldwide. In the U.S., it is estimated that more than 350,000 people have MS. But, because symptoms can be completely invisible, the actual prevalence of MS is not completely certain.

What Is MS?

MS is a chronic, unpredictable disease that attacks the central nervous system (the brain, spinal cord and optic nerves). It is an inflammatory nerve disorder in which myelin sheaths around axons of the brain and spinal cord are damaged, leading to loss of myelin and scarring. The damage is caused due to the destruction of the insulating layer surrounding neurons in the brain and spinal cord. This insulation, called myelin, helps electrical signals pass quickly and smoothly between the brain and the rest of the body. When the myelin is destroyed, nerve messages are sent more slowly and less efficiently. Patches of scar tissue, called plaques, form over the affected areas, further disrupting nerve communication. The symptoms of MS occur when the brain and spinal cord nerves no longer communicate properly with other parts of the body.

More than 2.1 million people are affected by MS worldwide.

There are four basic types of MS. Relapsing-remitting MS (RRMS) is the most common form of the disease. Approximately 85 percent of people with MS begin with a RRMS course. RRMS is characterized by clearly defined acute attacks with full recovery or with a residual deficit upon recovery. Periods between relapses are characterized by a lack of disease progression. Some people with RRMS go a year or more between relapses; others have them more frequently.

RRMS patients often develop secondary-progressive MS (SPMS) somewhere between 10 years and 15 years after their initial diagnosis of RRMS. When this happens, patients notice a change in the pattern of their disease. While some acute attacks (exacerbations) and periods of remission may still occur, they happen less frequently, recovery is less complete, and symptoms become chronic, gradually worsening over time.

Primary-progressive MS (PPMS) is diagnosed in approximately 10 percent of MS patients. With this form, exacerbations, or attacks, are rare, if they occur at all. Instead, MS symptoms worsen over time, gradually leading to disability.

Progressive-relapsing MS (PRMS) is the least common type of MS. Like PPMS, this form is characterized by a gradual worsening of symptoms over time, but patients also experience exacerbations and remissions. Unlike RRMS, however, people with PRMS do not typically regain complete functioning after a symptom relapse. Disability is caused by the combination of disease progression and incomplete recovery after an attack.
Symptoms of MS

MS is considered an autoimmune disorder because the immune system incorrectly attacks healthy tissue. It causes a wide variety of symptoms and can affect vision, balance, strength, sensation, coordination and bodily functions. In severe cases, patients can become paralyzed and/or blind, while in milder cases, there may be only numbness in the limbs.

MS is a chronic, unpredictable disease that attacks the central nervous system.

Symptoms present differently among patients. Some may have a single symptom and then go months or years without any others. It's even possible for a symptom to occur just one time, go away and never return. However, most patients with MS, particularly in the beginning stages of the disease, experience relapses of symptoms that are followed by periods of complete or partial remission. Partial remission can last weeks or even months.4

Early symptoms of MS may include blurred or double vision, lack of clarity in thinking or trouble concentrating, clumsiness or a lack of coordination, a loss of balance, numbness and/or tingling, and weakness in an arm or leg.

While no two people have exactly the same symptoms, there are some common indicators:

- About half of patients say they feel a “pins and needles” sensation. They may also have numbness, itching, burning, stabbing or tearing pains.
- About eight in 10 patients have bladder problems. They may need to urinate more frequently and urgently, need to go at night or have trouble emptying the bladder fully. Bowel problems, especially constipation, are also common.
- Patients may have difficulty walking because MS can cause muscle weakness or spasms. Balance problems, numb feet and fatigue can also make walking difficult.
- It’s common for patients to feel dizzy or lightheaded.
- About eight in 10 people feel very tired or fatigued and complain about weak muscles, slowed thinking or sleepiness. Some feel tired even after a good night’s sleep.
- Muscle spasms are another common symptom, usually affecting the leg muscles. For about 40 percent of patients, spasms are an early symptom. In PPMS and PRMS, muscle spasms affect about six in 10 people.5

Causes of MS

What causes MS is not clear, but the underlying mechanism is thought to be either destruction by the immune system or failure of the myelin-producing cells.6

Some studies suggest that genetic factors make certain individuals more susceptible to MS than others. Yet, while MS is not directly inherited (a great deal of evidence suggests that most people who are genetically susceptible must still be exposed to some other factor or factors in their environment or life experience for MS to develop), genes play an important role in who gets the disease. The risk of developing MS in the general population is one in 750, yet the risk rises to one in 40 in anyone who has a close relative (parent, sibling, child) with the disease. In families in which several people have been diagnosed with MS, the risk may be even higher. And, even though identical twins share the same genetic makeup, the risk for a person whose identical twin has MS is only one in four chance of contracting it — which again suggests that some factors other than genetics are also involved.7

More than twice as many women as men have MS. Dr. Brian Weinshenker of the Mayo Clinic and recipient of the 2011 John Dystel Prize for MS Research examined the genetic underpinnings of MS and found that more women had variations in the gene that instructs interferon gamma, a molecule that ratchets up the immune attack in MS. The genetic variations were associated with different levels of production of interferon gamma by immune cells.8

In all parts of the world, MS is more common at northern latitudes that are farther from the equator and less common in areas closer to the equator. Researchers are now investigating whether increased exposure to sunlight and the vitamin D it provides may have a protective effect on those living nearer the equator. While MS occurs in most ethnic groups, including African-Americans, Asians and Hispanics/Latinos, it is more common in Caucasians of northern European ancestry. However, some ethnic groups, such as the Inuit, Aborigines and Maoris, have few if any documented cases of MS regardless of where they live.9

Infectious agents are most often proposed as the triggering factors of MS, yet evidence suggests that geography, ethnicity, genes and other factors interact in some complex way to cause MS. Researchers have examined
environmental and industrial toxins, diet, trace metal exposures and certain climatic elements such as sunlight. But, none has been shown to be causally linked to MS, and exactly what factors are the cause remains an open question.\textsuperscript{10}

**Diagnosing MS**

The diagnosis of MS is notoriously tricky. There is no single test,\textsuperscript{11} and confirming a diagnosis can become a waiting game because the doctor must find evidence of two episodes of disease activity in the central nervous system that have occurred at different points in time.\textsuperscript{12} Most often, a diagnosis can take years because doctors must rule out all other possible explanations.

Dr. Weinshenker’s most important lesson for clinicians: “Make sure the diagnosis is right. We see a lot of people who are put on MS treatments but who do not have MS.” In fact, Dr. Weinshenker conducted a landmark study that tracked the course of MS, which helped develop ways to distinguish MS from look-alikes. His research resulted in a clarification of an MS diagnosis by defining no fewer than 79 “red flags” that point away from MS.\textsuperscript{13}

Most people are diagnosed between the ages of 20 and 50, although MS has been detected in patients as young as 2 and as old as 75. It is unclear why the disease appears so early in some children.\textsuperscript{14}

While more people are being diagnosed with MS today than in the past, epidemiologists have found no evidence to suggest that the disease is on the increase. More likely explanations include a greater awareness of the disease, improved medical care and more effective tools for making a diagnosis. In addition, the availability of effective treatments makes physicians more likely to communicate the diagnosis to their patients.\textsuperscript{1}

**MS Treatment Options**

Although there is no cure for MS, effective treatments are available to modify the disease course, treat attacks and relapses, manage symptoms and improve function and safety. In combination, these treatments enhance the quality of life for people living with MS.\textsuperscript{15}

Permanent damage to nerve fibers (called axons) occurs early in MS in association with the destruction of myelin. In addition, overall brain atrophy can occur early in the disease, and damage can be ongoing even when patients have no symptoms of an attack and feel well. Therefore, MS specialists advise the early use of a medication such as Gilenya (fingolimod), a once-daily oral capsule,\textsuperscript{16} that effectively limits lesion formation and brain atrophy. In the opinion of the National MS Society’s Medical Advisory Committee, limiting lesions may be a key to reducing future permanent disability for many people with MS.\textsuperscript{17}

The vast majority of people who experience acute attacks respond well to the standard high-dose corticosteroid treatment.\textsuperscript{18} For those unresponsive to steroids, plasma exchange therapy (also known as plasmapheresis, a blood-cleansing procedure) may be a possible alternative treatment.\textsuperscript{19} However, plasma exchange should be considered a treatment alternative for only the few who do not respond to treatment, and only for a short time. And, in people with relapsing forms of MS, it may be effective as a secondary therapy for exacerbations that have not responded to treatment with corticosteroids. Plasma exchange has not been found to be effective for SPMS or PPMS.\textsuperscript{20}

Intravenous immune globulin (IVIG) remains a controversial treatment option. In the late 1990s, there were a few studies that showed patients with RRMS had reduced exacerbations of their MS when treated with IVIG.\textsuperscript{21} Since that time, pharmaceutical companies have developed more disease-modifying therapies more specifically targeted
to MS (Avonex, Copaxone, Rebif, Tysabri, Gilenya, etc.), and additional studies have shown no statistical significance when IVIG is added to these treatment regimens.22

Some physicians treat MS patients with IVIG postpartum. MS patients seem to develop a natural disease-modifying effect when they are pregnant, so most go off of their therapy during pregnancy and do fairly well. After they deliver, however, the protective effect of the pregnancy is gone, and these patients may be at a higher risk of exacerbating soon after delivery. In addition, if patients are planning to breast-feed, they usually remain off of their current therapy.23

Some studies suggest that genetic factors make certain individuals more susceptible to MS than others.

