Coping with Chronic Illness
Understanding the Emotional Impact

The Healthful Way to Gain Weight
Understanding & Treating ITP
Parenting: Dealing with Delayed Puberty
How to Deal with Sleep Deprivation

www.IGLiving.com
August-September 2012
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--------------------------
• IgA deficient patients with antibodies against IgA and a history of hypersensitivity

---------WARNINGS AND PRECAUTIONS--------------------------
• IgA deficient patients with antibodies against IgA are at greater risk of developing severe hypersensitivity and anaphylactic reactions. Have epinephrine available immediately to treat any acute severe hypersensitivity reactions.
• Monitor renal function, including blood urea nitrogen, serum creatinine, and urine output in patients at risk of developing acute renal failure.
• GAMUNEX-C is not approved for subcutaneous use in ITP patients. Due to a potential risk of hematoma formation, do not administer GAMUNEX-C subcutaneously in patients with ITP.
• Hyperproteinemia, with resultant changes in serum viscosity and electrolyte imbalances may occur in patients receiving IGIV therapy.
• Thrombotic events have occurred in patients receiving IGIV therapy. Monitor patients with known risk factors for thrombotic events; consider baseline assessment of blood viscosity for those at risk of hyperviscosity.
• Aseptic Meningitis Syndrome (AMS) has been reported with GAMUNEX-C and other IGIV treatments, especially with high doses or rapid infusion.
• Hemolytic anemia can develop subsequent to IGIV therapy due to enhanced RBC sequestration. Monitor patients for hemolysis and hemolytic anemia.
• Monitor patients for pulmonary adverse reactions (transfusion-related acute lung injury [TRALI]).
• Volume overload
• GAMUNEX-C is made from human plasma and may contain infectious agents, e.g., viruses and, theoretically, the Creutzfeldt-Jakob disease agent.
• Passive transfer of antibodies may confound serologic testing.

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

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

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

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

Talecris Biotherapeutics, Inc.
Research Triangle Park, NC 27709 USA
U.S. License No. 1716
08939771/08939782-BS
Revised: October 2010
Important Safety Information for GAMUNEX-C

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

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

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

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

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

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

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

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

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

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

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

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


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

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

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PATRICIA CARROLL
IG Living’s Second Annual Essay Contest Winner

Winning Essay! Lessons from the Deep
“Getting in the pool for the first time taught me that my extra weight doesn’t matter and my limitations don’t define me.”

NANCY CREADON, RN
Vice President, VaxAmerica

Chronic Illness and Sleep Deprivation
“The best methods to treat sleep disorders in the chronically ill involve adherence to treatment programs to reduce symptoms and pain management.”

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

Parenting: Helping Kids Deal with Delayed Puberty
“Two immune deficiencies are specifically linked to delayed onset puberty: XLA with growth hormone deficiency (XLA/GHD) and X-linked hypogammaglobulinemia with growth hormone deficiency (XLH/GHD).”

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

Diagnosing Specific Antibody Deficiency: The Effects of the Conjugated-Pneumococcal Vaccine, Part 2
“Immunization with Prevnar or Prevnar13 has a great potential to make interpretation of pneumococcal polysaccharide antibody responses more difficult.”

AMY SCANLIN, MS
Freelance Writer

Coping with Chronic Illness
“Helping patients to realize that while they may not have control over their chronic illness, they do have control over the way they choose to view that illness, as well as control over their behaviors that can both negatively and positively impact their condition.”

JILL WEISENBERGER, MS, RD, CDE
Registered Dietitian

Gain Weight the Right Way
“Bodies rely on the proper mix of nutrients to feed the immune system, support growth, build muscle and other tissues and maintain function of every organ and body system.”

Connect with Other IG Living Readers through Monthly Teleforums!

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

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

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

Sign up for the Teleforums now by emailing rrhodes@IGLiving.com or calling (800) 843-7477, ext. 1362.
Illness Sparks Courage

It’s logical to think that when illness strikes, the human psyche will retreat in self-wallowing and fear. But, while that may initially happen for some, the indomitable human spirit is far too strong to allow it to be overpowered by fear for long in the face of an intense struggle. And that truth couldn’t be more clear from the entries in *IG Living*’s second annual essay contest.

This year, we asked entrants to submit essays with a lead sentence that began: “The last time I did something for the first time, I...” And, I am humbled by how courageous people are in the immune globulin community. After reading the entries, I was struck by the common gutsy themes running throughout the essays.

Not giving up is a particularly strong theme. Dana writes that she was told she wouldn’t live to see her 30th birthday. She learned not to “listen to those voices that resemble the ‘grown-ups’ on the Charlie Brown specials,” and at age 34, she is “keeping the flames of hope and positivity glowing” inside of her.

Others shared that because of their illness, they have learned to fight for what they believe in. Rachael was told she would never be able to walk again after being diagnosed with myositis and surviving a deadly bout of pneumonia. But, she writes, “I knew that I would walk again.” And, today, she can walk many steps, “all because of the one step I dared to take that day in the hospital.”

Many have learned to take a chance and try something new. Our second-place winner, Rachel, learned that “risk is the name of the game when the person you love most has a primary immune deficiency.” That person is her son who was diagnosed at 3 months old with severe combined immunodeficiency (SCID). Fortunately, she and her husband “took a gamble on a young resident’s hunch that it was not SCID,” decided against the recommended transplant and discovered through time and additional testing that a transplant was not medically necessary.

Others shared that they had gained a new perspective on life, and two decided to take that perspective and give back to others through what they have learned. Darlene put her health at risk and went to bat for the wildlife after the 2010 oil disaster in the Gulf of Mexico. She became a voice for the voiceless through her photographs of dolphins and turtles that were washing ashore and of birds and gulls sprayed with dispersants dying with eggs in their nests. Mike discovered that his illness is a “gold mine,” and he writes that it has helped him to understand suffering in ways that will help him to help other people. Today, he is a Buddhist priest.

In this issue, we share with you the winning essay on page 34. Patricia also decided to try something new, which required her to step out of her comfort zone. After sabotaging her attempts to feel better by attending a therapy pool strengthening and toning class, Patricia finally took the plunge and is crying “tears of relief” because getting into that pool made her realize that her limitations “don’t define me.”

Most people will never experience what illness can do to their spirit — thankfully so. But, those who do have some courageous stories to share. We think you will relate to this winning essay, as well as the second- and third-place essays that will appear on our *IG Living* blog in September.
Immunology 101:
Diagnosing Specific Antibody Deficiency:
The Effects of the Conjugated Pneumococcal Vaccine, Part 2
By Terry O. Harville, MD, PhD

AS PREVIOUSLY DISCUSSED, the goal of conjugating a polysaccharide polymer to a protein is, in essence, to “trick” the immune system to respond to the polysaccharide antigen better than would be achieved by natural exposures to the microorganism. T lymphocytes recognize and respond to protein antigens, but typically do not have an inherent ability to recognize polysaccharide antigens. However, a B lymphocyte is capable of producing an antibody directed against the polysaccharide antigen. So, if a B lymphocyte is near the activated T lymphocytes, they could recognize the conjugated protein and generate the necessary stimulants for impelling the B lymphocytes toward producing the corresponding anti-polysaccharide antibodies.

In practice, this works well, and most children with otherwise normal immunity produce useful anti-polysaccharide antibodies, which keeps them free from serious pneumococcal infections from the strains of pneumococci incorporated into the vaccine.

The initial protein-conjugated pneumococcal vaccine was Prevnar (pneumococcal 7-serotype conjugate vaccine to diphtheria CRM197 protein). The classic non-conjugated pneumococcal polysaccharide vaccine contains 23 different pneumococcal polysaccharide components. Each polysaccharide component is from a different strain of pneumococcus, and each is termed as a serotype due to how it was determined. These 23 serotypes, selected from all the strains available in nature, have been thought to be the ones most responsible for human infections with pneumococcal bacteria. From these 23 serotypes, seven were selected that were thought to be the most virulent responsible for most of the infections in infants and children. About 21 years ago, the Centers for Disease Control and Prevention approved Prevnar. Within a few years, the reduction in serious pneumococcal infections in infants and children was dramatic. Indeed, the effect was so good that a new pattern of specific serotypes emerged as responsible for causing infections in children. This led to the development and current use of Prevnar 13, which contains 13 serotypes, rather than the original seven serotypes. At some point, there may be a Prevnar 23. However, this will likely be very problematic for the diagnosis of antibody deficiency.

The diphtheria CRM197 protein is a very immunogenic protein to which most persons with normal immunity can produce vigorous T lymphocyte and antibody responses. For these reasons, it was chosen to be the conjugate protein. Since it can create such a vigorous immune response, it was anticipated to produce a significant bystander response, activating B lymphocytes capable of forming antibodies to its conjugated pneumococcal polysaccharide component.

Further, diphtheria toxoid protein is part of the routine immunizations during infancy, thereby boosting immunity against this protein, which further results in the vigorous immune activation directed toward it. Obviously, this has all worked out to the great benefit of most infants and children.

During normal testing for the possibility of an antibody deficiency, the capacity of the patient to produce specific antibodies is measured. This is generally accomplished by 1) obtaining serum from the patient (pre-immunization serum), 2) immunizing with the specific vaccine to be tested and 3) approximately four weeks after the immunization, reobtaining serum (post-immunization serum). The pre- and post-immunization sera are analyzed, and the magnitude of the antibody response is used to assess whether age antibody production is normal or not.

The vaccine antigens, as well as the immune system’s ability to react, can be divided into responses to 1) protein antigens/vaccines and 2) polysaccharide antigens/vaccines. Thus, immunization with Prevnar or Prevnar 13 could potentially result in anti-polysaccharide antibodies via the bystander-effect of the protein-activation mechanism of the immune response, rather than a response elicited solely by the polysaccharide serotype antigen. This has a great potential to make interpretation of pneumococcal polysaccharide antibody responses more difficult. For instance, can the immune system inherently make anti-pneumococcal antibodies, or is the apparent production a result of “trickery” via the diphtheria protein (a protein response rather than a polysaccharide response)? We will continue this discussion next issue.

TERRY HARVILLE, MD, PhD, is medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, and a consultant for immunodeficiencies, autoimmunities and transplantation.
The World Health Organization has identified chronic illness as the leading cause of mortality worldwide. Life insurance can help individuals with chronic illness to pay for care and, in the end, to care for their family. Some individuals are fortunate enough to have purchased life insurance, long-term life insurance or an added chronic illness rider before being diagnosed. For those individuals who did not, however, these policies can be very difficult and expensive to obtain—despite the fact that over the years, better healthcare, improved lifestyle, advances in medical care and improved diagnostic procedures have increased life expectancies for those with chronic illness.

Why Purchase Life Insurance?

“The basic reason for life insurance, which is to provide liquidity at death, allows for well-planned folks to spend the other assets they have for the care they need,” explains Logan Ostrander, a life insurance agent. And, as life expectancy and the number of people with chronic illness increase, it makes sense that there will be a greater need among the population to prepare for both expected and unexpected healthcare expenses. Just look at the statistics: The U.S. Department of Commerce estimates that more than 100 million people in the U.S. have a chronic illness; more than 20 percent of the population, about 54 million people, have some level of disability; and nearly 10 percent, or 26 million people, have a severe disability. By 2020, approximately 157 million Americans will be afflicted by chronic illnesses, according to the U.S. Department of Health and Human Services.

Can Life Insurance Pay for Treatment?

Permanent and term life insurance policies can be used as a secondary benefit to help pay for healthcare costs. “A person can take out the cash value with a loan or just cash in the policy [and] then use the funds to pay for care,” says Ostrander. “If they use a loan, the death benefit will still be there, reduced by the amount of the loan.” However, it is more customary for healthcare costs to be covered through stand-alone long-term care insurance.

Individuals also can purchase a long-term care, or chronic illness, rider. A long-term care rider “allows for a stream of funds to pay for custodial care,” explains Ostrander. However, he says, in order to qualify to use the rider, a person has to have lost two of the six activities of daily living, which include bathing, continence, dressing, eating, toileting and transferring, or have suffered from a severe cognitive impairment for at least 90 consecutive days within the previous 12 months.

Obviously, the premiums for permanent or term life insurance, stand-alone long-term care insurance and chronic illness riders are cheapest when healthy individuals purchase the policy. In fact, says Ostrander, most riders need to be purchased...
before diagnosis. Therefore, members of a family with a history of chronic illness are encouraged to purchase life insurance prior to any chance of becoming ill, if possible.

**Life Insurance Options Post-Diagnosis**

Individuals with chronic illness who have not previously purchased life insurance do have options. It is possible to purchase permanent, term and long-term care insurance. But, says Ostrander, premiums for the same coverage cost more with the shorter life expectancy that most often comes with any chronic illness. This is especially true today, as insurance companies are careful not to sell contracts that will not stand up actuarially (when the premium fails to match the risks), which has led many companies to discontinue offering these policies.