Managing Day to Day

While MS is not considered a fatal disease since the vast majority of patients live a normal life-span, patients may struggle to live as productively as they desire, often facing increasing limitations.24

Medication can help manage disease symptoms, yet there also needs to be a focus on function — improving or maintaining the ability to perform effectively and safely at home and at work. Rehabilitation is an important component of comprehensive, quality healthcare for patients with MS at all stages of the disease. Rehabilitation programs include physical therapy, occupational therapy, cognitive rehabilitation and vocational rehabilitation. Rehabilitation professionals focus on overall fitness and energy management, while addressing problems with accessibility and mobility, speech and swallowing, and memory and other cognitive functions.25

Future Outlook

Many advances have been made in the fight against MS. And, each advance interacts with the others, adding greater depth and meaning to each new discovery. The National MS Society supports and funds research activities spanning all research stages. This research provides information about relationships among factors so that the disease can be better understood and helps explain who gets MS and why.26

Finding the underlying causes of MS brings researchers closer to developing a cure. Because MS is significantly more common (at least two to three times) in women than men, the gender difference has stimulated important research initiatives looking at the role of hormones in MS. How hormones influence inflammation and neuron and glial function is being slowly unraveled. There is increasing evidence that estrogen, progesterone and testosterone contain immune responses and influence damage repair in the nervous system. Hormones such as prolactin and vitamin D are being explored as immunomodulators and how they may influence MS or may be used therapeutically to modulate the immune response. More recently, hormones such as leptin and ghrelin have been found to possibly influence the course of disease.27

Alberto Ascherio, MD, DrPH, professor of nutrition and epidemiology and associate professor of medicine at the Harvard School of Public Health, is conducting research to identify causes and risk factors for MS and other neurodegenerative diseases, as well as biomarkers that may provide information about susceptibility, and that may lead to earlier diagnosis. One infectious factor that Dr. Ascherio’s team and others continue to pursue is Epstein-Barr virus (EBV), which causes several disorders, including infectious mononucleosis. Most people in the U.S. show signs of having been exposed to EBV. In one study, the team reported that individuals with signs of significant exposure to EBV were twice as likely to develop MS up to 20 years later.28

Research also continues to be conducted to find new therapies to treat MS. U.S. and German researchers have developed a therapy that stops the autoimmune attack against myelin in its tracks without impairing the normal function of the immune system. The experimental treatment targets T cells in the brain that are responsible for the disease. According to Stephen Miller, a microbiologist and immunologist at Northwestern University in Evanston, Ill., the therapy has a different mode of action from current MS treatments, which suppress the immune system. The current treatments “will not only try to down-regulate the autoimmune response that’s actually causing the disease, but will also make patients, in the long run, susceptible to everyday infections and increased rates of cancer,” states Miller. In effect, they sweep away the dead T cells. The
experimental treatment resets the patients’ immune systems, halting the leukocytes’ attack on the nerve sheaths and reducing the assault on the myelin by 50 percent to 75 percent. While not a cure for MS, Miller believes it’s a step in the right direction.\(^{29}\)

**The Hope for a Cure**

As research continues, determining the cause of MS, as well as ways to prevent it, becomes more likely. But, that will require a greater understanding of the genetics that make people susceptible to developing the disease, and an identification of the environmental triggers that should be avoided or otherwise derailed. In the meantime, new therapies are being developed to help patients live healthier lives.

“Having MS has helped me learn to balance and cherish the things that are important in life,” says Mabelle. But, “I’m hoping the researchers find something that not only stops the progression of MS from happening, but also reverses the damage that has already been done … because I still have to live every day with the symptoms that were caused by previous attacks.”

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

**References**

Rituximab: An Autoimmune Disease Therapy

Autoimmune disease patients who do not respond to standard therapies are increasingly being treated with rituximab with safe and effective results.

By Amy Ehlers, BS, PharmD, BCPS

Rituximab (Rituxan) is a monoclonal antibody that was first approved in 1997 for the treatment of B-cell lymphoma. It was considered a breakthrough treatment for B-cell lymphoma due to its lower side-effect profile when compared with traditional chemotherapy treatments alone. Antibody therapies are different from chemotherapies because they target malignant cells while sparing healthy cells. Typically, chemotherapies kill any cell, good or bad, that is rapidly dividing, which is what leads to the well-known chemotherapy side effects of hair loss, bone marrow suppression and inflammation of
gastrointestinal membranes (mucositis). Antibody therapies, on the other hand, kill only those cells identified by the immune system and are known as immunomodulatory agents. Because rituximab is an immunomodulatory agent, doctors have recently begun prescribing it for the treatment of several autoimmune disorders.

How Rituximab Works

Understanding how rituximab works requires a brief overview of the B cells (also known as B lymphocytes) and their role in the immune system. Lymphocytes are white blood cells that are responsible for the activity of the immune system. They are initially formed in the bone marrow and either migrate to the thymus gland and develop into T cells (also known as T lymphocytes) or remain and develop into B cells. T cells are the main regulators of immune function. Specific subsets of T cells provide signals to stimulate B cells. The B cells are responsible for producing antibodies to the antigens. Some of the activated B cells will form memory B cells that allow for a quicker response to the antigens upon further exposure.

Rituximab works by specifically targeting the CD20 protein on B cells. When rituximab attaches to the CD20 protein, the B cells may be further attacked by complement proteins, which result in their destruction. The B cells coated with rituximab may be removed from the circulation by a particular part of the immune system known as the RES (reticulo-endothelial system), or in some instances, the B cells merely internalize the CD20 protein with the rituximab antibody attached. Because the CD20 protein is found on mature B cells, it does not affect the formation of future B cells, thereby avoiding ongoing or continuous suppression of the immune system.

The side effects of rituximab include infusion reactions such as fever, headache, chills and nausea. More serious adverse events include hepatitis B reactivation, cardiac arrhythmias, renal toxicity, tumor lysis syndrome (TLS) and progressive multifocal leukoencephalopathy (PML). However, these events rarely occur, and they may be due to some other factor than rituximab.

Rituximab as an Autoimmune Therapy

As rituximab’s successful use began to increase, interest grew in its use as a treatment for autoimmune diseases due to its action as an immunomodulating agent. In 2006, rituximab received its FDA-approved indication for moderate to severe rheumatoid arthritis in combination with methotrexate in patients who have had an inadequate response to TNF (tumor necrosis factor) antagonist therapies.

Since then, there have been hundreds of small studies and case reports involving the use of rituximab in many autoimmune disorders with varying degrees of success. This use of rituximab is considered by the FDA as off-label, but it may be considered when other traditional first-line therapies have failed for autoimmune diseases such as pemphigus, Guillain-Barré syndrome, polymyositis/dermatomyositis and myasthenia gravis.

Because rituximab is an immunomodulatory agent, doctors have recently begun prescribing it for the treatment of several autoimmune disorders.

Pemphigus is an autoimmune disorder in which there is significant blistering of the skin and mucous membranes due to antibody formation against the proteins that join skin cells together. Blisters may form anywhere on the body, including the mouth and throat. Standard treatment for pemphigus includes high-dose oral steroids (prednisone) and/or an immunosuppressant such as azathioprine (Imuran). In cases that are refractory to these, intravenous immune globulin (IVIG) is used. However, if patients are not candidates for IVIG, there are nearly no viable options, which led to the trialing of rituximab in these patients. In a retrospective study on 47 patients with pemphigus who were treated with biweekly rituximab infusions, the results showed a remission rate of 76 percent after the first cycle of two infusions of rituximab at days one and 15, and the remission rate reached 91 percent after additional rituximab cycles.1

There also have been some successful reports of using rituximab in patients with Guillain-Barré syndrome (GBS) who had other comorbid conditions. In a case study of a patient with acute lymphoblastic leukemia in remission who underwent allogeneic peripheral blood stem transplantation and developed GBS, the patient was treated...
with rituximab once a week for four weeks, which depleted the B lymphocyte targets of the Epstein-Barr virus and improved muscle strength.\(^2\) Therefore, while the No. 1 treatment for GBS is plasma exchange and, in many instances IVIG, rituximab may be another possibility.

Polymyositis and dermatomyositis are autoimmune diseases in which there is inflammation of the muscles (polymyositis) or the skin, joints, esophagus or lungs (dermatomyositis) accompanied by weakness in the affected areas. As with pemphigus, standard treatments include corticosteroids or immunosuppressant drugs with IVIG use in patients unresponsive to these therapies. In one study of 13 patients with refractory myopathies who were treated with rituximab twice within a two-week interval, positive results were sustained for an average of 27 months in the primary outcomes of disease activity as measured by creatine phosphokinase, lactate dehydrogenase and muscle strength.\(^3\) In yet another pilot trial of seven patients receiving rituximab infusions once weekly for four weeks, patients were followed up after one year without additional rituximab infusions, and all evaluable patients exhibited significant clinical improvement in muscle strength. However, return of symptoms varied among patients.\(^4\)

**Expanding Its Use**

As more autoimmune diseases are diagnosed, the future use of rituximab will be expanded. There is already interest in using it for systemic lupus erythematosus, idiopathic thrombocytopenia purpura and Sjogren’s syndrome. Most autoimmune disease treatments include corticosteroids and/or immunosuppressants. But, if these therapies are unable to control or minimize the disease process, or if patients develop intolerable, unwanted or unmanageable side effects, patients have few options. IVIG may be an alternate therapy, but due to some patients’ comorbid health conditions and insurance coverage concerns, it may not be appropriate for all. In these patients, rituximab has the potential to be part of their medication regimen. To date, however, very few controlled clinical trials for the use of rituximab in autoimmune diseases have been conducted, and much of what is known about the drug’s potential is based upon case reports. But, these reports have shown rituximab to be safe and tolerable for most patients with most side effects being infusion-related, allowing physicians and patients the option to try rituximab when other choices have failed.

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The process of diagnosing an immune deficiency is very complex and, many times, labor-intensive. There are so many unknowns that come with a diagnosis, beginning with its cause. Once diagnosed, patients must receive regularly scheduled treatments of immune globulin (IG), one of five antibodies produced by the body and the main antibody defense against bacteria that can lead to frequent infections. Essentially, IG is immune-deficient patients’ lifeline.

Many restrictions are placed on immune-deficient patients and their families as they try to remain infection-free, which can lead to a feeling of hopelessness and helplessness, especially for caregivers. Many caregivers want to know what they can do to help. And, there is something very important they can do: Donate plasma!

The Plasmapheresis Process
IG is manufactured with donated plasma. Plasma, the clear portion of the blood, is donated through a process called plasmapheresis, a sterile and self-contained process that separates whole blood into cellular parts with the use of a special machine. During plasmapheresis, a needle is inserted into the donor’s arm while the donor is connected to the machine. Blood is extracted and passed through a filter in the machine to separate out the red blood cells and platelets. The red blood cells and platelets are infused back into the donor, while the plasma goes through yet another filter and then is collected into a bag. The collected plasma goes through a rigorous screening process to ensure that it is infection-free so it can be manufactured into IG and other lifesaving therapies and then safely infused into patients.

By Sharon Burton-Young, RN

Plasma: How and Why You Should Donate

Donating plasma is one of the best ways caregivers can help patients who rely on immune globulin.
**Donating Plasma**

Several donors are needed to provide the amount of plasma necessary to manufacture the prescribed dosages of IG for one patient. For example, 130 donations are needed to make enough IG to treat a 150-pound adult primary immunodeficiency disease patient for one year.

The plasmapheresis donation is regulated by the U.S. Food and Drug Administration (FDA), which has stringent guidelines that must be followed to protect both the donor and the recipient. Before becoming a donor, an individual must be able to meet certain criteria. He or she must be at least 18 years old, but not older than 65 years. That individual also must weigh at least 110 pounds, pass a physical examination, be pre-screened for communicable diseases, provide an extensive medical history and provide a valid ID at the time of donation (these criteria may vary from state to state). Donors also cannot have any piercings, tattoos or tattoo touch-ups within 12 months prior to donating. In addition, individuals who are sick, women who are or may be pregnant, and those who are insulin-dependent diabetics, or who have heart disease, cancer, hepatitis, malignant tumors or HIV/AIDS shouldn’t donate plasma. Last, those with low protein levels or high fat levels in their plasma won’t be able to donate.

**Increased Demand = Increased Need for Donors**

The demand for IG is increasing as more individuals are diagnosed with an immune deficiency and as IG therapy is used to treat additional disease states. Indeed, the use of IG tripled between 1992 and 2004, with an annual increase of 11 percent.

An awareness about the importance of plasma donation is paramount. The last major shortage took place in 1998, and it is hoped that another shortage can be avoided. Those interested in becoming a plasma donor can visit [www.donatingplasma.org](http://www.donatingplasma.org) and conduct a Zip code search to find the location of a plasma center closest to them. They can then contact the center to learn of any specific requirements for donating plasma.

Caregivers are our secret weapons, and plasma donation can be another part of their arsenal. It takes commitment to be a plasma donor. Donors must go through a rigorous screening process and dedicate themselves to leading healthy lifestyles so they remain in good health and can continue to donate. By committing to being a part of this lifesaving effort, caregivers can find comfort in knowing that patients will continue to receive the IG therapy necessary to maintain productive and fulfilling lives.

**SHARON BURTON-YOUNG, RN, is the founder and CEO of Infusion Care of Delaware. She is an infusion nurse who completed her formal nursing education at the University of Delaware in Newark, and she has dedicated her career to advocating for her patients, but even more important, educating them so they can be advocates for themselves.**

**References**

Ask the Experts

Reader: I had to switch intravenous immune globulin (IVIG) providers, and they substituted the 50 grams of Flebogamma 5% I’d been receiving with 48 grams of 3% Carimune NF. Is this comparable?

Michelle and Leslie: The product switch may be related to your provider’s formulary or to reimbursement. From the standpoint of providing you with the antibodies provided by IG, the two products are comparable. However, there are some differences in the way the two products are manufactured, which your provider should be able to explain to you. The two major differences between Flebogamma and Carimune NF are the stabilizers and the final format of the product (liquid versus powder). Flebogamma is manufactured as a 5% liquid ready-to-use solution, whereas Carimune NF is a powder product that must be mixed with sterile water in order to be administered. Flebogamma is stabilized with d-sorbitol, and Carimune NF is stabilized with sucrose. IVIG products stabilized with sucrose have been linked to an increased risk of kidney problems. This doesn’t mean the new product may not be right for you. It would be a good idea to talk again with your new provider to ask about the sucrose stabilizer and whether or not there is any concern for you. You may also want to discuss the product selection with the physician who orders your IVIG infusions. Last, how you tolerate the infusions of the different products is important. If you have increased side effects with one product versus another, you should discuss this with your provider.

Reader: I am 68 and have common variable immune deficiency, and I have been receiving 30 grams of intravenous immune globulin (IVIG) monthly for four years. The three-hour hospital clinic routine is successful, but I’m considering trying subcutaneous IG (SCIG) to achieve more consistent IgG levels (avoiding month-end fatigue) and to allow myself more flexibility in travel plans. Before making the switch, I have several questions: 1) Do more frequent infusions actually reduce cyclical energy dips for most patients? 2) Does infusing once a week increase emotional stress because it requires confronting the disease more often and keeping medical equipment at home? 3) What about product choice? I’m weighing the advantage of using an IG product that requires no refrigeration (easier for travel) versus another that does, which is the same brand I currently use. 4) Does Medicare impose restrictions on switching back and forth between different infusion methods?

Leslie: Some people who infuse weekly have indicated they feel better on the weekly infusion schedule and don’t experience the just-prior-to-infusion cyclical dips you describe. Whether there will be increased emotional stress infusing at home really depends upon the person. There is not a lot of medical equipment required to infuse subcutaneously, so it would be easy to store out of sight when not being used. Many of the current IVIG and SCIG products have some stability at room temperature, which may help with travel. If you travel frequently, using a product that doesn’t require constant refrigeration may prove to be beneficial for you. A change in product should be discussed with your physician and IG provider.