There also are life insurance policies that have been created specifically for people with chronic diseases, but again, the costs for these policies are high. The first is life insurance without a medical exam. The price of this type of policy will depend upon life expectancy. If a person has a very high chance of passing away within the term, the cost will be higher. The second is term life insurance without a medical exam. For this type of insurance, people will need to answer only a few health questions, such as their height and weight and whether they smoke. But, they will not have to give blood or urine samples. A third type is guaranteed issue life insurance. The only individuals who can be turned down for this type of policy are those who are residents of a hospital or nursing home.

**Tips for Purchasing Insurance Post-Diagnosis**

Should individuals with a chronic illness want to purchase permanent or term life insurance, there are steps that can be taken to help increase their ratings to reduce their premiums. An impaired-risk specialist is an insurance broker who is familiar with life insurance underwriting and the variables that companies will consider when insuring applicants based on their particular health issues. These specialists can point individuals in the right direction when it comes to finding the right carrier for the insurance they want, they can serve as advocates and negotiate with insurers to get the best price possible, and they can help individuals find impaired-risk life insurance coverage.

Individuals can talk to their physicians about their health and alert them to the likelihood of a request from the insurer for their medical records. If physicians think patients have progressed favorably and that they are following a prescribed health plan to control or prevent the health condition, having that opinion in the medical records will help. Individuals also should consider the timing of applying for a policy. For instance, those who have recently had treatment or surgery might consider waiting until they are fully recovered.

**Individuals with chronic illness who have not previously purchased life insurance do have options.**

**Beneficial, but Costly**

Today, life insurance companies’ actuarial tables have been expanded to a life expectancy of 121 years versus the old table that capped at age 100. So, with increased life expectancies, as well as the ability of the healthcare system to better manage chronic conditions, insurance companies are not as resistant to providing life insurance to the chronically ill. But, while this insurance can be very beneficial for the chronically ill, that coverage won’t come without increased premiums for those who purchase post-diagnosis.

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

**Gamunex CoPay Card for CIDP Patients Only**

In the June-July issue of *IG Living*, we incorrectly reported that the Gamunex CoPay Card Program offered by Grifols is a coupon program that helps patients with common variable immune deficiency (CVID) cover the copay costs for Gamunex-C. However, at this time, the CoPay program is intended only for patients with chronic inflammatory demyelinating polyneuropathy (CIDP), not CVID. The program provides up to $2,500 per patient per 12-month period, and it is open to CIDP patients who are not using any state or federally funded healthcare program such as Medicare, Medicaid, Medicare Advantage and Tricare; who are not residing or receiving treatment in the state of Massachusetts; and who are not getting treatments in hospitals or hospital-associated outpatient clinics. For additional information, go to gamunexcard.com.

**Research**

**Personalized Immune Mouse New Tool to Study Autoimmune Disease**

Columbia University Medical Center scientists have developed a new “personalized immune mouse,” a new tool that allows them to recreate an individual’s immune system to study autoimmune diseases. The mouse model also could have clinical applications, such as predicting how a particular patient might respond to existing drugs or immunotherapies. And, it could prove useful for developing individualized immunotherapies for fighting infection or cancer or for lessening a patient’s rejection of transplanted tissue.

The mouse model is made by transplanting human bone marrow stem cells (also known as CD34+ cells), along with a small amount of HLA-matched immature thymus tissue, into an immunodeficient mouse. The thymus tissue is implanted into the mouse’s kidney capsule, a thin membrane that envelops the kidney and serves as an incubator. Within six to eight weeks, the transplanted thymus tissue is seeded by circulating human CD34+ cells (which are infused into the mouse’s bloodstream), and begins generating human immune cells from the CD34+ cells.

While the researchers intend to use the personalized immune mouse to study type 1 diabetes, Dr. Megan Sykes, director for the Columbia Center for Translational Immunology, says that they “hope to find out what is fundamentally different about patients’ immune systems, compared with those of healthy individuals, before any disease develops.”

**Medicines**

**IDF Urges FDA to Exempt IG from Biosimilars Pathways**

At the U.S. Food and Drug Administration (FDA) public hearing on draft guidance of biosimilar products in May, the Immune Deficiency Foundation (IDF) urged the FDA to exempt immunoglobulin (IG) therapies from the biosimilars pathways in order to protect the safety of patients with primary immunodeficiency disease (PIDD). The FDA recognizes each IG product as unique with no generic equivalent because current science cannot demonstrate that two products will provide the exact same clinical results for a large number of patients or that switching patients from one product to another will pose no additional risks.

IDF President and Founder Marcia Boyle’s testimony urged the FDA to follow the example set by the European Medicines Agency and exempt IG therapy from the biosimilars pathways, or to, at the least, require that biosimilar drug products undergo clinical trials to determine whether a proposed interchangeable therapy will offer patients the same clinical outcome. Additionally, Boyle requested that the FDA prohibit automatic substitution of a biosimilar drug with an original biologic. “We believe the FDA’s foremost responsibility is to ensure that biosimilars are manufactured and prescribed safely,” said Boyle. “All medicines must be thoroughly tested and meet the highest safety standards set by the FDA.”
The United States Senate unanimously passed a resolution in February recognizing May 16, 2012, as Hereditary Angioedema (HAE) Awareness Day. The resolution is the result of a year-long advocacy effort to generate recognition of the need for increased professional education regarding HAE, which is a rare and potentially fatal genetic disorder. The advocacy effort also highlights the need for further research aimed at improving diagnosis and treatment options for patients.

The goals of an annual HAE Awareness Day are to increase awareness of HAE among the general public and medical community; support better care and an earlier and more accurate diagnosis for HAE patients; raise funds for further national and international initiatives; and enhance the understanding that HAE patients can lead a healthy life. HAE Awareness Day helped launch the first HAE Global Conference held in Copenhagen, Denmark in May. Findings from the conference, to be held biannually, will be the impetus for additional HAE research.

“This first annual HAE Awareness Day will put a spotlight on HAE, its symptoms, and the impact this challenging disorder has on patients and their families,” said Janet Long, executive vice president of the Hereditary Angioedema Association. “We hope this national recognition will broaden awareness of HAE and prompt anyone who suffers from repeated bouts of swelling to seek appropriate diagnosis and treatment.”

Support for the public policy program of the Hereditary Angioedema Association, which encouraged the Senate to acknowledge the need for increased awareness and research, was provided by CSL Behring through the company’s Local Empowerment for Advocacy Development (LEAD) program. For more information about HAE Awareness Day, please visit www.haeday.org.

Eight scientists each received a one-year $50,000 Discovery Grant, for a total of $400,000, for pilot projects that have potential to lead to breakthroughs in the understanding of psoriatic diseases and to discover better treatments. The grants are also intended to lay the groundwork for additional, long-term funding from the National Institutes of Health and other funding agencies. Six scientists each received a two-year $200,000 Translational Grant, for a total of $1.2 million, for studies that aim to move laboratory and clinical discoveries into projects and treatments that benefit patients. And, 12 early-career dermatologists were awarded Psoriasis Foundation medical dermatology fellowships totaling $465,000. These fellowships provide support of up to $40,000 per year to new doctors training to do research in psoriasis and psoriatic arthritis. The fellowships are intended to increase the number of doctors studying psoriatic diseases.

“National Psoriasis Foundation is committed to funding promising research,” said Chip Newton of the National Psoriasis Foundation’s Scientific Advisory Committee. “This year, due to a record number of applicants, we awarded the highest number of grants and dollars in our organization’s history. Each of these projects has tremendous potential to advance our knowledge of psoriatic diseases, lead to new treatments and, we hope, even a cure for these diseases.”
AAN Releases IVIG Guideline for Neurology

The American Academy of Neurology (AAN) has released a new evidence-based guideline on the efficacy of intravenous immunoglobulin (IVIG) — used to treat a range of immune-mediated neurological diseases — for neuromuscular disorders, based on a comprehensive review of the literature by the AAN Therapeutics and Technology Assessment Subcommittee in the 43-year period between 1966 and 2009. The guideline answers the following questions: What are the significant findings for treatment of neuromuscular disorders with IVIG? How would neurologists use these guidelines in practice? What are some of the side effects of the treatment? What are some of the alternate treatments, and how do they measure up? Why is the benefit from IVIG often short-lived? Where should further research be done? The review was published in the March 27 print issue of Neurology.

New Guidelines Issued for Severe Lupus

The American College of Rheumatology has issued new guidelines for the screening and management of lupus nephritis (kidney inflammation). According to the guidelines, patients who have not received treatment for lupus nephritis who show signs of kidney involvement should get a kidney biopsy. If there is kidney involvement, patients should be given the drug hydroxychloroquine. And, if there is any sign of protein in the urine, patients should be prescribed blood pressure-lowering medications called ACE inhibitors or angiotensin-receptor blockers.

When diagnosed with lupus, one in three patients already has kidney inflammation, and during the first 10 years with the disease, as many as 60 percent of patients will have some kidney problems. “Without treatment, lupus nephritis can lead to end-stage renal disease, which requires dialysis or a kidney transplant. But, not all types are this serious. It depends on the pattern of damage to the kidneys,” says Dr. Bevra Hahn, lead author of the new guidelines and a professor of medicine and chief of rheumatology at the David Geffen School of Medicine at the University of California, Los Angeles.

The guidelines were released online May 3 and were published in the June 2012 issue of Arthritis Care & Research.

People and Places

A researcher at the University of Texas Health Science Center at Houston has been awarded a $1.9 million grant from the National Institutes of Health’s National Heart, Lung and Blood Institute to study a novel cell therapy that could help avoid autoimmune problems after stem cell transplantation, as well as potentially treat other autoimmune diseases.

Dr. Jordan Orange, an internationally recognized leader in studying and treating primary immunodeficiency disorders in children, will lead the new Center for Human Immunobiology at Texas Children’s Hospital. His research, which will be funded by the National Institute of Allergy and Infectious Diseases, as well as the United States Immunodeficiency Network, will focus on the biology of natural killer cells and the innate immune system, with a clinical focus on primary immunodeficiency disease.

Baxter International announced plans to spend up to $1 billion over the next five years to open a new production center near Atlanta that will employ more than 1,500 workers. Construction will begin in 2012, and the plant is expected to start production in 2018.

Did You Know?

A new column titled “IG Chronicles” will debut in IG Living magazine’s December-January issue featuring autobiographical stories written by a reader about his or her life with a chronic illness. Submit your stories to editor@IGLiving.com.
Organizations

**New Organization for PIDD in United Kingdom**

The United Kingdom Primary Immune Deficiency Patient Support (UKPIPS) is a new national patient-run and patient-led organization in the United Kingdom dedicated to people with primary antibody and immune deficiencies and their caregivers. The organization, which offers them information, advice, support and hope, is currently seeking charitable status and has a highly regarded medical advisory panel.

The launch of UKPIPS will coincide with World Primary Immune Deficiency Week. “There is currently little support or information for people living with a primary antibody deficiency in the U.K., with many people often suffering from extreme ill health for a number of years — or even decades — before they are diagnosed,” says Liz Macartney, UKPIPS coordinator and trustee. “There is also a lot of misunderstanding of the condition, including even the basic principles of caring for a patient with a compromised immune system.”

For more information on UKPIPS, visit www.ukpips.org.uk.

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FDA Approval

FDA Approves Advate to Treat Hemophilia A

The U.S. Food and Drug Administration (FDA) approved Baxter International Inc.'s Advate (antihemophilic factor [recombinant] plasma/albumin free method) for routine prophylaxis to prevent or reduce the frequency of bleeding episodes in patients with hemophilia A. Advate is the only antihemophilic factor approved in the U.S. for prophylactic use in both adults and children.

The approval is based on a Phase IV prophylaxis study that demonstrated a statistically significant reduction in the median annual bleeding rate. Patients receiving on-demand treatment experienced 44 bleeds (per patient per year) compared with one bleed (per patient per year) while on either of the prophylactic regimens evaluated (a 98 percent reduction in annual bleed rate). Forty-two percent of study patients experienced zero bleeds during one year on prophylaxis. And, of the two prophylactic regimens approved for use, the dosing schedule of every three days (a pharmacokinetic-driven regimen based on patients' clinical response) offered some patients the option of fewer infusions over one year of treatment.

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Patients are very pleased and are offering us this type of feedback:

“I have done 2 infusions with the new Soft-Glide SClg sets you sent. Both times all 4 insertions were pretty much pain free. Also, my infusion times were down about 15 minutes for 8 gms of Hizentra. It usually takes about 2 hours for my infusions, and the last two times have taken about 1 hr 45 mins. I really like the new soft glide sets and will request them on my next order from my provider.