As for Medicare, Medicare Part B does not restrict switching products or routes of administration, nor is there a prior authorization process for Part B-covered diagnoses. For SCIG, Medicare covers only four products: Hizentra, Gammagard Liquid, Gamunex-C and Gammaked. Switching between these four products is easy, but if you are not currently infusing one of these products, you would not be able to use it for subcutaneous administration. Coverage varies based on the site of care. Under Medicare Part B, more diagnoses are covered in the outpatient hospital and doctors’ office settings than at home.

It may benefit you to speak directly with a patient who has had the same questions and who has made the transition. One way you could do that is to enroll in CSL Behring’s Voice2Voice peer-to-peer support program at Hizentra.com/V2V or by calling (877) 355-4447.

Michele Greer, RN, is vice president of sales for NuFACTOR Specialty Pharmacy.

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Let’s Talk!

By Annaben Kazemi

Ida Wagner caught our attention when she commented on our Facebook page that she was reading IG Living magazine in Norway. Because the primary immunodeficiency disease (PIDD) population is so small in Norway, Ida began searching outside of her country for resources and information. She uses Facebook as a way to connect with others about her disease.

Ida Wagner has CVID and lives in Norway, where less than 500 people have a PIDD diagnosis. She uses Facebook as a way to connect with others about her disease.

Ida: It’s pretty lonely. I have a few contacts, actually one in my own country and one in Sweden, our “neighbor” country, who I can say are my friends. I have always looked pretty healthy outside, but I am struggling with the feeling of being alone and the isolation that an invisible disease brings. There are only five million people in Norway, and less than 500 have been diagnosed with a PIDD. I know there are some other people with PIDD in my city (I’m in Oslo), but I don’t know them. I have a membership with the Swedish Society, which is a wonderful way to get information by post and ordinary papers. Facebook is an absolutely amazing way to get in touch with people all over the world. We can all learn from one another and share information. It’s surreal to find we have so much in common with others, even though they may be halfway around the world.

Annaben: What is your diagnosis? Ida: I was very lucky to get a diagnosis at all! I was 47 when I was finally diagnosed with common variable immune deficiency (CVID). I administer my own subcutaneous infusion of Gammanorm (similar to Gamunex-C in the U.S.) at home once a week, but more often when I am traveling. Getting immune globulin (IG) infusions have changed my life, enabling me to travel, swim and enjoy life.

Annaben: What is it like to have a rare disease in a country where less than 500 others have PIDD?

Ida: I have learned that having a PIDD in the U.S. can be extremely challenging economically. We have a completely different healthcare system in Norway, where we all get free medical help, dental help and free medications, as well as equipment. I have also learned that invisibility and isolation are barriers many people feel no matter where they live. It’s wonderful to be included and surrounded by brilliant, caring people online on Facebook, where I’ve found the most support.

Annaben: What have you learned from your U.S. friends on Facebook?

Ida: I have learned that it’s important to be your own advocate. Never give up: You know your body better than anyone else. It took years for me to get my diagnosis. I had doctors tell me it was all in my head and that my problems were psychological. Through persistence, I finally found a doctor who listened, and I was diagnosed by the medical chief of immunology and infection at the Oslo University Hospital. My message is to never ever give up your fight to get the right diagnosis and the medication that can change your life.

Annaben: What inspires you and keeps you going?

Ida: I feel that I am very fortunate to swim every day. I am able to get in a treatment pool at the University Hospital, where it’s heated to a temperature of 96.8 degrees Fahrenheit. I do exercises in the pool every day with instructions from a physiotherapist and the help of two nurses. I feel this is one of the wonderful things that helps my body to heal and keeps me going every day. As far as I know, I am the only adult outpatient with the diagnosis of CVID who’s using this treatment pool. The results while using the pool for the last two years have been very motivating for me.

Annaben: What would you like others living with a PIDD to know?

Ida: I would like others to know that it’s important to be your own advocate. Never give up: You know your body better than anyone else. It took years for me to get my diagnosis. I had doctors tell me it was all in my head and that my problems were psychological. Through persistence, I finally found a doctor who listened, and I was diagnosed by the medical chief of immunology and infection at the Oslo University Hospital. My message is to never ever give up your fight to get the right diagnosis and the medication that can change your life.

Annaben Kazemi is a patient advocate for IG Living magazine.
“WHY ME?” That was a question I used to ask myself, among others such as “Why do I have to go through this?” “Will life ever get better for me?” “What’s even the point anymore?” and, worst of all, “Why should I keep on living?” I know there are probably many of you who have asked yourselves these same types of questions. It’s hard enough being a teenager, but when you have an illness, it can be very hard, and it’s easy to become depressed.

I have common variable immune deficiency (CVID), which has made life very difficult for me. CVID has caused me a lot of pain; I get sick easily and often, and I have to get treatments. But, most of the time, the emotional pain has been much worse than the physical pain. I’ve had a lot of negative emotions, and I’ve felt angry and sad that I have to live like this.

I think the worst part about having this disease was being lonely. I am enrolled in the homebound program at school, which makes it hard to meet new people or make friends. I’m content with the few friends I have, since I’m not much of a social person, but for a while, I was still lonely. No one I knew had a condition like me. I didn’t have someone who could understand what I go through or who I could relate to and share my pain and trials with. That made me feel I was in a very dark, sad and lonely place, and it seemed like there was no escape. I spent all of my time playing video games. I still play video games, because it’s a good hobby for me, since I can’t go out or do things like play sports, but when I was depressed, video games completely consumed my life. They could take me out of my depression, or at least make me forget about it for a while. I thought I was going to live in sorrow for the rest of my life, and at that point, I honestly didn’t care how much longer my life was going to be. It is very sad and a little bit scary thinking back on that.

Now, my life has gotten so much better, and I wouldn’t change a single thing. That’s why I want to write this article — to show other teenagers that things can and will get better. There are people out there who can understand what you go through. I found this out when I met my girlfriend, Mary. She also has CVID, which is how we met. It helps us that we both have the disease because we can better relate to each other, and we understand how each other feels. Mary fills me with so much happiness that spending one second with her makes up for all of the pain and sadness I’ve ever had. I know most teenagers say those types of things when they are in a relationship, and those relationships usually never last, but it is different for me and Mary; we truly are soulmates. I have also grown religiously, which has helped me a lot.

If you are feeling like I did, know that you don’t have to feel alone; there are others out there who understand what you are going through. When you are feeling down and are having a bad day, just remember that there is always tomorrow; life will get better. You haven’t experienced half of the good life has to offer, and you still have your whole life ahead of you. Don’t let your problems get the better of you and keep you from living a good life. I hope all of you can become as happy as I am now.

KONNER LIVELY lives in Princeton, W.V., where he is enrolled at Princeton Senior High School in the homebound program. He has been receiving weekly, self-administered subcutaneous immune globulin (IG) treatments for the past two years, and he enjoys hunting, painting, playing video games and NASCAR.

Check out the IG Living Teen page at IGLiving.com/IGLTeen.aspx

Teens with a chronic illness who rely on IG therapy have unique life experiences. This column is an opportunity for them to share their stories and to connect with other teens. Teens are invited to submit their stories of 600 words or fewer to editor@IGLiving.com.
HAVING A DISEASE like common variable immune deficiency (CVID) is frightening. Constant illness can be overwhelming; it is extremely expensive (for me, it's $7,200 per week), frustrating (very few physicians are knowledgeable about the disease), and it affects not only the patient (constantly saying at the last minute we cannot do something because we are sick again), but also our families, friends and, usually, our employment. Here is what we need to do to cope with this largely misunderstood illness:

**Attitude.** Yes it is rare, but there are hundreds of others around the globe who are facing this illness right now. If you have been properly diagnosed and have found a physician who is experienced, it is a miracle! Most do not get a proper diagnosis until they have faced constant serious illness for years. Look for support groups on Facebook. There are 1,200 others from every country on the globe who face the same ups and downs we do. Knowledge is power! It guides us to prepare and helps to alleviate fear.

Share the sorts of things you face frequently. Discuss the pain, the feeling that you have the flu five out of seven days, the harshness of the needles, as well as the exhaustion. Share the amount of time it takes to feed your body replacement therapy and visit the doctor every few days. Get it all out, and then rejoice! Get past the “Woe is me!” “What am I going to do?” “I could die!” thought process. You still have choices. Those choices make the difference in how you view this disease and react.

**Health.** The word “health” may seem like an oxymoron when used in the same sentence as CVID, but there is a lot you can do to make your life easier. I’ve studied natural health for years and have greatly benefited from it. We are all familiar with physicians who, after sitting and hearing all about our symptoms, aches and pains, digestive issues, congestion, headaches, insomnia, dermatological problems, etc., prescribe another medicine to add to the many we already take. The problem is that side effects from these medicines can tumble us into a long list of unnecessary drugs.

Treatments are an invasion of our bodies and rough on our kidneys. We must drink a lot of water to protect our organs from damage. Flavor it lemon, mint, orange, cherry or whatever you enjoy, but absolutely do it without question. Keep yourself nourished with a lot of fruits and vegetables. The vitamin C in them is great for your immune system and your skin. Antibiotics deplete the digestive system of good bacteria. All CVID patients have taken a lot of antibiotics. I personally was on antibiotics for 12 months in a row before diagnosis. Probiotics have literally been a lifesaver.

Listen and teach. Manage your illness by listening to yourself and what works. Only you truly understand the details of what it means to be sick. Most of us do not “appear” sick every day. Those around you need to be taught what it’s like to feel the way you do in order to empathize and support you. Pretending you are normal doesn’t help. It’s OK to complain that your legs hurt, you’re weak, you cannot go to the party this time, or you cannot concentrate today because your IgG levels are down. It is not your fault you are ill, nor is it anyone else’s. Your inner process should allow yourself to cry, sleep, pout, take a hot bath or whatever it takes to have a few hours of self-service.

After that, listen to everyone else. It takes your mind away from illness and toward healthy living. Smile, go out into the sunshine, watch a funny movie, play with the kids, hug your husband, get your hair cut, have a picnic and believe you control this disease through bits and pieces of time. You may not be able to go out and play a volleyball game, but you might be able to get a lawn chair and watch others play! There is joy in deciding to participate by watching them smile and listening to them giggle. It teaches them you love them despite how bad you sometimes feel.

**Healing.** The word “health” may seem like an oxymoron when used in the same sentence as CVID, but there is a lot you can do to make your life easier. I’ve studied natural health for years and have greatly benefited from it. We are all familiar with physicians who, after sitting and hearing all about our symptoms, aches and pains, digestive issues, congestion, headaches, insomnia, dermatological problems, etc., prescribe another medicine to add to the many we already take. The problem is that side effects from these medicines can tumble us into a long list of unnecessary drugs.

**Healing.** The word “health” may seem like an oxymoron when used in the same sentence as CVID, but there is a lot you can do to make your life easier. I’ve studied natural health for years and have greatly benefited from it. We are all familiar with physicians who, after sitting and hearing all about our symptoms, aches and pains, digestive issues, congestion, headaches, insomnia, dermatological problems, etc., prescribe another medicine to add to the many we already take. The problem is that side effects from these medicines can tumble us into a long list of unnecessary drugs.
Who’s Afraid of the Big Bad … Tubing?

By Cheryl L. Haggard

Note to reader: What you are about to read is real. This has been written from true accounts of the unexplainable. Do not adjust your magazine or call your doctor; nothing will help you now, because you are in “The Home Care Zone.”

“BEEP! BEEP! BEEP!” Caleb’s pump rang out, signaling the last precious drop of immune globulin (IG) had infused successfully into our 14-year-old son’s once immune-suppressed body.

Not only does our primary immune deficiency disease (PIDD) kid’s pump let us know he’s done, it also tells Caleb’s home care nurse, Nancy, and me to get our calendars ready to schedule his next infusion.

“OK, so four weeks from now, let’s see, looks like the … uh … 13th. Friday the 13th,” I said, matter-of-factly.

Nancy shot me a look like I had just missed something important, like an elephant skipping rope in my kitchen while giving her trunk a sinus wash. I studied my calendar as if it were my entrance exam for the School of Hard Knocks, and there “it” was; I couldn’t believe I had missed such an important date! “Thanksgiving Day!” I announced with gusto.

“If you’re Canadian,” Nurse Nancy answered back with deep disappointment in me. “Seriously, Cheryl! Haven’t you heard of Friday the 13th? It is a very unlucky day!”

“Only in Canada,” I snickered. Nancy wasn’t impressed with my ability to shrug off a 24-hour span of time known for causing otherwise everyday “normal” folks to avoid black cats, breaking mirrors or walking under ladders. “Well, how ‘bout you and I plan on consuming inhuman amounts of garlic and keep the date?” Nancy challenged.

“Uh, sure!” I agreed, despite a slight
hesitation in my gut. “I’m not worried about the boogeyman tampering with the tubing!”

“Alrighty then,” Nancy chimed. “We’re on!”

The hesitation in my gut went from a dull roar to a full-blown screamfest. What if I’m not taking this seriously enough? Roaring aside, I wrote down the date. Nancy plunked confidently at her computer keyboard.

“See ya’ on the 13th!” Nancy chirped on her way out the door.

“Alrighty then!” I chirped right back, sort of.

Four weeks had come and gone, and Friday the 13th was upon us. I swallowed my first sip of coffee, which felt like liquid velvet comforting my early-morning routine. Tubing, pumps, mini-spikes and IG bottles lined the kitchen table ready for Nurse Nancy to do her thing. My sleepy-eyed PIDD kid Caleb made his way to the couch, EMLA secured over his port-a-catheter by Tegaderm. Cinnamon, sugar and butter filled the air as the traditional cinnamon roll breakfast treats were almost ready to come out of the oven. It’s intravenous IG (IVIG) day, and everything looked, smelled and felt like any other IVIG day. However, in the back of my mind, I hadn’t forgotten about Nancy’s reminder that this day just happened to be Friday the 13th.

“So, we ready to roll?” Nancy asked, like always. The routine was intact, and everything seemed to run like clockwork. Until…

“No way!” Nancy gasped. “No, no way!”

“What ‘No, no way!’?” I copycatted, delivering the sharps container Nancy requested.

Caleb shot up from his port access position, lying chest-up on the couch, to see what all the excitement was about.

“This is the ‘no, no way’ I was talking about!” Nancy dangled a disgraced, horribly bent, once razor-sharp 16-gauge needle used for safe port-a-catheter access from her pointer finger. The needle looked more like a craft store dollar-bin clearance item, a mere faint memory of its once-magnificent purpose.

“I don’t mean to state the obvious but, what the…?” I asked, feeling a bit foolish.

“This has never, and I mean never, in the 38-plus years I’ve been nursing, ever happened to me! All I did is what I’ve done a billion times before: Wipe EMLA, prep area, sterilize and in goes the needle. As soon as the tip hit the port, it felt like a hot knife through butter. And this is what it looked like when I pulled it from Caleb’s port,” Nancy explained, studying the crumpled mess she placed on a chuck pad.

“You don’t think…?” I paused before I completed my thought.

“No, go on. What ‘don’t you think?’” Nancy inquired with a knowing grin. “You don’t think that today’s date has anything to do with what just happened, do you?”


After the second, and might I add successful, attempt to access Caleb’s port, Nancy and I both went on with business as usual, both feeling an eerie tension in the air. After a few more sips of coffee and a cinnamon roll or two, Nancy and I got our groove back and went about solving all of the world’s problems.

“What’s your Internet password?” Nancy asked. “I’ve got to recertify Caleb.”

I rattled off the letters and numbers I’ve used day in and day out for the past 17 years — letters and numbers I could recite in my sleep, while driving, eating, jumping out of airplanes and, well, doing just about anything.

After an hour of calling my husband, Mark, at work, the cable company, the computer company, our priest and, out of desperation, an Internet dating service just to get any type of connection to our computer: nothing.

“You don’t think…?” Nancy asked me this time. I shook her off like a pitcher shaking off her catcher — bases loaded, bottom of the ninth, the winning run 90 feet from home plate with full count to none other than Big Papi himself. “No, I do not think this has anything to do with Friday the 13th,” I said with confidence, hoping that whoever was messing with my computer password would hear how annoyed (read: completely freaked out!) I was.

Beep! Beep! Beep! Beep! Caleb’s pump squawked at us unmercifully.

“What now!” Caleb complained. “Whadda I do to shut this thing off?!”