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

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

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

New research reveals that patients suffering from both breast cancer and arthritis have a more aggressive cancer, a finding that could suggest a possible treatment. Experiments by researchers at the University of North Carolina at Charlotte have shown an intimate relationship between mast cells — immune system cells that are located in various tissues and that can cause inflammation — and metastatic tumors.

Researchers worked with two strains of mice. The first group had spontaneous arthritis and the second group had spontaneous breast cancer. They found that the population of mast cells within the bone and lung microenvironment was significantly higher in those mice with arthritis and breast cancer versus those without arthritis and breast cancer. The differentiation of mast cells from bone marrow-derived stem cells also was significantly higher in the arthritic versus the non-arthritic tumor-bearing mice.

Their findings point to a relationship between the c-Kit receptor found on mast cells and on the transmembrane stem cell factor (SCF) ligand found on metastatic breast cancer cells. The interaction between SCF and c-Kit appears to play a critical role in facilitating metastasis. Because of the suspected relationship, the researchers tested the effect of blocking the receptor by treating the mice with an anti-c-Kit receptor antibody and celecoxib, an anti-inflammatory medication, which resulted in a greatly reduced incidence of breast cancer metastasis to the bone and lung.
The psychological needs of chronically ill adult patients include taking a positive outlook about their disease, learning how to control anxiety and feelings of helplessness, understanding the disease’s impact on family, and establishing good communication.
Chronic conditions affect more than 162 million people in the U.S. each year, and they are the main cause of death and disability, with economic costs topping $1.3 trillion in 2003. In addition to the health and financial tolls that chronic illness imposes on patients and their families, the psychological costs are astronomical. And, while great strides have been made in understanding and treating many chronic conditions, the emotional impacts are less well understood.

The uncertainty of chronic illness, including how and when it will manifest, poses one of the biggest coping challenges for patients. Patients often grapple with how they will live with the emotional stress of their condition, what type of support they will need from their families, and what types of concessions they will need from their employers and how that need will affect relationships within the workplace. “Dealing with disabilities or chronic illness is a very complicated process,” explains Matthew Purinton, MSW, LSW, staff therapist for the Council for Relationships. “It’s what’s known as ambiguous loss, because society does not have a ritual for the loss of function or health in the same way that it does when a person dies. Therefore, dealing with this loss and finding positive ways to cope are made more difficult by a lack of a pre-existing path or ritual for coping with the loss.”

For a mental health professional, the goal of helping patients manage their chronic conditions, as well as the emotional and social implications, requires a well-rounded approach. This includes helping patients understand the influences and effects of their chosen behaviors on their own health and that of their families. “The most difficult thing is the idea that the chronic illness will last for the rest of their life,” explains Purinton. “Usually a chronic illness doesn’t have a known end point. Instead, the person is told that they will always have to deal with the symptoms of the illness for the rest of their life. It’s also difficult to see how an individual’s chronic illness affects the lives of the people they love. A chronic illness isn’t a static experience; instead, it ebbs and flows, with periods of greater symptom expression — good days and bad days. These bad days often will impact the person’s personality, making them angry at others or shortening their fuse. The difficult thing is learning how to deal with their illness without unjustly taking it out on the people who are trying to help them.”

Making Sense of Life Beyond the Diagnosis

Helping patients realize that while they may not have control over their chronic illness, they do have control over the way they choose to view that illness, as well as control over their behaviors that can both negatively and positively impact their condition. This realization is crucial to long-term success. A response shift, defined as a change in internal values, is often needed in those with chronic illness. Therefore, a valuable tool for mental health professionals helping chronically ill patients is facilitating a subsequent transformation of learning and then a changing of belief systems, feelings and knowledge that reflects the new norms and values for patients about their condition, prognosis and their role in achieving long-term success.

The uncertainty of chronic illness, including how and when it will manifest, poses one of the biggest coping challenges for patients.

Says Purinton: “When the person with a chronic illness believes that there are things that they can do to improve their situation, they’re much less likely to develop learned helplessness. For example, for people with chronic pain, the intensity level of the pain and the level of distrust that it causes in the person’s life is poorly correlated. This means that a person with the pain level of 4 out of 10, which is considered to be significant in needing treatment, may not be able to work, while someone with an 8 out of 10 is able to. The difference often comes down to whether they believe they are in control of their life.”

Patients who display resilience after a diagnosis tend to have three major commonalities: memory, hope and meaning, with memory being the link between hope and meaning. Professionals can help patients develop skills that allow them to view their illness as part of life and a challenge worth facing — far healthier than viewing their illness as a debilitating diagnosis that will soon take over their lives.

A study that looked at psychological adjustments to chronic illness, published in The Lancet, identified four
specific factors that help patients have a healthier adjustment to their condition: physical activity, as much as is reasonably possible; finding a healthy way to express emotions; taking initiative in the self-management of their condition; and finding the positive with regard to their condition. All of these will provide the best chances for patients to experience a successful adjustment to their current and future challenges.4

**Anxiety and Feelings of Hopelessness**

Many challenges lie ahead for patients with a chronic illness, and the inability to face these challenges head-on can lead to anxiety, feelings of helplessness and depression. Even those with the best self-care techniques can be struck with periods of feeling blue and deep depression. Recognizing these symptoms early and seeking professional help can make all the difference. “Anxiety cannot be made manageable when families think of it in terms of ‘for the rest of their lives,’” says Purinton. “When considering the chronic illness in its entirety, it can be especially overwhelming. By taking one day at a time, the person with a chronic illness and the family are better able to mobilize their coping resources.”

Patients should be encouraged to seek counseling, even on a short-term basis, because the ability to anticipate what feelings are coming and how to best manage both the emotions and physical stress can help patients enormously. The highest risk of depressive symptoms occurs within the first two years following diagnosis.5 Once depression starts, other behaviors come into play that can further set patients back, such as the elimination of exercise, the addition of poor eating habits and even becoming less inclined to take medication.

“Dealing with a chronic illness changes as each individual and the family progress in their development, and the family as a whole progresses through the family life cycle,” says Purinton. “As a therapist, I often help families to anticipate what challenges the next stage of life will bring, as well as help them to transition and mobilize different coping resources as the needs and the circumstances change.”

**Impact on Families**

Families of patients with chronic illness are an important concern. Though patients may feel alone in their condition, they are not. Families feel the same uncertainty over the future and how the illness will impact their relationship, their financial future, the spouse’s evolving role as a caretaker and more. Finding successful stress-management and coping skills for families is crucial to the overall success for patients and their relationships. Explains Purinton: “As a profession, social work believes that the social environment that the person finds themselves in plays a large role in their symptoms. In this context, a chronic illness is a very stressful environmental stimuli that the family must adapt to.”

Families need an action plan, just as patients do, and this action plan must take into account where each member of the family is emotionally, not only relating to the illness itself but with the patients. “I look at how the family has done with periods of adversity in the past — how other members of the family have dealt with negative circumstances even if they’re not a chronic illness — to help determine the resiliency of the family system,” explains Purinton. “I’m also looking for other vulnerabilities that would make coping with a chronic illness more difficult: for example, economic difficulties, being victimized by prejudice, and a lack of social support. I’m looking for views that family members may have about chronic illness. Sometimes people have difficulty with the stigma of having a chronic illness. When each member of the family has a proactive coping posture, they’re much more resilient against the many challenges that chronic illness often brings with it.”
If you are a Hizentra patient or caregiver

Sometimes talking to someone who “gets it” is KEY.

Voice2Voice

Your key to explore Voice2Voice online

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

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

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

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

Sign up for Voice2Voice.

You can enroll online at Hizentra.com/V2V or call 1-877-355-IGIQ (4447) for assistance.

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

Important Safety Information

Hizentra treats various forms of primary immunodeficiency (PI) in patients age 2 and over.

Hizentra should not be used if you have had serious negative reactions to immune globulin (Ig) preparations or a deficiency of an Ig known as IgA. Because Hizentra contains the amino acid proline as stabilizer, patients with hyperprolinemia (too much proline in the blood) should not take Hizentra.

Infuse Hizentra under your skin only; do not inject into a blood vessel.

Allergic reactions can occur with Hizentra. If your doctor suspects you are having a bad allergic reaction or are going into shock, treatment will be discontinued. Immediately tell your doctor or go to the emergency room if you have signs of such a reaction, including hives, trouble breathing, wheezing, dizziness, or fainting.

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

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

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

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

Important Safety Information (continued)

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

The most common drug-related adverse reactions in the clinical trial for Hizentra were injection-site reactions (swelling, pain, redness, heat or itching); headache; back pain; diarrhea; tiredness; cough; rash; itching; nausea and vomiting.

Hizentra is made from components of human blood. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent, cannot be completely eliminated.

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

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

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

Reference: ¹ Data on File. Available from CSL Behring as DOF HIZ-103.
Before prescribing, please consult full prescribing information, a brief summary of which follows. Some text and references refer to full prescribing information.

1 INDICATIONS AND USAGE

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

2 CONTRAINDICATIONS

Hizentra is contraindicated in patients who have had an anaphylactic or severe systemic reaction to the administration of human immune globulin or to components of Hizentra, such as polysorbate 80. Hizentra is contraindicated in patients with hyperprolinaemia because it contains the stabilizer L-proline.

3 WARNINGS AND PRECAUTIONS

3.1 Hypersensitivity

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

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

5.2 Thrombotic Events

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

Because of the potentially increased risk of thrombosis, consider baseline assessment of known or suspected hyperviscosity, and/or those who use estrogen-containing products.

5.3 Aseptic Meningitis Syndrome (AMS)

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

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

5.4 Reactions Reported to Occur With IGIV Treatment

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

Renal Dysfunction/Failure

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

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

Hemolysis

Hizentra can contain blood group antibodies that may act as hemolysins and induce in vivo coating of red blood cells (RBCs) with immunoglobulin, causing a positive direct antiglobulin (Coombs’) test result and hemolysis. Delayed hemolytic anemia can develop subsequent to the use of globulin therapy due to enhanced RBC sequestration, and acute hemolysis, consistent with intravascular hemolysis, has been reported.

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

Monitor recipients of Hizentra for clinical signs and symptoms of hemolysis. If these are present after a Hizentra infusion, perform appropriate confirmatory laboratory testing. If transfusion is indicated for patients who develop hemolysis with clinically compromising anemia after receiving Hizentra, perform adequate cross-matching to avoid exacerbating on-going hemolysis.

5.6 Laboratory Tests

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

6 ADVERSE REACTIONS

The most common adverse reactions (ARs), observed in ≥5% of study subjects receiving Hizentra, were local reactions (e.g., swelling, redness, heat, pain, and itching at the injection site), headache, diarrhea, fatigue, back pain, nausea, vomiting, abdominal pain, heartburn, cough, flushing, urticaria, and injection site induration.

Subjects were treated with Hizentra at weekly median doses ranging from 66 to 331 mg/kg body weight (mean: 181.4 mg/kg) during the wash-in/wash-out period and from 72 to 379 mg/kg (mean: 213.2 mg/kg) during the efficacy period. The 49 subjects received a total of 2264 weekly infusions of Hizentra.

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

Table 4 summarizes the most frequent ARs (experienced by at least 2 subjects) occurring during or within 72 hours after the end of an infusion. The proportion of subjects reporting local reactions decreased over time from approximately 20% following the first infusion to <5% by the end of the study. Three subjects withdrew from the study due to ARs of mild to moderate intensity. One subject experienced injection-site pain and injection-site pruritus; the second subject experienced injection-site reaction, fatigue, and feeling cold; and the third subject experienced injection-site reaction and hypersensitivity. All reactions were judged by the investigator to be “at least possibly related” to the administration of Hizentra.

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

Hizentra
The following adverse reactions have been identified during postmarketing use of Hizentra. This list does not include reactions already reported in clinical studies with Hizentra (see Adverse Reactions [6.1]).