Nancy rushed to Caleb’s side. “Tubing occlusion. This should be an easy fix,” Nancy said — for the very last time. For the next hour and a half, Nancy executed enough ups and downs that’d make a pro football player lose his lunch. “I guess I don’t need to go to the gym tonight!” Nancy joked through her frustration. “I think that pump is possessed!” Caleb’s pump even managed to stump the pharmacy’s nurse, pharmacist, billing clerk and even the driver. In the end, Nancy
resorted to a wire hanger, a sturdy safety pin and gravity to infuse Caleb’s precious medicine. The pump managed to burn through 30 feet of tubing, four mini-spikes, 20 (give or take) alcohol wipes and two chuck pads. When looking at all the spent medical supplies, Nancy and I both thanked our specialty pharmacy’s patient coordinator, Britini, for making sure we had plenty of supplies on hand — not to mention the name of a good exorcist.

Six hours after the first “beep!” Nancy and I found ourselves completely exhausted, hair mangled, mascara glops dotting our cheeks — both of us finding comfort in holding a grilled cheese sandwich in one hand and a braid of garlic in the other. Nancy and I were resolved not to allow the date and the barrage of bad luck to convince either of our well-educated, clear-thinking brains to succumb to the possibility that this day was anything more than coincidence. We managed to find ourselves back at our original spots at the breakfast table, calendars and computers ready and waiting right where we left off some seven hours before.

When I counted off four weeks to schedule Caleb’s next infusion, Nancy interrupted: “Cheryl, my computer just accepted your password. Du du du du…”

“And guess what’s so special about the next infusion day?” I asked while Nancy continued to du du (the theme from “The Twilight Zone” for those of you not getting it). “Give up?” Nancy shrugged her shoulders and nodded while still “du du-ing.” “April Fools’ Day!”

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caring for people with hemophilia around the world—one at a time.
Sometimes having an autoimmune disease feels like a cruel game of tag that you don’t even know you’re playing. One day, you’re running around carefree in the game of life, and then an invisible hand reaches out and says: “Tag! You’re It!” You develop debilitating symptoms and then spend too much time trying out too many doctors to be part of your caregiving team. Your life is forever entrenched in playing this new game with rules you don’t understand, and you’re stuck having to manage a team you don’t want to be on.

But you are. That means you need the best teammates to help you win. Friends, family and support groups are all great players to have, but the captain of your team will be your doctor, or doctors, depending on how many specialists you need. I was lucky from the start to have a neurologist who is a strong captain of Team Neuropathy Girl. My rheumatologist, internist and psychiatrist are all great teammates working with her to ensure I bring my A game to life. I’ve heard so many stories from other people whose doctors aren’t as compassionate, committed or willing to get help from their colleagues to find out how to best treat their patients.

That doesn’t mean I’ve been exempt from trying out a few doctors who left me baffled that they ever chose medicine as a career. Once, after I waited three hours past my scheduled appointment time to see a hematologist, she waltzed into the room and bellowed: “Well, I don’t know why you’re here!” Then, there are the doctors who spent our whole appointment talking about themselves — prompting me to want to bill them for wasting my time! How can I ever forget the occupational therapist who kept repeatedly saying: “You don’t look sick; I’m not sure why you’re here.”

Maimonides, a fifth century physician and philosopher, wrote: “May I never forget that the patient is a fellow creature in pain. May I never consider him merely a vessel of disease.” The trust I place in my medical professionals is based on the premise they care for me as a human being. It truly matters to them whether I live or die. It is evident to me that they, too, realize that at any moment they could be tagged “You’re It!” My medical professionals aren’t separate from me; they are part of a team that deciphers the rules of a game that is little understood and keeps changing.

As the team manager, I have also learned not to be afraid to cut the dead weight. If you’re not happy with a doctor you’re seeing, I don’t care how many people recommended the person or how great their record is, if they aren’t treating you with the level of care you deserve, then they’re out. You call the shots when it comes to everyone in your organization to give you the results you want. Speak up, speak out and be the example of the change you want to see. We are only as strong as the people we choose to surround us. No matter how rare the disease, how few people have it or how few have even heard of it, we need to go to bat for one another. All of us receiving intravenous immune globulin therapy are on the same team. We need to be helpful and supportive of each other to win in this crazy game we’ve been chosen to play. So “Goooooo team!” I hope we all triumph over the hurdles that are yet to come.

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, and she is working on her supersecret identity as Neuropathy Girl, who will one day save the world after her infusion and a nap.
Parenting:
A Balancing Act:
PIDD Kids and “Typ-Sibs”

Healthy siblings often suffer negative emotions when a sibling has a chronic illness. But, parents can help them overcome that, and oftentimes, those “typ-sibs” become more compassionate human beings.

**AS PARENTS OF KIDS** with primary immunodeficiency diseases (PIDDs), we spend a lot of time tending to their needs. But what about our children who are healthy: the “typical siblings,” or as I like to call them, the “typ-sibs”? Does the quantity of time we must commit to our PIIDD kids rob the typ-sibs of their quality of childhood and cause them great psychological damage? Or does growing up with a sibling who is chronically ill somehow make typ-sibs better people? It all depends on how we, the parents, approach this difficult situation by balancing the needs of PIIDD kids and typ-sibs.

**Psychology of the Typ-Sib**
According to the American Academy of Pediatrics, when attending to the needs of our children with a chronic illness, we may be neglecting — or creating unfair expectations for — healthy children. When parents have to pay extra attention to an ill child, the typ-sibs often feel neglected. They also may have difficulty learning to live with the stresses of having a brother or sister with a chronic health problem.

Younger typ-sibs may express guilt that they themselves are not sick, asking “Why him and not me?” Or, they may feel anxious about becoming sick themselves. And, they may sometimes wish that they, too, were sick so they could be the center of attention. Older typ-sibs might become angry if they are asked to assume more household chores than their ill brother or sister, or feel guilty for resenting the additional responsibility. And, they may become embarrassed when strangers continually ask about their ill sibling’s condition. Any number of feelings may overwhelm the family’s typ-sibs, and parents need to channel those feelings into something positive.

According to John V. Lavigne, PhD, and Michael Ryan of Children’s Memorial Hospital and Northwestern University Medical School in Chicago, typ-sibs do seem more likely to experience adjustment or behavioral problems than children with healthy siblings. They appear to be at risk for certain types of disturbances at certain ages, and they tend to be more socially withdrawn and more irritable than children in families without chronically ill children.

In a study by Lavigne and Ryan, the risk of disturbances was different for boys and girls and different at different age groups. Children between the ages of 3 and 6 were more likely to show elevated incidences of overall psychopathy. Males between 7 and 13 showed more of a tendency toward emotional problems than females, and they showed higher levels of psychopathy than the siblings of healthy males, but this difference was not significant. Younger females had more problems adjusting to medical issues than younger boys. This may be because dependence is often tolerated in younger girls, so when an older sibling gets more attention, the younger sister has more problems adjusting. At the other extreme, older females do a better job of adjusting to medical issues than older males; older females usually have more household responsibilities, and taking care of a sick younger sibling, directly or indirectly, is considered just another responsibility to the older sister.

Phoebe D. Williams, RN, PhD, FAAN, who conducts research on the care of families and children with chronic illness at the Kansas University Medical Center, has found in interviews with parents who have both chronically ill children and typ-sibs that typ-sibs often have more stress than siblings in families without chronically ill kids. Specifically, she found that typ-sibs may feel they are not loved as much as the ill child or that they are pushed aside because the ill child comes first. They feel robbed of time with their parents
because of attention given to medicines, treatments and routines for the ill child. And, they are bothered by the numerous trips to the hospital that disrupt their schedule. Some parents report that an ill child’s disease might force siblings to stay home and not go out with friends, that rules might be stricter with typ-sibs or that typ-sibs may feel “put upon” because they have to help with treatment, care, etc.

What Parents Can Do

As typ-sibs become older, many of these negative psychological repercussions usually work themselves out. However, it is important for parents to be on the lookout for warning signs that might require crisis intervention strategies. For instance, is the typ-sib showing certain psychological issues such as anxiety, anger, rebellion or depression? Are they losing interest in their friends or activities that once brought them pleasure, like sports or music? Are they acting in ways to draw attention to themselves such as doing poorly in school or pushing themselves too hard to achieve?

To help overcome these negative emotions and actions, the American Academy of Pediatrics recommends that typ-sibs be allowed to participate in family decision-making and be taught to feel a sense of pride and love in helping their brother or sister with their health problem. It is also recommended that parents provide typ-sibs with honest information about the illness and listen to and answer their questions. And, parents need to establish balance between the needs of their ill child and those of their typ-sibs, making an effort to develop a special relationship with each one of their children.