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

General
The following adverse reactions have been reported during postmarketing use of immune globulin products1:

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

To report SUSPECTED ADVERSE REACTIONS, contact CSL Behring Pharmacovigilance at 1-866-915-6958 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

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

<table>
<thead>
<tr>
<th>AR (≥2 Subjects)</th>
<th>Number (%) of Subjects (n=49)</th>
<th>Number (Rate) of ARs (n=2264 Infusions)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Local reactions</td>
<td>49 (100)</td>
<td>1322 (0.584)</td>
</tr>
<tr>
<td>Other ARs:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>12 (24.5)</td>
<td>32 (0.014)</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>5 (10.2)</td>
<td>6 (0.003)</td>
</tr>
<tr>
<td>Fatigue</td>
<td>4 (8.2)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Back pain</td>
<td>4 (8.2)</td>
<td>5 (0.002)</td>
</tr>
<tr>
<td>Nausea</td>
<td>4 (8.2)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Pain in extremity</td>
<td>4 (8.2)</td>
<td>6 (0.003)</td>
</tr>
<tr>
<td>Cough</td>
<td>4 (8.2)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Vomiting</td>
<td>3 (6.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Abdominal pain, upper</td>
<td>3 (6.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Migraine</td>
<td>3 (6.1)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Pain</td>
<td>3 (6.1)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
</tr>
<tr>
<td>Urticaria</td>
<td>2 (4.1)</td>
<td>3 (0.001)</td>
</tr>
</tbody>
</table>

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

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

European Study
In a clinical study conducted in Europe, the safety of Hizentra was evaluated for 10 months (ITT Population), US Study

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

* 15 to 45 minutes after the end of infusions administered at regularly scheduled visits (every 4 weeks).
† For multiple injection sites, every site was judged, but only the site with the strongest reaction was recorded.
‡ Rate of injection-site reactions per infusion.
§ Number of infusions administered regularly scheduled visits.

Table 4: Incidence of Subjects With Adverse Reactions (ARs)* (Experienced by 2 or More Subjects) and Rate per Infusion, European Study

<table>
<thead>
<tr>
<th>AR (≥2 Subjects)</th>
<th>Number (%) of Subjects (n=51)</th>
<th>Number (Rate) of ARs (n=1831 Infusions)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Local reactions</td>
<td>24 (47.1)</td>
<td>105 (0.057)</td>
</tr>
<tr>
<td>Other ARs:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>9 (17.6)</td>
<td>20 (0.011)</td>
</tr>
<tr>
<td>Rash</td>
<td>4 (7.8)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Pruritus</td>
<td>4 (7.8)</td>
<td>13 (0.007)</td>
</tr>
<tr>
<td>Fatigue</td>
<td>5 (9.9)</td>
<td>5 (0.003)</td>
</tr>
<tr>
<td>Abdominal pain, upper</td>
<td>2 (3.9)</td>
<td>3 (0.002)</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>2 (3.9)</td>
<td>2 (0.001)</td>
</tr>
<tr>
<td>Erythema</td>
<td>2 (3.9)</td>
<td>4 (0.002)</td>
</tr>
<tr>
<td>Abdominal discomfort</td>
<td>2 (3.9)</td>
<td>3 (0.002)</td>
</tr>
<tr>
<td>Back pain</td>
<td>2 (3.9)</td>
<td>2 (0.001)</td>
</tr>
<tr>
<td>Hematoma</td>
<td>2 (3.9)</td>
<td>3 (0.002)</td>
</tr>
<tr>
<td>Hypersensitivity</td>
<td>2 (3.9)</td>
<td>4 (0.002)</td>
</tr>
</tbody>
</table>

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

* Excluding infections.
† Rate of ARs per infusion.
‡ Includes injection-site reactions as well as bruising, scabbing, pain, irritation, cysts, eczema, and nodules at the injection site.
§ Number of infusions administered regularly scheduled visits.
In dealing with emotions, “the patient has a right to feel angry and frustrated, but not a right to direct it at family members who are trying to help,” says Purinton. “I tell them that it’s a process that takes time to adjust to for both the person with a chronic illness and each member of the family. I also tell family members that it’s OK to feel as though the care that the person with a chronic illness requires is a burden. And that’s not the same thing as saying that the person is a burden or that the person doesn’t want the family member to come along, even though it may mean that there are added challenges.”

Only 50 percent of patients in developed countries follow therapies prescribed by their health professionals.

Enhancing Behavior Compliance

Only 50 percent of patients in developed countries follow therapies prescribed by their health professionals. And yet, the quality of the patient-doctor relationship can be the most important factor relating to patient compliance for both psychological and physical behaviors. A look at the current state of communication between chronically ill patients and their providers showed a positive correlation for patient compliance by displaying affective behavior, or simply asking patients about their feelings and responding to them in a way that is caring and understanding and provides a proactive approach for patients. It also is important that providers try to match the communication style of patients.

Patients will be far more successful in managing their conditions when they feel their behaviors positively affect the outcome. Providers who relay the importance of external and internal responsibility, such as taking medications, exercising and other external factors that influence the disease, can guide patients to understand that they must take charge of these responsibilities.

In some instances, patients and providers have the same concerns about the patients’ condition, but an inability to effectively communicate those concerns leaves questions unanswered and both sides feeling frustrated. A collaborative care communication model allows patients and their providers to develop a protocol for the patients’ health that takes into account the patients’ priorities, as well as the doctors’ care instructions. This model will enhance compliance for patients and, in turn, enhance health outcomes. Patients and providers can use the stages of change model to develop the steps patients need to take to participate in their own caretaking.

“Good” communication, one that is understandable to patients, can enhance the end result of common goals and move patients forward in the health continuum. While physicians and patients can set those goals together, mental health professionals can assist patients in taking the next steps toward the mutually agreed-upon goals and help patients to understand their feelings, how to overcome setbacks, and how to keep their treatment a family affair with plenty of positive support and encouragement.

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

References
Individuals who are underweight can experience as many health risks as overweight individuals. Therefore, good nutrition habits that can increase weight to a healthful level are essential.

By Jill Weisenberger, MS, RD, CDE
While most people are focused on overweight and obesity and their health risks, being at the other end of the scale also is associated with medical problems. Being underweight can cause bone loss and osteoporosis, delayed wound healing, poor growth in children and infertility in women. Many underweight individuals celebrate that they are not battling the bulge and believe that their low weight gives them license to eat anything they want. This is a bad tactic for anyone to take, but especially for those with chronic diseases. Bodies rely on the proper mix of nutrients to feed the immune system, support growth, build muscle and other tissues and maintain function of every organ and body system.

What can good nutrition achieve? It can help individuals fend off an infection before it has a chance to take hold, as well as to recover more quickly. Good nutrition can help mood, sleep, mental focus and energy level. For individuals with gastrointestinal problems that cause malabsorption and nutrient losses, it’s even more important to make every bite count. The bottom line is that no matter what an individual’s weight, eating healthfully matters. Here are some simple strategies to boost calories and nutrition for healthful weight gain. Aim for 500 to 1,000 additional calories daily. If this goal is out of reach, consuming 200 or 300 extra calories daily will help with weight gain, although at a slower pace.

Sip on It
It’s a common misconception that the primary drink needs to be plain water. As healthful as water is, for someone needing to gain weight, other beverages are better choices. Replace several glasses of water with 100 percent fruit juice and nonfat or 1 percent lowfat milk. Each offers fluids for hydration and nutrients for overall health. By trading in an 8-ounce glass of water for an equal amount of grape juice, for example, an additional 150 calories will be consumed. Variety will provide better nutrition and keep taste buds happy. Here are some swaps to sip on and the extra calories provided in 8 fluid ounces.
- Orange juice: 110
- Pineapple juice: 130
- Cranberry juice: 115
- Peach nectar: 130
- Pomegranate juice: 130
- Nonfat milk: 80
- 1 percent lowfat milk: 100
- 1 percent lowfat chocolate milk: 160

Eat Bigger Portions
Increasing your portions of rice, chicken and broccoli by just a little or even doubling them can increase calorie intake without having to prepare additional food or eat an extra snack. Milk or juice can be poured into a taller glass, cereal can be eaten from a larger bowl, and bigger bananas, potatoes and other fruits and vegetables can be chosen. Even making sandwiches on thicker or denser bread or on bigger rolls counts.

The bottom line is that no matter what an individual’s weight, eating healthfully matters.

Add Healthful Fats
Don’t fear fat. Fat sometimes gets a bad rap, but it is undeserved, especially for someone trying to gain weight. Ounce for ounce, fat provides more than double the calories of protein or carbohydrates, making it ideal to slip a lot of calories into a relatively small portion of food. The key is to choose foods rich in the healthful fats and light on the saturated and trans fats. High intakes of trans fatty acids and most saturated fatty acids are linked to increased levels of LDL (bad) cholesterol and reduced insulin sensitivity. Additionally, trans fats lower HDL (good) cholesterol. Trans fats can be recognized on food labels by looking for the words “partially hydrogenated oils” in the ingredients list. These processed foods should be left on the supermarket shelves, and another brand without trans fats — or, better yet, whole, unprocessed foods — should be selected. Foods rich in saturated fats are easy to recognize by their firmness at room temperature. When bacon grease has cooled, it solidifies. Even after sitting on the counter for a few hours, butter is still firm. These are hints that bacon and butter contain a lot of saturated fat. Dairy fat and the tropical oils (coconut, palm and palm kernel) also are largely saturated. Cheese is nutrient-packed, but because of the saturated fat, small amounts should be used or reduced-fat varieties should be purchased.
Some ways to add healthful fats to a diet include:
• Slip ping avocado onto sandwiches; dicing it into salads, including chicken and tuna salads; and mashing it for a spread on bread and a dip for chips and raw veggies.
• Dipping bread into seasoned olive oil.
• Stirring pesto sauce into soups and pasta dishes.
• Using a heavy hand when cooking with good-for-you oils like olive and canola oils.
• Drizzling a little extra oil onto salads.
• Adding olives to salads, chicken and fish dishes, or simply enjoying them as a snack.
• Sprinkling on all types of nuts — sprinkling them into oatmeal, dry cereals, fruit salads and sautéed vegetables; combining them with dried fruit and dry cereals for a one-of-a-kind trail mix; and adding peanuts to chicken dishes.
• Using peanut butter and other nut butters liberally, and thinking beyond crackers, breads and English muffins: filling celery sticks with peanut butter or almond butter; mixing either of them into fruit smoothies; adding peanut butter to noodles with chile peppers and cilantro for an Asian flair; whipping up a peanut dipping sauce for chicken, beef or shrimp; and sipping on peanut soup.

**Turn Snacks Into Mini Meals**
Instead of snacking on just an apple, an apple can be diced into yogurt and topped with granola. The goal is to aim for three food groups for each snack, such as whole wheat pita bread, hummus and carrot sticks, or whole grain crackers, reduced-fat cheese and fruit. A peanut butter and banana sandwich on whole grain bread is both super easy and super fast.

**Fat sometimes gets a bad rap, but it is undeserved, especially for someone trying to gain weight.**

**Add Higher Calorie Foods**
Lower-calorie nutrient-dense foods like spinach, plums and vegetable soup shouldn’t be eliminated completely, because an array of nutrients are provided by a varied diet.

But often opting for the higher-calorie nutrient-dense choices is better. For example, corn has more calories than spinach, a banana has more calories than a plum, and black bean soup packs more calories than vegetable soup.

These calorie-rich foods offer lots of nutrition. Their approximate calories per serving sizes are listed:
- Nuts and nut butters: 190 calories per 2 tablespoons
- Avocado: 160 calories per 1/2 avocado
- Corn: 130 calories per cup
- Peas: 80 calories per cup
- Black beans, kidney beans and other dried beans: 200 calories per cup
- Bran flakes: 125 calories per cup
- Granola: 425 calories per cup
- Dried fruit: 80 to 140 calories per 1/4 cup

Corn, peas and beans are tasty additions to salads. Dried cherries, cranberries and raisins are delicious in green salads, as well as chicken salad. Baked or roasted chicken pairs well with dried apricots and prunes. For breakfast, bran flakes should be opted for over corn flakes, or two cereals could be mixed together.

**Sneak It In**
Sometimes the best additions are hidden — those ingredients that add a nutritional punch without drastically
changing the taste or texture of the original food. Examples of this are powdered milk added to fluid milk and mashed potatoes, macaroni and cheese and other casseroles. Or, one cup of powdered milk added to each quart of fluid milk and one tablespoon of powdered milk added to one cup of a casserole or other food. Additionally, pureed white beans can be whisked into soups or stews, and ground flax seed can be mixed into muffin mix and oatmeal.

Do It Any Way
Breakfast foods don’t have to be limited to the morning meal. Eggs, pancakes and waffles can be eaten any time of day. Likewise, meatloaf and mashed potatoes can be eaten for breakfast. Food should be eaten at any time, even if it’s not mealtime. And, there’s no need to conform to the typical pattern of a large dinner and small breakfast.

Drink Water at the End of a Meal
Water before or during a meal might take up precious stomach space that could otherwise be filled with nutrient-packed, calorie-rich foods.

Perk Up a Poor Appetite
Staying active, like taking a walk before mealtime and lifting weights to maintain muscle mass, can give a person a healthy appetite. Those who are not physically able to do these things should talk to a doctor about a referral to a physical therapist.

Sometimes medications take away an appetite. Physicians might be able to switch medications, prescribe a lower dose or recommend it be taken several hours from mealtime. However, any changes should first be discussed with a member of the healthcare team.

Stay Balanced
Many people feel that to gain weight healthfully, they need to eat extra protein. While some illnesses and injuries may increase an individual’s protein needs, most people do not need to focus on consuming extra protein-rich foods. Eating a balanced diet with additional calories is sufficient.

Consider Supplements
Regular foods are usually better than supplement beverages. In some circumstances, however, beverages like Boost and Ensure have a place. Carnation Instant Breakfast or other instant breakfast powder also may be helpful. Before selecting any of these, individuals should check with their physician or a registered dietitian. Additionally, protein powders should be avoided unless a member of the healthcare team has recommended a specific brand or reviewed the ingredients label carefully. Many of these products are not what they seem. A trained professional should evaluate any supplement before it is taken.

Staying active, like taking a walk before mealtime and lifting weights to maintain muscle mass, can give a person a healthy appetite.

Maintain Health Habits
Usual health habits should be maintained while trying to gain weight, including getting adequate sleep, exercising as able, not smoking, and taking time for relaxation and recreation. Those who require additional or more specific guidance than these general recommendations should make an appointment with a registered dietitian. One can be found in most areas at www.eatright.org.

JILL WEISENBERGER, MS, RD, CDE, is a registered dietitian, certified diabetes educator, nutrition and health writer, speaker, spokesperson and culinary expert based in southeast Virginia. Her website, All That’s Nutrition, can be accessed at www.allthatsnutrition.com.

Sources
Understanding Idiopathic Thrombocytopenic Purpura

By Ronale Tucker Rhodes, MS

This rare bleeding disorder often resolves on its own, but for those who develop chronic ITP, treatment and resources are available.

Try saying the words idiopathic thrombocytopenic purpura to someone and then wait for the perplexed look. The name of this disease sounds baffling enough to someone inside the medical profession, much less to a layperson who has likely never heard of a fraction of the autoimmune diseases affecting millions of people these days. So, imagine what these words sound like to someone who is told that they have the condition.

Idiopathic (immune) thrombocytopenic purpura, or ITP for short, is rare. It occurs in 50 to 150 per million people each year and affects children and adults equally.¹ According to the Platelet Disorder Support Association (PDSA), estimates of the prevalence and incidence of ITP vary since they are often based on small population samples or the review of insurance records. However, the incidence of ITP among children is approximately 4.3 to 5.3 per 100,000 per year (equally affecting boys and girls). And, since children with ITP usually recover, the prevalence of childhood ITP is about equal to the incidence. In adults, the incidence of ITP is between 1.6 and 6.6 per 100,000, and the prevalence is approximately 9.5 cases per 100,000. While more adult women than men have the disease, the gender difference disappears in people over 60 years old.²
What Is ITP?

Idiopathic means there is an unknown cause; thrombocytopenic means the blood has a lower than normal number of platelets; and purpura refers to bruises caused by bleeding from small blood vessels under the skin. In short, ITP is a bleeding disorder caused by a low number of blood cell fragments called platelets (or thrombocytes). Platelets, which are made in bone marrow along with other kinds of blood cells, stick together (clot) to seal small cuts or breaks on blood vessel walls and stop bleeding. A normal platelet count ranges between 150,000/µL and 450,000/µL of blood. With ITP, the platelet count is less than 100,000/µL. By the time significant bleeding occurs, the platelet count is less than 10,000/µL. The lower the platelet count, the greater the risk of bleeding.

Those with mild ITP may have few or no symptoms. Those who do experience symptoms may bruise easily; may have excessive bleeding following minor cuts; may have joint pain; may bleed in the urine, vomit, bowel movements or white parts of the eyes; and may bleed under the skin, which appears as tiny red or purple dots on the skin (known as petechiae). A lot of bleeding can cause hematomas, a collection of clotted or partially clotted blood under the skin that looks or feels like a lump. Many people who have ITP may get nosebleeds, have bleeding from the gums during dental work or other bleeding that is hard to stop. Women may have heavier than normal menstrual bleeding. And, in rare instances, bleeding in the brain can result, which can be life-threatening.

Researchers believe that ITP is an autoimmune disorder caused when antibodies, which typically fight infection, attack and destroy the body’s healthy platelets. What causes this is unknown. There are two types of ITP: acute and chronic. Acute ITP, the most common type, is temporary or short-term, generally lasting less than six months. This type occurs mostly in children between 2 and 4 years of age, and often occurs after a child has an infection or is sick with a virus. Chronic ITP is long-lasting, usually lasting six months or longer, and mostly affects adults, although sometimes teenagers or children develop it.

Diagnosing ITP

To diagnose ITP, doctors typically begin by excluding other possible causes of bleeding and a low platelet count, such as leukemia myelophthisic marrow infiltration, myelodysplasia, aplastic anemia and adverse drug reactions. If no other causes are found, then three other tests are typically conducted. A complete blood count (CBC) determines the number of white and red blood cells and platelets. With ITP, white and red blood cell counts are usually normal, while the platelet count is low. A blood smear confirms the number of platelets observed in a CBC. And a bone marrow examination helps to determine the cause of a low platelet count. This exam can include a bone marrow biopsy in which a sample of solid bone marrow is removed and/or a bone marrow aspiration, in which the liquid part of the marrow is removed. Many times, both procedures are performed at the same time, as both the solid and liquid samples are frequently taken from the sample place on the back of one of the hipbones via a needle or through an incision. With ITP, the bone marrow will be normal because the low platelet count is caused by the destruction of platelets in the bloodstream and spleen, rather than due to a problem with the bone marrow.

Because ITP is a diagnosis of exclusion, it is important that the patient and physician communicate well to arrive at the correct diagnosis so that the wrong disease is not treated. Patients who experience an episode of low platelets can help in this process by providing as much information as possible, such as a history of platelet count; certain food ingestions (i.e., wood ear mushrooms, quinine water and bitter melon); new prescription or nonprescription medications; vaccines; chemical exposure; other diagnoses (i.e., lymphoma, lupus, hepatitis C, HIV); recurrent stomachaches, fevers or ulcers; insect or animal bites; poison ivy exposure; travel outside the country; family history of autoimmune disease or bleeding disorders; easy bruising; frequent colds or flus; hearing problems; swelling
or aching joints; thyroid gland problems; recent stress; hospitalizations; new diets or exercise programs; excessive alcohol consumption; and periodic cycles of low platelets.8

**Treating ITP**

While there is no cure for ITP, there are a number of treatments that usually help boost platelet levels. Prednisone, administered orally, usually results in a gradual increase in platelet levels and helps strengthen the walls of veins and arteries, which helps prevent unwanted bleeding. After tapering off of prednisone after weeks or months, some people’s increase in platelet levels is permanent. However, those whose platelet levels drop as the prednisone dosage is reduced may require long-term low-dose prednisone to keep platelets at acceptable levels. The problem with prednisone is its side effects, which include water retention, mood changes, weight gain, gastrointestinal tract irritation and suppressed immune response, all of which increase in number and severity the longer prednisone is taken.9

**Today, IVIG is the drug of choice to treat severe or chronic ITP.**

Intravenous immune globulin (IVIG) was initially shown to be effective in treating ITP in 1981, when it was noted that dose administration of IVIG promoted a rapid recovery in children with ITP.10 Today, it is the drug of choice to treat severe or chronic ITP. There are five IVIG products that are FDA-approved, including CSL Behring’s Carimune NF and Privigen, Baxter Healthcare’s Gammagard S/D, Kedrion’s Gammaked and Grifols’ Gamunex-C.11 Nobody knows precisely how IG treatment works, but it is believed that it blocks platelet removal and, thus, increases the number of platelets. Side effects are typically rare and minor, but very rarely (in one-tenth of 1 percent), a severe anaphylactic reaction may occur. IVIG is used during pregnancy because of the decreased risk to the health of the mother and the baby as compared with other treatments.9

All five IVIG products were approved by the FDA as a result of pivotal clinical studies. In one study of Privigen, conducted in Europe, 57 subjects with a platelet count of less than or equal to $20 \times 10^9/L$ received 1 g/kg of Privigen twice on each of two consecutive days and were observed for 29 days. A total of 46, or 80.7 percent of subjects, responded to Privigen therapy with an increase of platelet count to greater than or equal to $50 \times 10^9/L$ within seven days after drug administration. Hemolysis occurred in eight subjects, and all cases resolved uneventfully.12

Anti-D antibodies in the Rho(D) immune globulin WinRho SDF work similarly to IVIG. Also administered intravenously, its effects are short-term (lasting about one month), and it is effective only for people who are Rh positive (up to 85 percent of people are) and have a spleen. Occasionally, anti-D antibodies result in a long-term platelet count increase. Unlike IVIG, it may not be suitable for some pregnant women.9

Splenectomy (removal of the spleen) has been used to treat ITP since the middle of the 20th century, and it works in two-thirds of ITP patients. However, doctors usually do not resort to splenectomy before trying other treatments. And, since the spleen plays an important role in cleaning the body of infection, those undergoing splenectomy are usually at a higher risk of infections. In addition, a lot of people relapse after a splenectomy and their platelet counts go down again. Some studies suggest that people age 40 and under usually fare better with splenectomy.9

Other drugs and treatments used for ITP include chemotherapy drugs, antibiotics, Danazol, Rituxan and immunosuppressant drugs.9

The American Society of Hematology (ASH) has published its 2011 Clinical Practice Guideline on the Evaluation and Management of Immune Thrombocytopenia (ITP), which was published in Blood on April 21, 2011. In addition, ASH has developed a pocket-sized quick reference guide to provide physicians with an easy reference tool for its practice guideline, which can be obtained by contacting Patrick Irelan at pirelan@hematology.org.13

**Living with ITP**

ITP patients can prevent complications by avoiding medications such as aspirin or ibuprofen that can affect platelets and increase bleeding; protecting themselves from injuries that can cause bruising or bleeding; and seeking treatment immediately for infections or symptoms of infection. Lifestyle changes also can be made to prevent complications. For instance, contact sports or other sports that can cause injury, especially head injuries, should be avoided. And certain precautions should be taken such as
WARNING: ACUTE RENAL DYSFUNCTION AND ACUTE RENAL FAILURE

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

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

- For patients at risk of renal dysfunction or failure, administer GAMMAKED at the minimum concentration available and the minimum infusion rate practicable. (see Warnings and Precautions)
**WARNINGS AND PRECAUTIONS**

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

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

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

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

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

**Rx Only**
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Research Triangle Park, NC 27709
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Fort Lee, NJ 07024
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wearing seatbelts and wearing gloves when working with knives or other tools.

If bleeding does start, ITP patients should sit or lie down and apply pressure to the wound if it can be seen. An ice pack also can slow the bleeding. If the wound is to an arm or leg, that limb should be elevated above heart level. If there is blood in the urine, increased fluids should be ingested. If there is vaginal blood, tampons should not be used; instead, the number of sanitary pads should be kept track of and clots should be watched for. A physician should be contacted if the patient has a headache, confusion or dizziness; if there is blood when coughing or difficulty breathing; if there is blood in the urine, vomit or bowel movement; and if there is unusually heavy vaginal bleeding or vaginal bleeding after menopause.  

ITP is a serious disease, but the prognosis is good.

For pregnant women with ITP, treatment is not usually needed. However, treatment is needed for those with very low platelet counts to prevent serious heavy bleeding during delivery and afterward. And, while babies of women with ITP aren’t usually affected, some babies are either born with low platelet levels or develop low platelets soon after birth. These babies’ platelet counts almost always return to normal without any treatment.

The PDSA (www.pdsa.org) has a variety of resources for ITP patients, including a 26-page booklet titled Coping with ITP and online discussion groups.

The ITP Prognosis

ITP is a serious disease, but the prognosis is good. More than 80 percent of children with untreated ITP have a spontaneous recovery with complete normal platelet counts in two to eight weeks. Fatal bleeding occurs in only 0.9 percent of children upon initial presentation of ITP, and fatal intracerebral hemorrhage occurs rarely in children with ITP who have been treated with prednisone and anti-D antibodies or IVIG for at least two days. In adults with ITP, 60 percent to 90 percent respond with an increased platelet count after treatment with prednisone, a combination of prednisone and anti-D antibodies, or IVIG. And, of those who don’t maintain an increased platelet count and who require splenectomy, approximately two-thirds have a sustained response and 10 percent to 15 percent have a partial response. 

There is no question that ITP is a more chronic, persistent disorder in adults, but almost all patients do well. With a correct diagnosis, proper treatment and use of available resources, ITP patients can live relatively normal lives.