Patty Curran, a former psychologist who now homeschools her children — one of them chronically ill — agrees that it is important for siblings not to be isolated from the medical events unfolding around them. Seeing what happens firsthand tends to make a big impression. It is also important for parents to let the typ-sibs know that if they were ever sick or needed hospitalization, that the same thing would be done for them. This helps them to understand that the sick child is not receiving preferential treatment.

Positive Outcomes

The presence of a family member with a chronic illness provides typ-sibs with opportunities for increased empathy, responsibility, adaptability, problem-solving and creativity. And, although many studies have focused on the negative outcomes of typ-sibs in a family with an ill child, the same studies reveal an even greater positive impact for typ-sibs. Interviews conducted by Williams and her co-workers indicate that having a sibling who is chronically ill has reportedly increased family closeness, increased a typ-sib’s sensitivity to the ill child and toward caregiving, and increased the typ-sib’s personal growth and maturation. Typ-sibs were described by their parents as being “diplomatic,” “outgoing,” “sensitive and caring about other people’s feelings” and “generous in giving, helping, caring and volunteering.” “Surprisingly enough,” says Curran, “we have watched our oldest turn into a wonderful young man. He is full of compassion and has become a wonderful volunteer in our community.”

Even more encouraging is a study from Donald Sharpe, PhD, and Lucille Rossiter, MA, both from the University of Regina in Canada. Their study reveals that, in interviews concerning typ-sibs’ emotional and psychological development, parents tended to overstate the problems that their typ-sibs were having. The upshot: Things might not be as bad as they seem. But, that shouldn’t keep parents from seeking help if they have a concern.

Added Responsibility

As parents of PIDD kids, we have been given added responsibility. Not only do we have to tend to the needs of our PIDD kids to ensure they are getting the appropriate amount of medicine at the appropriate time, planning their trips to the hospital and caring for them when they are sick, but we have to tend to the feelings of others in the home as well. The time that we spend with our PIDD kids and their typ-sibs will have a great impact on their future well-being.

My oldest son, the typ-sib of our family, has turned out well. This last summer, he went on a weeklong mission in San Francisco to help the homeless. I would like to believe that my excellent parenting instilled in him the compassion to go on such a trip, but it had to be something bigger than me. With the help of his brother and sister, both of whom have a PIDD, he has learned compassion, not just for his own family, but for the larger human family with all kinds of ailments.

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

Book Corner

When Doctors Don’t Listen: How to Avoid Misdiagnoses and Unnecessary Tests
Author: Leana Wen, MD
Publisher: Thomas Dunne Books, us.macmillan.com/ThomasDunne.aspx

In When Doctors Don’t Listen, Dr. Leana Wen, an emergency physician at Brigham & Women’s and Massachusetts General and a clinical fellow at Harvard Medical School, teaches patients that they need to advocate for their own health by doing one simple thing: asking for a diagnosis when they go to see their doctor. The book provides the 8 Pillars to Better Diagnosis, including: tell your story, even if your doctor is steering you away from a narrative and toward the chief complaints; practice your pitch before going, including writing it out; make the differential diagnosis together by continuing to ask what else could be going on; evaluate with your doctor the likelihood of each possible diagnosis; use common sense to confirm the working diagnosis (a working diagnosis should be reached at the end of every visit) and make sure it makes sense.

Fierce Joy: A Memoir
Author: Ellen Schecter
Publisher: Greenpoint Press, www.greenpointpress.org

In her late 30s, Ellen Schecter, married and the mother of two small children, began experiencing pain in various parts of her body. Her left foot felt numb, her fingers tingled, her ears ached, she saw white flashes in one of her eyes. The pain was so cosmic and so difficult to pin down that she sometimes thought it might be a figment of her imagination, that she might be a hypochondriac or a hysteric. But after two years of trying to ignore the ever-increasing anguish colonizing her body, she and her husband finally sought medical attention and a diagnosis. What she had turned out to be not in her head at all, but systemic lupus marked by inflammation of her peripheral nerves. The disease is progressive. The commonly used treatment — heavy doses of steroids — failed to slow its march through the corridors of her body. And Schecter proceeds to fight her deterioration with every scrap of will and humor she can muster. Fierce Joy is the story of this courageous woman and her battles to maintain her spirits even while she was losing her nerves.

Past Tense: 365 Daily Tools for Putting Stress Behind You
Authors: Shawn Kilgarlin and Ron Kilgarlin

This book provides guidance and useful tips that are positive, fun, lighthearted and profound so readers can get the peace of mind they really need. It is designed for easy reading and fast relief, providing 365 engaging “page-a-day” stories to put stress to rest. Each story brings a healthy dose of feeling better every day of the year. It is sprinkled with inspiring quotes, thoughtful sayings and profound wisdom from the world’s great philosophers, modern thinkers and major religions.

The Autoimmune Paleo Plan: A Revolutionary Protocol to Rapidly Decrease Inflammation and Balance Your Immune System
Author: Anne Angelone, LAc
Publisher: Self-published, www.anneangelone.com

This book explores how genetics, lifestyle and nutrition play a role in the origin of all autoimmune reactions and what individuals can do with diet and natural medicine to halt the progression and radically reduce flare-ups, attacks and inflammation. The Paleo Autoimmune Protocol is designed to be used along with treatment strategies from functional medicine and nutrigenomics to significantly alter the course of an autoimmune condition. This book was written for patients who have failed to receive adequate management of their autoimmune condition in the conventional healthcare system.
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**FAMILIES FIGHTING FLU (FFF)** is a nonprofit, 501(c)(3) volunteer-based advocacy organization dedicated to protecting the lives of children by helping to increase annual influenza vaccination rates among families. Our members include families whose children have suffered serious medical complications or died from influenza, as well as health care practitioners and advocates committed to flu prevention.

Share in our mission to protect all children against influenza and save lives. Visit [www.FamiliesFightingFlu.org](http://www.FamiliesFightingFlu.org) Call (888) 2ENDFLU (888-236-3358)
Managing GERD Symptoms

By Carla Schick

GASTROESOPHAGEAL REFLUX DISEASE (GERD) is an aggravating condition that affects many immune globulin (IG) patients. It is a more severe form of acid reflux that plagues heartburn sufferers at least twice a week with symptoms that range from the feeling of a lump in the throat and a dry cough, to chest pain and difficulty swallowing.

Experts in the fields of immunology, nutrition and alternative medicine have several suggestions on how those with GERD can successfully manage their symptoms.

Lifestyle Changes

One of the best ways patients can help manage their GERD symptoms is to examine their lifestyle, specifically their exercise habits and diet.

Krista Sheehan, author of the article What Are Good Exercises To Do When You Have Acid Reflux?, suggests three forms of exercise for those with GERD: cycling, Pilates and walking. Cycling is an energetic workout choice that allows riders to stay in an upright position. However, it is recommended that cyclists not hunch forward on the bike, as this would put pressure on their stomach and force acid into the esophagus. Pilates exercises utilize many GERD-friendly positions such as cat, side leg kicks, plank and saw poses, which are gentle stretches and slow movements that are relaxing and will not aggravate the esophagus. And, gravity involved in moderate, low-intensity walking helps with digestion.

Diet can have a profound effect on those with GERD. Common trigger foods to avoid are tomato-based sauces, fried foods, citrus fruits, chocolate, peppermint gum and carbonated beverages. Items on the “good” list include lean chicken and pork, ginger, oatmeal, low-fat milk, yogurt, ice cream, cinnamon gum and eggs. Symptoms can also be controlled by eating several small meals throughout the day instead of fewer large meals.¹

In addition to steering clear of trigger foods, GERD sufferers can turn to cookbooks that specialize in heartburn-conscious meals. Oftentimes, these books highlight informational sections that list best and worst foods for heartburn, medical guides and natural healing programs.

Friendly Bacteria

Probiotics are friendly bacteria responsible for maintaining a healthy digestive tract, which is especially important for IG patients since they are frequent users of antibiotics. Probiotics are known to restore the balance of the intestinal microflora that can become unbalanced due to illness, stress, age, traveling or the use of medication. A number of probiotic strains are currently available, but the two most common are Lactobacillus plantarum and Bifidobacterium bifidum. These are offered in an assortment of products from supplement capsules and granola bars, to yogurt and breakfast cereal. It is important that patients discuss the use of probiotics with their physician to make sure this is the best course for them.²

Over-the-Counter and Prescription Solutions

Over-the-counter (OTC) and prescription treatments also may help to control GERD symptoms. There are three different types of drugs: antacids, histamine (H2) receptor blockers and proton pump inhibitors (PPI). Antacids help by neutralizing stomach acid. Yet, while these will help to counteract the acid, they will not repair an inflamed esophagus damaged by stomach acid. H2 receptor blockers work by reducing acid production. And, although they do not function as quickly as antacids, they provide longer relief from symptoms. Stronger H2 blockers can be obtained with a doctor’s prescription if the OTC version is unsuccessful in providing relief. PPIs block acid production in the stomach and give the esophagus tissue time to heal from excessive acid exposure. Again, if the OTC versions of these treatments do not provide relief, they are also available through a doctor’s prescription.³

Controlling GERD Is Possible

It is possible for IG patients to successfully manage GERD symptoms. By making adjustments in nutrition, altering exercise routines and adding supplements, the burden of severe acid reflux can be offset.

CARLA SCHICK is a staff writer for IG Living magazine.

References

**Directory of GERD Products**

<table>
<thead>
<tr>
<th>Nature’s Bounty Advanced Probiotic 10</th>
<th>The Acid Reflux Solution: A Cookbook and Lifestyle Guide for Healing Heartburn Naturally</th>
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<tr>
<td>Nature’s Bounty Advanced Probiotic 10 is a pre- and probiotic blend that contains 10 distinct naturally derived probiotic strains and 20 billion live probiotic cultures per serving. A daily dose of two capsules can help to provide support for digestive and intestinal health and overall improved immune function.</td>
<td>The Acid Reflux Solution: A Cookbook and Lifestyle Guide for Healing Heartburn Naturally is a combination medical guide and cookbook by gastroenterologist Jorge E. Rodriguez, MD, and dietitian and food writer Susan Wyler. The 224-page paperback book features reflux-friendly recipes along with a three-step program to help manage heartburn naturally.</td>
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<th>Attune Probiotic Bars</th>
<th>Nexium</th>
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<td>Attune Probiotic Bars deliver 6.1 billion CFUs of clinically supported strains of probiotics in natural and certified gluten-free chocolate bars. Each bar weighs 0.7 ounces and is offered in dark chocolate, chocolate crisp, and mint chocolate flavors. They are available in the following quantities: individually, seven pack or 28 pack.</td>
<td>Nexium is a proton pump inhibitor (PPI) that is prescribed to treat the symptoms of acid reflux disease and to help heal esophageal damage. With one pill a day, it can repair most erosive esophagitis in four to eight weeks; however, results may vary.</td>
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<td>shop.attunefoods.com/attune-Probiotic-Bars/c/AttuneFoods@Attune</td>
<td>(800) 236-9933, <a href="http://www.purplepill.com/index.aspx">www.purplepill.com/index.aspx</a></td>
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<th>Dropping Acid: The Reflux Diet Cookbook &amp; Cure</th>
<th>Pepto-Bismol Max Strength Liquid</th>
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<tr>
<td>Dropping Acid: The Reflux Diet Cookbook &amp; Cure is a nontraditional diet approach to help treat the symptoms of acid reflux. The 200-page hardcover cookbook is written by otolaryngologists Jamie Koufman, MD, and Jordan Stern, MD, and French Master Chef Marc Bauer. It features a collection of 75 delicious, healthful recipes using only good-for-reflux foods, and also includes a section of best and worst foods for reflux sufferers.</td>
<td>Pepto-Bismol Max Strength Liquid is an antacid that helps relieve the symptoms of heartburn, indigestion, nausea, upset stomach and diarrhea. It is designed to treat individuals 12 years of age and older, is available in 4-, 8- and 12-ounce bottles, and comes in a fresh wintergreen flavor. Each 30-milliliter dose contains 1,050 milligrams of bismuth subsalicylate.</td>
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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/fleitas/bandaides
- eMedicine from WebMD: emedicine.medscape.com
- FamilyDoctor.org: www.familydoctor.org
- Johns Hopkins Medicine: www.hopkinsmedicine.org
- KeepKidsHealthy.com (pediatrician’s guide to children health and safety): www.keepkidshealthy.com
- Mayo Clinic: www.mayoclinic.com
- National Committee for Quality Assurance (detailed report cards on health plans, clinical performance, member satisfaction and access to care): www.ncqa.org
- National Heart, Lung and Blood Institute: www.nhlbi.nih.gov/health/health-topics/by-alpha
- National Institutes of Health: health.nih.gov/see-all-topics.aspx
- National Organization for Rare Disorders (disease-specific support groups and virtual communities for patients and caregivers): www.rarediseases.org
- Office of Rare Diseases Research: rarediseases.info.nih.gov
- Patient Advocate Foundation (patient access to care, maintenance of employment and financial stability): www.patientadvocate.org
- WebMD (medical reference): www.webmd.com
- IG MANUFACTURER WEBSITES
  - Baxter: www.baxter.com
  - Bio Products Laboratory: www.gammalplex.com
  - CSL Behring: www.cslbehring.com
  - Grifols: www.grifolsusa.com
  - Kedrion: www.kedrionusa.com
  - Octapharma: www.octapharma.com

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

### Disease-State Resources

#### Ataxia Telangiectasia (A-T)

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

#### Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

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

#### Evans Syndrome

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

#### Guillain-Barré Syndrome (GBS)

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

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

#### Idiopathic Thrombocytopenic Purpura (ITP)

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

#### Kawasaki Disease

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

#### Mitochondrial Disease

**WEBSITES**
- United Mitochondrial Disease Foundation: www.umdf.org
- MitoAction: www.mitoaction.org
Multifocal Motor Neuropathy (MMN)

WEBSITES
- The Neuropathy Association: www.neuropathy.org

Multiple Sclerosis (MS)

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

ONLINE PEER SUPPORT
- Friends with MS: www.FriendsWithMS.com
- MSWorld's Chat and Message Board: www.msworld.org

Myasthenia Gravis (MG)

WEBSITES AND CHAT ROOMS
- Myasthenia Gravis Foundation of America (MGFA): www.myasthenia.org

ONLINE PEER SUPPORT
- Genetic Alliance: www.geneticalliance.org

Myositis

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

Peripheral Neuropathy (PN)

WEBSITES
- Neuropathy Action Foundation: www.neuropathyaction.org

Pemphigus and Pemphigoid

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

Primary Immune Deficiency Disease (PIDD)

WEBSITES
- The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/health/topics/Primary_ImmuneDeficiency.cfm
- American Academy of Allergy, Asthma & Immunology: www.aaaai.org
- International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: www.ipopi.org
- New England Primary Immunodeficiency Network: www.nepin.org
Sources

OTHER RESOURCES

- Rainbow Allergy-Immunology: www.uhhospitals.org/rainbow/services/allergy-immunology
- 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://idffriends.org/forum
- IDF Friends: http://idffriends.org
- Jeffrey Modell Foundation Message Board: www.info4pi.org

• Michigan Immunodeficiency Foundation: www.facebook.com/groups/108048062584350
• Rhode Island peer group: health.groups.yahoo.com/group/RhodeIslandPIDD

Scleroderma

WEBSITES

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

ONLINE PEER SUPPORT

- Scleroderma Support Forum: curezone.com/forums/l.asp?f=404

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.livingwithspes.com
- Stiff Person Syndrome: www.stiffpersonsyndrome.net

Other Resources

EDUCATION AND DISABILITY RESOURCES

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

MEDICAL RESEARCH STUDIES

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

FOOD ALLERGIES

- Allergic Disorders: Promoting Best Practice: www.aaaai.org
- American Partnership for Eosinophilic Disorders: www.apfeld.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

Stiff Person Syndrome:

- IVIG Flebogamma 5% DIF and 10% DIF: www.grifols.com/portal/en/US/bioscience?div=8883
- IVIG/SCIG Gammmagard Liquid: www.gammagardliquid.com
- IVIG Gammmagard S/D: www.baxter.com/patients_and_caregivers/products/gammagard_sd_5.html
- IVIG/SCIG Gammmaked: www.gammaked.com
- IVIG Gammaplex: www.gammaplex.com
- IVIG Privigen: www.privigen.com
- SCIG Hizentra: www.hizentra.com

PUMP AND INFUSION SETS WEBSITES

- EMED Technologies: www.emedtc.com
- Marcal Medical Inc.: www.marcalmedical.com
- Intra Pump Infusion Systems: www.intrapump.com
- Micrel Medical Devices: www.micrelmed.com
- Norfolk Medical: www.norfolkmedical.com
- RMS Medical Products: www.rmsmedicalproducts.com
- Smith Medical: www.smiths-medical.com/brands/cadd

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The **Products** you need when **you need them.**

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