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

References
When Laurieann Skinner was diagnosed with chronic inflammatory demyelinating polyneuropathy (CIDP), doctors immediately hospitalized her and began a five-day course of high-dose intravenous immune globulin (IVIG). In an unusual turn of events, Laurie contracted hemolytic anemia, a condition that occurs when red blood cells are prematurely destroyed or removed, sending her back into the hospital. Laurie hopes to make others aware of this rare but documented adverse reaction to IVIG.

Trudie: Tell me about your disease state.
Laurie: I have a rare, slowly progressive autoimmune neurological disorder called chronic inflammatory demyelinating polyneuropathy, or CIDP. CIDP affects the peripheral nerves and nerve roots. It is very hard to diagnose because there are no specific tests for CIDP. Diagnosis is based on symptoms, clinical exam, increased protein in spinal fluid and nerve damage seen in an electromyography (EMG).

Trudie: How were you diagnosed?
Laurie: In 2007, I was diagnosed with optic neuritis, which caused inflammation of my optic nerve and, as a result, I have vision loss in the central part of my right eye. Optic neuritis can be an indication of multiple sclerosis (MS), so I was sent to a neurologist who specializes in MS. I was given a magnetic resonance imaging (MRI) test, which came back normal, and was told by my doctor that I had a 40 percent chance of developing MS in 14 years.

As time progressed, I began to experience symptoms that mimicked MS, including numbness, tingling, pain, fatigue and problems with my balance. So I was admitted into the hospital for some tests that, again, came back normal. At my next doctor visit, it was apparent that my symptoms were getting worse. I started having terrible pain in my feet that made walking difficult. It also felt like someone was pouring hot water down my leg. That's when my doctor ordered a spinal tap and an EMG. As a result, I was referred to a neuromuscular specialist, who was certain that I had CIDP, and he began treatment right away.

Trudie: When did you start intravenous immune globulin (IVIG)?
Laurie: My first treatment was high-dose IVIG that was administered in my arm over a five-day hospital stay. The IVIG caused headaches, fever, chills and vomiting. These symptoms lessened by reducing the rate of infusion and receiving what is called a migraine cocktail.

Trudie: In an IG Living Facebook post, you said IVIG triggered a rare condition called hemolytic anemia. Tell us about that experience.

Laurieann Skinner developed hemolytic anemia, a rare side effect of high-dose IVIG therapy, after being diagnosed with CIDP.
Laurie: A few days after being released from the hospital, I began to feel very sick. I was unable to stand for long periods of time, and my heart raced uncontrollably. My urine turned brown. I called my neurologist, and he sent me to my local doctor for lab work. The next day, I was admitted to the hospital with hemolytic anemia and needed blood transfusions.

Trudie: What is hemolytic anemia?
Laurie: It is a condition in which there are not enough red blood cells in the blood due to the premature destruction of them. This is a rare but documented adverse reaction of IVIG.

Trudie: What were the risk factors?
Laurie: Apparently, I met all the risk factors for hemolytic anemia caused by IVIG because I was a female with blood type A who received high-dose infusions. Studies have shown that anyone with a non-O blood type is at higher risk for developing this complication.

Trudie: How was the condition treated?
Laurie: I was given three blood transfusions over five days. While being transfused, I asked my hematologist why I was receiving blood type O when I was blood type A. He explained that if they gave me type A, my antibodies would attack and destroy the new blood. I don’t quite understand all of what happened, but I am very blessed to have had skilled doctors who knew exactly what to do. I’ve never met anyone who experienced hemolytic anemia from IVIG, and I can’t find out a lot about it on the Internet, but I would like to make others aware of this very rare side effect of high-dose IVIG.

Trudie: How are you managing your illness now?
Laurie: IVIG was initially very helpful in improving my symptoms and my energy level. It also stopped the numbness and tingling I was experiencing in my hands and feet. Unfortunately, because of the side effects, my doctor decided it would be safer for me to try another treatment. He prescribed pulsed high-dose dexamethasone 40 mg for four days every 28 days for six cycles. I only had my first dose a few weeks ago, and so far it hasn’t been very successful. I am hopeful for better results in the future.

Trudie: How has CIDP changed your life?
Laurie: I’m not the same person I was a year ago. I look healthy, but I struggle with pain and weakness almost every day. I can’t do a lot of the same things I used to do, but I am learning to accept this disease and not let it define me. I work full time as a bookkeeper for a middle school and struggle some days with fatigue and pain, but my co-workers are very understanding and supportive, so I am very lucky.

Trudie: What advice do you have for other patients?
Laurie: I try to stay positive and laugh as much as I can because it makes me feel better. I am blessed with a devoted husband and loving son who will do anything for me. I get my strength from God and pray every day for his help and guidance.

“Hemolytic anemia is a condition in which there are not enough red blood cells in the blood due to the premature destruction of them. This is a rare but documented adverse reaction of IVIG.”

TRUDIE MITSCHANG is a staff writer for IG Living magazine.

If your life depends on immune globulin and you have a unique experience to share, we want to feature you in this column! Email us at editor@IGLiving.com.
Patient: I have been infusing Hizentra for almost two years now. I no longer have respiratory issues or arthritis, and my fibromyalgia has improved. However, I have a new issue. I have become extremely sensitive to multiple foods, supplements and medications. Doctors are skeptical that my system is changing, but it is becoming quite debilitating at times. I get diarrhea, headaches, muscle cramping and spasms. I’ve tried eliminating corn products and gluten products, and while that seems to help, it does not completely take the symptoms away.

Dr. Terry Harville: From your description, it is extremely difficult to understand what is truly occurring. Individuals with primary immune deficiency disease (PIDD) who are treated with immunoglobulin replacement may have changes in the gastrointestinal (GI) tract organisms based on the types of antibodies present in the product they are using. Further, patients who are IgA deficient may have little secretory IgA getting into the GI tract, but they may have some of the IgG from their replacement getting there, which can result in changes in the organisms present. In addition, the organisms in the GI tract of patients who have taken, or continue to take, antibiotics may be greatly altered. Therefore, your sensitivities to certain foods may be due to altered metabolism of the food products by the altered organisms that now inhabit your GI tract.

The therapeutic process that may be of benefit is to use one or more probiotic supplements. Many of these are available at drug stores and health food stores. Kefir may be of benefit and can be found in health food stores, as well as some grocery stores. I would suggest trying different brands and combinations until your symptoms improve. For instance, try any one brand for a week or so before changing. For advice on products, ask a representative at your local health food store.

The Immunodeficiency Foundation has multiple patient advocacy forums. You may try posting a question about which probiotics have been most useful at http://idffriends.org/forum. This could elicit responses from patients with PIDD as to what has worked best for them.

Patient: I was hospitalized for six days with a pulmonary infection when I caught Stenotrophomonas maltophilia, and I just can’t get better. Is this normal? My physician assistant (PA) told me to go back to the emergency room after listening to my lungs. I gurgle, bubble and rumble, and it’s hard to breathe. My pulmonologist just doesn’t seem to care, and I don’t know what to do. I’m afraid of going back into the hospital and going through the same thing. A sputum culture was performed for the second time a few days ago, so maybe something has changed. I don’t have a high temperature, but I feel horrible.

Leslie Vaughn: As you may know, it is pretty rare to have a Stenotrophomonas maltophilia infection. The infection is an aqueous-loving bacterium and tends to colonize in fluids such as intravenous fluids, irrigation solutions, etc. — all of which are found in a hospital setting. There is a difference between being colonized with S. maltophilia and actually being diagnosed with an active S. maltophilia infection. Your pulmonologist might be trying to determine if you are colonized versus infected by conducting the second sputum culture. Since the PA detected abnormal sounds in your lungs and you are having difficulty breathing, I would definitely recommend further follow-up with either your pulmonologist, immunologist or the PA again to rule out pneumonia and start appropriate treatment.

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

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Here Comes the Bride?

By Ever Fecske Mazza

BY THE TIME you read this, I will be Mrs. Ever Mazza! But, as I write this, I am 32 days away from my wedding day in fabulous Hollywood, Calif. I have meetings every other day with vendors such as photographers, DJs and the baker. In the midst of a migraine, I am writing thank-you cards, and during my infusion, I am making a seating chart; who sits next to whom can make or break me at this point.

I have a list of things to do that seems three miles long — with no end in sight. And, I’m trying desperately to keep my cool, because I know if I don’t, I risk getting sick. No one wants a snotty bride, and, believe me, the bride doesn’t want to be snotty either! I never knew I could cry from stress, and I never dreamed I could have a temper tantrum over a veil that didn’t look right. In my defense, I made it myself five different times! I never expected one of my bridesmaids to drop out of my wedding because of a guy, nor did I foresee another bridesmaid getting pregnant and not fitting into her dress! But it has happened, and I can only imagine what is next.

After seven years of being chronically ill, you would think I would know how to deal with curve balls and roller-coaster rides. I don’t think anything could have prepared me for this. But, in retrospect, nothing could have prepared me for a chronic illness either.

In the midst of coping with all the stress of wedding planning and worrying about staying well for the big day, a realization has come to me: Three years ago, my fiancé and I had to postpone this very event because we didn’t know if I would even be here. We were just engaged, and I was on 60 mg of prednisone and undergoing experimental treatment for my lung disease. There were long periods when I couldn’t even get out of bed. I could barely breathe, and I didn’t know if I was ever going to get better.

But, as my original wedding date came and went, I never gave up hope that one day I would walk down the aisle in a big white cupcake of a dress! There was excitement to make plans and create the perfect day, not stress. I surrounded myself with positivity and bridal magazines. I distracted myself, instead of worrying about survival. I dreamed about a five-tiered cake with pink sparkling icing. In reality, my body was fighting for a chance to survive, but in my mind, I was imagining walking toward my handsome man in his swanky black suit.

In 32 days, I will wake up, and my dreams will be reality. I will walk down the aisle in a white pastry-like gown toward the man who has stuck by me through sickness. We will cut our pink sparkling cake, and in that moment, all the stress and the struggle will be like a distant memory. Now I see that all I needed was a little perspective to realize how far I have come. I had to stand back from the chaos to recognize that, at this moment, I am successfully living with a chronic illness, and no seating chart, pregnant bridesmaid or ugly veil can take that away from me!

EVER FECSE MAZZA was diagnosed with CVID and interstitial lung disease in 2004. She is a fashion design student, loves spending time with her husband, family and bulldog, Dunkin, and can’t get enough of writing, cake decorating and anything that sparkles!
IG Living’s Second Annual Essay Contest: Winning Entry!

“The last time I did something for the first time, I…”: This was the leading sentence we asked IG Living readers to finish when writing their entries for our second annual essay contest. These words sparked entrants to share courageous stories about their personal journeys of living with a chronic illness. From dozens of entries, one first place and two runners-up were chosen. Featured here is the winning entry. The runners-up will be published on IG Living’s blog in September at www.IGLiving.com/blogengine.

Lessons from the Deep

THE LAST TIME I did something for the first time, I walked slowly down the ramp into the 90-degree therapy pool for my first strengthening and toning class. As I moved deeper into the water, I was amazed at how soothing it felt. I thought: Why on earth did it take me so long to get here?

It’s been two years since my chronically sore nurse’s back turned out to be something more: severe degenerative spinal disease. Three months later, it was H1N1 and then a cough that would not go away. I began a familiar routine: productive cough, feel crummy, drop off a culture, take antibiotics, feel better and then in four or five weeks, rinse and repeat.

After about six months, I was so short of breath, I couldn’t lie flat to do my back exercises. Thus began a year of increasingly exotic antibiotics and increasing doses of prednisone. I woke up one morning and wondered: “Where did that 30 pounds come from? Yikes!” Hmmm…did I want to breathe easily or get my girlish figure and low cholesterol back?

Not a month later, I was sitting in a recliner, as my homecare nurse started my IV, looking up at that little intravenous immune globulin (IVIG) bottle. I learned that MedicAlert introduced a “fashionista” collection (who knew?), but selective immunodeficiency wasn’t on its list of recognized conditions. It is now!

After a few months, my physical therapist wanted me in the pool. I talked a good fight, but for the first time in my life, I was scared to do something new. I was a crusty, old, fearless emergency room nurse, after all. Yet, there I was, afraid to get in the water. The night before registration, I thought I was on the bottom stair but I wasn’t, and I stepped into space and landed on my ankle. My husband said he’d never seen anything swell so fast (or heard me yell so loudly!). No pool for me. Six weeks later, I fell again. My physical therapist challenged me: “What is it that you don’t want to step into? Something is going on here.”

She was right. That afternoon, I took out the tape measure and put numbers to the depressing physical changes from the prednisone. I paged through the catalog, picked out swim shorts and a long swim top, and hoped I would be covered enough.

So there I was in the water. I couldn’t believe how great it felt. I looked around at the others in the pool. I was surely the youngest, but not the heaviest. Two ladies introduced themselves and welcomed me. Once class started, I marveled at the camaraderie and support. When I turned toward the ramp during a stretch, I saw four canes, two walkers and — by the bench along the wall — a wheelchair!

As we performed the cool-down balance exercises, tears rolled down my cheeks. My balance was awful, but in the water, I didn’t have to risk another fall. I was safe. I cried when I realized how much strength I’d lost and how I had been living my life in fear. I was afraid to fall, afraid of getting sick again. But those tears also were tears of relief. Getting in the pool for the first time taught me that my extra weight doesn’t matter and my limitations don’t define me. Nobody there cares about my shape. It’s clear that, like cameras and food, self-pity is not permitted in the pool area. And that’s just what I needed to learn to muster the courage and strength I’ll need the next time I do something for the first time.

PATRICIA CARROLL was a registered nurse for more than 20 years. She currently designs educational programs for medical manufacturers and teaches online for her alma mater, Excelsior College. Her immunodeficiency was diagnosed in October 2011, and since then, she has developed a fondness for zebras.
“ARE YOU SERIOUS?” I exclaimed while reading the latest newsletter from my drug company. “Can’t you come up with anything better than ‘bring a book or magazine’ to keep yourself from being bored while infusing?” After reading the other “primo” suggestions (bring your favorite snack? Duh!), I began an unrelenting yawning attack.

I like being bored on my infusion days because, frankly, I’m tired. It should be my time to indulge in a monthly nap. I can’t think of anything more boring and exhilarating at the same time than a nap. But the only time I get a good nap is under anesthesia for various surgical procedures. On the other hand, the last thing my primary immune deficiency disease (PIDD) kids, Caleb and Molly, want to do on infusion day is take naps. Instead, Caleb and Molly play video games, watch cartoons and gulp Happy Meals. My kids have even gone so far as to play art class with recycled medical equipment just so
they don’t have to rest (or do homework) on infusion days. For example, Molly likes to decoupáge with alcohol wipes, 4-by-4s, Tegaderm and leftover EMLA cream. Caleb enjoys measuring how far he can squirt water from used saline syringes (the 60 mL luer lock syringes “can really go the distance,” according to Mr. Mensa). Lock syringes “can really go the distance,” according to Mr. Mensa). Despite the multitude of activities our infusion company ships to our house once every four weeks (aka the Haggards’ “party box”), even we run out of ideas for what to do with leftover durable medical equipment. I knew my kids were beginning to feel desperate because they actually started doing their homework. Caleb and Molly were about to concede to that nap until I introduced “Susie Scooter” to them.

As I mentioned, the last nap I took was under twilight anesthesia for foot surgery. When I finally came to, I couldn’t help but notice that my right foot adorned a preordered Pepto-Bismol pink cast. Mark, my husband, was going over my discharge papers with the nurse.

“Looks like we need to stop by the pharmacy to fit you with some kind of a scooter, hon,” Mark told me while I chomped on ice chips.

“Sounthsss like a plan, thsssSam,” I slurred, dripping cold water from my bottom lip.

Now, I have been known for my outlandish hair color, bright clothes and funky accessories; it’s how I was wonderfully knit together 40-something years ago. I am the kind of person who comes into a room with my mouth wide open, talking even though people aren’t listening. Knowing I had to pick out boring, flesh-colored medical equipment to keep me upright and balanced for the next few weeks didn’t make me happy. So when the salesperson came out with a bright red scooter with a darling basket, you can imagine my delight! She (yes, SHE) was as bright as a penny (and worth quite a penny as well!). Susie Scooter and I became fast — and I do mean fast — friends.

Despite the fact Susie Scooter was designed for only one foot to be on the ground while the recovering leg/foot rested comfortably on her knee pad, I could whip through the supermarket in no time flat. I’d gather all my ingredients for dinner plus a gallon of 2 percent and still have room in Susie’s ginormous basket to grab a bag of fresh-baked chocolate chip cookies. A quick run through the self checkout and, before you could say “Wonkadoodle,” we were on our way. Susie was light-years ahead, “scientifically speaking,” from crutches and could easily sweep the Mother’s Choice Awards. Susie Scooter just might be (sniff) the Eighth Wonder of the World! Everyone loved Susie Scooter so much that when I was asked to help judge at our son Calvin’s debate tournament, I decided to charge $1 to ride Susie up and down the venue’s hallways (not a bad idea, as Susie put a small dent in Calvin’s college fund, not to mention a few holes in the walls!).

So when the most recent infusion day rolled around, I’m sure you can imagine what was asked to help keep boredom at bay: Susie Scooter.

“Awwww, come on, Mom!” my PIDD kids begged. I looked at our beloved Nurse Nancy for a little support. She waved her hands in front of her face like she was putting out a fire.

“Thanks a lot,” I mumbled at her, frustrated that our very wise homecare nurse wouldn’t make the call. Out of nowhere, the “fun mom” in me appeared and plopped herself onto my right shoulder and whispered, “It’ll be grrreeeeaaat! Besides, it’s not like they’re lugging around IV poles; those waist packs are snug around their tummies, and they won’t get hurt!”

“True that, my sistah!” I giggled.

Then, the “sensible mom” screamed in three different languages from my left shoulder: “Have you lost your mind?!”

After a very dramatic and borderline-painful pregnant pause, I had made my decision. I looked into the eyes of my beloveds and announced: “We’re gonna have to go get Dave.”

Dave is our neighbor who just happens to be a city police officer. Police officers have radar guns and if a radar gun can clock the neighborhood kids on their bikes, I had a hunch it could also clock an infusing PIDD kid racing Susie.

To this day, I don’t know what was more fun: watching Officer Dave giggle like a little girl or my PIDD kids as they took turns trying to beat their best time one-legged, with IV needles (and smiles) intact. One thing I do know: It’s gonna be pretty hard to top “The Haggard 500.” It’s also going to be tough beating the top time: Susie Scooter and I clocked in at 22 miles per hour! ❗️

CHERYL L. HAGGARD is a stay-at-home mom and has three children, two of whom have CVID. She and her husband, Mark, also operate Under the Hood Ministries at www.underthehoodministries.org.
Patients beginning immune globulin (IG) treatments often deal with countless emotions and questions. At the receiving end of these emotions and questions are the patients’ healthcare providers, whose job it is to make their transition to this life-saving therapy as smooth as possible.

Dr. Robert Hellmers is a pediatric allergist and immunologist who has been involved in intravenous IG (IVIG) therapy research since 1967, when he worked in the infectious disease department at Children’s Hospital Los Angeles. For the past 12 years, Dr. Hellmers has been infusing children and adults, answering their questions, addressing their concerns and helping them get the best treatment possible. He currently practices at Arizona Allergy Associates in Chandler, Ariz. His nurse, Jerry Hunter, RN, has been working with IG patients for the past six years.

Transitioning New Patients

Dr. Hellmers’ practice treats approximately 350 IG patients, 290 of whom are prescribed IVIG therapy and 60 percent whom are on subcutaneous IG (SCIG) therapy. About 15 percent of his patients come into his office for treatment. However, due to the high cost of treating in the office, the other 85 percent of patients are treated at hospitals or in the home environment.

When patients are new to IG therapy, their treatments are closely monitored by both Dr. Hellmers and Hunter. Dr. Hellmers prefers to see his patients every four to six months to review updated lab work to make sure that they’re not experiencing kidney issues, that their serum levels are within range and that they’re doing clinically well overall. For instance, adds Hunter, they look for whether patients are experiencing “less fatigue, fewer infections and fewer hospitalizations per year.”

Typically, once patients begin either IVIG or SCIG, “98 percent are doing very well,” says Dr. Hellmers. They have to go through fewer surgeries, they have fewer sinusitis infections and they feel like they have more energy to cope with day-to-day activities. This, says Dr. Hellmers, not only makes the patients feel better, but it makes him and the other staff members feel good as well. And, as Hunter notes, “we don’t always get that in nursing.”

Dealing with Emotions

Receiving a diagnosis is a relief for patients, but it also produces a lot of emotions, both positive and negative. According to Dr. Hellmers, most patients feel happy to find an answer to why they are so sick. They realize that “there’s hope at the end of the tunnel,” he explains. They realize that they can now receive treatment to help prevent them from getting sick as often. But, as Hunter notes, many patients fear the unknown. Dr. Hellmers and Hunter have to answer patients’ questions regarding the therapy, how much the therapy will cost, how the treatment will improve their lifestyle and how they are going to feel. Their goal, says Dr. Hellmers, is to give patients “all the options and help guide them to what’s best” for their health.

Once treatment begins, side effects, which are often a complication, can result in another series of emotions. Therefore, Dr. Hellmers and Hunter work with patients to help reduce side effects. They suggest pre-medicating with ibuprofen, Tylenol or Benadryl and drinking plenty of fluids before, during and after infusions to ward off or minimize negative reactions. They also try to minimize patients’ reactions by infusing very slowly when first starting so they can determine patients’ infusion-rate tolerance levels. Dr. Hellmers asks patients to inform him if they experience chills, fever or severe headaches the day after infusing. When this happens, he prescribes low-dose steroids that usually resolve the issues.

The stress of dealing with insurance companies is another source of emotions for patients. Part of Dr. Hellmers and his staff’s job, then, is
to guide patients in their search for insurance help. Sometimes, he refers them to IG manufacturers that have assistance programs for patients. His office also has a full-time staff member whose sole purpose is to get patients authorized for treatment. According to Hunter, “she has an amazing demeanor and calming effect” when discussing insurance coverage and out-of-pocket charges with patients or their guardians.

When asked if patients’ emotions take their toll on the staff, Dr. Hellmers explains that “from my standpoint as a physician, I want to be very objective about [my patients’] care. Obviously, you feel bad for them, but we’re offering a treatment that will alter their life” and make them feel better. Once patients begin therapy, he points out, negative emotions usually go away because they start to feel the positive effects of the treatments, even if they have to cope with mild side effects or tough financial situations.

**Kids Versus Adults**

While Dr. Hellmers and Hunter both began their careers working with children, they don’t have a preference when it comes to working with children versus adults. But, there are differences. For instance, when first starting treatment, adults are more aware, whereas children don’t initially make the connection between their illness and treatment, observes Hunter. It also can be difficult to get IVs into children. And, parents of children are anxious about the treatment. Their goal is to make the infusion process as comfortable as possible. In their office, patients have their own private rooms with Wi-Fi, cable television and a library. And, they encourage parents to bring games, such as a console game system like Nintendo, to keep their kids entertained.

**A Rewarding Job**

Getting to know patients well is a priority for Dr. Hellmers and Hunter. This includes understanding patients’ issues and their response to the things they are doing in their practice. According to Dr. Hellmers, when patients come in for infusions, it becomes a “bonding experience” because there always is time for a lot of communication.

Dr. Hellmers gives the example of one of his patients, an 86-year-old woman who has common variable immune deficiency (CVID), whom he’s been seeing since she began treatments. “She’s just a sweetheart of a lady,” he says. “She comes in all decked out and brightens your day.” When she told him that her daughter was getting sick often, he “convinced her to get her daughter checked out.” It turned out that her daughter also has CVID, as well as her grandchildren.

Working with IG patients is a rewarding experience, say Dr. Hellmers and Hunter. They have the ability to see patients transition from when they first come to the office, suffering from infections, multiple surgeries and constant visits to the ER and urgent care, to “a new lease on life because their health is so much improved,” Dr. Hellmers affirms.

**CARLA SCHICK** is a staff writer for IG Living magazine.

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**If you have a life-transitioning story brought about with the help of IG, we want to share it with our readers. Email us at editor@IGLiving.com.**
TJ USED TO really like going to school. Even though he missed a few days due to infections caused by X-linked agammaglobulinemia (XLA), his intravenous immune globulin (IVIG) infusions have kept him healthy and active in the school community. However, at the start of high school, TJ didn’t enjoy school as much, particularly PE. He had not grown as quickly as the other boys in his class, his voice was still high and he had yet to need to use a razor. And, while he waited for changes to other parts of his body, other boys had already seen those changes in themselves.

XLA had delayed the beginning of puberty for TJ. For boys, puberty begins at around 9 years of age when the brain signals the release of gonadotropin-releasing hormone (Gn-RH) into the body. The Gn-RH causes the pituitary gland to release two other hormones, luteinizing hormone (LH) and follicle-stimulating hormone (FSH). These stimulate the sex organs to produce sex hormones, estrogen in women and testosterone in men. These two hormones are responsible for sparking the development of adult male and female characteristics and the changes of puberty.

Two puberty disorders are common to both boys and girls, though. Precocious puberty, or early onset puberty, occurs when hormonal imbalances cause puberty to come early. Delayed puberty, a possible result of immunodeficiency, occurs when puberty comes later in the teen years.

Precocious Puberty

In precocious puberty, the process of puberty begins prematurely, before the age of 7 years in girls and 8 or 9 in boys. Precocious puberty may cause growth of pubic or underarm hair, acne and “mature” body odor. For girls, it may also cause breast development and the onset of menstruation; for boys, it may cause growth and development of the penis and testicles. Children also may exhibit a growth spurt. However, because their bones do not grow fully, they will ultimately be passed by their peers, and they will remain short as adults.

Psychologically, girls may become confused or embarrassed about the physical changes in their body and become moody or irritable; boys may become more aggressive and develop an inappropriate sex drive. Teasing from other children may lead to social and emotional problems such as low self-esteem, depression and substance abuse.

There are two forms of precocious puberty. In central precocious puberty, the process simply begins too soon. Some causes may be a process in the brain present at birth such as hydrocephalus, excess fluid buildup against the brain, or hamartoma, a noncancerous tumor near the brain. Other causes may be a brain or spinal cord injury or exposure to radiation. Certain diseases may cause precocious puberty such as McCune-Albright syndrome, congenital adrenal hyperplasia, which is abnormal adrenal hormone production by the adrenal glands, or hypothyroidism, in which the thyroid does not produce enough hormones.

Less common than central precocious puberty, peripheral precocious puberty occurs without the involvement of the hormone Gn-RH. This is caused by the release of testosterone

Parenting:
Helping Kids Deal with Delayed Puberty

Immune deficiencies can cause delayed puberty in children, but medicine can help and, more importantly, parents can help their kids get through that trying time.

By Mark T. Haggard
or estrogen into the body because of problems with sex organs. Peripheral precocious puberty in both boys and girls may be caused by a tumor in the adrenal glands that secretes estrogen or testosterone, or, more commonly, exposure to external sources of estrogen or testosterone such as creams, ointments or chemicals from the environment. Ovarian cysts or ovarian tumors may start puberty in girls; in boys, early puberty may be the result of a tumor in the cells that make sperm (germ cells) or testosterone (Leydig cells), or by a rare gene mutation called “familial gonadotropin-independent sexual precocity,” which causes early testosterone production in boys between the ages of 1 and 4. Girls are at higher risk than boys for precocious puberty. Because fat is an important catalyst for puberty, those who are overweight are at risk. Fat acts as a storage site for sex hormones, so that those obtained from medications or environmental sources may be kept at higher levels for longer periods of time, and increase the likelihood of early pubertal changes. To help reduce the chances of early onset puberty, parents should keep children away from external sources of estrogen or testosterone. They also should help children maintain a healthy diet and weight. To treat early puberty, doctors may prescribe medications such as LHRH analog, a synthetic hormone that suppresses the pituitary hormones; girls may be prescribed drugs that block estrogen production, such as testolactone.

**Delayed Puberty**

As opposed to precocious puberty, delayed puberty occurs when the process starts later in life. Most of the time, delayed puberty is a “constitutional delay,” which means the child might simply be a “late bloomer.” This is usually genetically inherited from the family. Puberty also may come late as a result of a medical condition. Immune deficiencies are specifically linked to delayed onset puberty: e.g., XLA with growth hormone deficiency and x-linked severe combined immunodeficiency with reduced insulin-like growth factor activity (these male children are not very responsive to growth hormone). Children with other immune deficiencies may find puberty delayed as well, because sickness makes it harder for their bodies to grow and develop. Proper treatment and control over these conditions can make delayed puberty less likely. For example, girls who are extremely active may not start puberty because they are so lean and their bodies require a certain amount of fat to start puberty.

There are two different conditions that cause delayed puberty. In hypergonadotropic hypogonadism, which can result from frequent infections, autoimmune problems or as a side effect of certain medications, the brain senses that the ovaries or the testicles are not making puberty hormones and sends repeated signals to the sex organs to try to get them to work. The result is that the LH and FSH are high, but estrogen or testosterone levels are low. In hypogonadotropic hypogonadism, which may result from chronic illness, the brain does not make the hormones necessary to start puberty. The result is that both LH and FSH are low, while estrogen and testosterone also are low. The sex organs have the ability to work normally, but they do not receive the messages from the brain to begin making puberty hormones and, thus, delay puberty.

**Helping Children with Puberty Issues**

For children who are considerably behind their peers, a pediatric endocrinologist may offer hormone therapy in an effort to try to “jump start” puberty. This course may be enough to get puberty hormones going. “Puberty is an arduous process for adolescents when normal, but it is more difficult in children with aberrant puberty,” says Dr. Richard D. Blondell of the University of Louisville. “Many physical and biochemical problems associated with disorders of puberty may be successfully treated. These children benefit from management by a knowledgeable and sensitive clinician.”

Beyond medical help, Dr. Blondell suggests that “psychotherapy can
LifeStyle

play an important role in assisting these patients as they develop physically and emotionally.” Steven Dowshen, pediatric endocrinologist at the du Pont Hospital for Children in Wilmington, Del., tells children: “If you’re feeling depressed or having school or other problems related to delays in your growth and development, talk to your mom or dad, your doctor or another trusted adult about finding a counselor or therapist you can talk to. This person can help you sort out your feelings and suggest ways to cope with them.” Randall Phelps, MD, of the University of Michigan, suggests that children need to be reminded that all people go through changes — some go through them earlier and some go through them later. He adds that parents need to give children openings to discuss any worries or concerns, and they should encourage kids to take part in regular physical and social activities.

TJ has now graduated from high school. He really enjoyed his senior year, and he is shaving. His parents listened to him, supported him through this difficult time in life and helped him become a better adult, regardless of his XLA and his late start to adult life. Parents who are there for their children and are willing to listen may be enough to settle the frustration in their children’s hearts. If not, there are doctors willing to help. The connections between delayed puberty and XLA are clear; the connections between early or late puberty and other immune deficiencies are not. Nevertheless, parents actively listening to their children can see them through a tough time in becoming an adult.

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Caring for people with hemophilia around the world—one at a time.
Your Medical Mind
Authors: Jerome Groopman, MD, and Pamela Hartzband, MD
Publishers: The Penguin Press, booksellers.penguin.com

Drs. Groopman and Hartzband show patients how to chart a clear path to make the best medical decisions and cut through the confusion caused by the healthcare system, the media and gaps in individuals’ reasoning. They interview patients who have struggled with medical decisions and provide reviews of research and insight from doctors, psychologists, economists and others to help reveal the array of forces that can aid or impede patients’ decisions. In addition, they show the subtle strategies drug advertisers use to influence choices; they unveil the extreme, and sometimes dangerously misleading, power of both narratives and statistics; and they help patients to understand how to improve upon a universal human shortcoming: assessing the future impact of the decisions they make. Dr. Groopman, a New Yorker writer and bestselling author, is an oncologist who guides patients through life-or-death decisions. Dr. Hartzband is a noted endocrinologist and educator at Harvard Medical School who helps patients make critical decisions about their long-term health.

The Patient’s Checklist: 10 Simple Hospital Checklists to Keep You Safe, Sane & Organized
Author: Elizabeth Bailey
Publisher: Sterling Publishing, www.sterlingpublishing.com

This book provides guidance to healthcare consumers and family members on how to go about the process of ensuring they are getting the best and safest hospital care possible. The basic checklists help patients without access to a personal health record stay organized. The 10 checklists include: Before You Go (tips to be better informed, prepared and proactive); What to Bring (both practical and personal items); During Your Stay (tips to increase comfort and to partner more effectively with the healthcare team); Master Medication List (a user-friendly log to track all prescribed medications during the hospital stay); Daily Medication Log (a daily log to help ensure that for any medication given, it’s the right drug, the right dose, at the right time — and that it is meant for the patient); Daily Journal (a brief but important way to chronicle each day of hospitalization); Discharge Plan (tips to begin planning for discharge in anticipation of the complexities of home care); Insurance (tips to navigate the overly complicated insurance system); Doctor Contacts (how to keep an organized list of all doctors); and Family & Friends Contacts List (a way to consolidate contact information for family and friends so everyone can help in the best ways possible). For administrators, the book also includes stories of how errors, oversights and miscommunication affected patients and their families.

Andy & Sofia: Stem Cells, Scientific Miracles and One Fit Savior
Author: Andrés Trevino
Publisher: CreateSpace, www.createspace.com

Written by a father, this story is about a couple who embarked on a desperate quest. They needed to create a baby with the right genetic profile, whose umbilical cord blood could save their 2-year-old son, Andy, who was dying from a rare genetic disease. The baby also would have to be free of the inherited disease killing their son. After three years and five in vitro fertilization (IVF) cycles, and with the critical intervention of a novel medical technology called pre-implantation genetic diagnosis (PGD), their daughter, Sofia, was born. Her cord blood stem cells, transplanted into her brother via blood transfusion, replaced his faulty immune system and saved his life. Today, Andy, 11, and Sofia, 6, are both healthy, beautiful children. In gratitude, Andrés and his wife, Paulina, donated the remaining frozen IVF embryos — the ones that could never be used because they carried the flawed gene causing the disease — to the Stem Cell Research Program at Children’s Hospital Boston, where scientists there were able to develop two new stem cell lines whose unique genetic properties will help researchers learn how Andy’s disease, and many others, develop. It is hoped that from that knowledge will come treatments and cures.
HOW DOES ONE know if they are getting enough sleep and, if not, how can sleep deprivation be treated? Many factors contribute to sleeplessness, including anxiety, stress, depression and pain. To determine whether one is getting adequate sleep, it is important to understand the average sleep requirement, as well as the many health factors that determine the amount of sleep hours we need in a day. If sleep deprivation is evident, treatment programs are available.

How Much Sleep Is Enough?

Infants require 16 hours of sleep in a day, teenagers require nine hours per day, and most adults need seven to eight hours, although some require as few as five hours or as many as 10 hours. Pregnant women in their first trimester of pregnancy require several hours of additional sleep.

The best methods to treat sleep disorders in the chronically ill involve adherence to treatment programs to reduce symptoms and pain management.

Experts say that feeling sleepy during the day, even during boring activities, is a sign of not getting enough sleep. People maintain a “sleep bank.” Often, sleepless nights accumulate withdrawals to an individual’s sleep bank. When the sleep bank account is debited and not replenished, an individual may start to experience symptoms of sleep deprivation, including memory loss, depression, increased sensitivity to pain and a weakening of immune defenses.

Sleep deprivation can be quite dangerous. Sleep-deprived individuals tested in a driving simulator had hand-eye coordination activities as bad or worse than intoxicated individuals. The National Sleep Foundation states that if one has trouble keeping their eyes focused, can’t stop yawning or can’t recall driving the last few miles, they are too drowsy to drive.

Chronic illness plays a key role in sleep deprivation. Often pain and depression associated with chronic illness compounds the inability to sleep and creates a vicious cycle. Neurological conditions such as Parkinson’s disease and Alzheimer’s disease contribute to an inability to sleep.

Treating Sleep Deprivation

The best methods to treat sleep disorders in the chronically ill involve adherence to treatment programs to reduce symptoms and pain management. At times, pain medication may be indicated to help an individual fall asleep.

Good sleep hygiene is critical to a peaceful night. Sleep hygiene refers to practices that an individual can follow to ensure a peaceful and restful night. These practices include sticking to a regular bedtime schedule, avoiding napping during the day, sleeping each night in a noise-free bedroom that is dark, controlling the bedroom temperature so that it is comfortable, avoiding eating or drinking foods with caffeine, and avoiding alcohol consumption and nicotine. A psychologist specializing in sleep disorders may recommend biofeedback or relaxation training.

It is recommended that an individual with sleep deprivation try non-drug-related therapies first. If non-drug-related treatments are not successful, an individual should see a physician. A physician will first try to determine the cause of insomnia or the inability to fall asleep or stay asleep during the nighttime. He or she also will evaluate an individual’s state of physical or emotional health to help guide treatment. A sleep diary, in which periods of time when sleep deprivation occurs, may be suggested. And, drug therapy may be prescribed, including those to assist in aiding sleep and treating depression.

Adequate Rest is a Must

Regardless of the cause of sleep deprivation, it is essential that an individual gets the necessary treatment needed to ensure he or she gets adequate rest. This is especially true for the chronically ill whose conditions will likely be exacerbated by a lack of sleep.

NANCY CREADON, RN, is vice president of VaxAmerica, a subsidiary of FFF Enterprises Inc.
Sleep Innovations  
Novaform gel memory foam mattresses and toppers are designed to provide pressure point relief and spinal alignment, more support at higher compression levels, and increased air circulation with gel beads that attract and distribute heat for a cooler night’s sleep. The mattresses measure 12 inches and the toppers measure 2.5 inches.  
www.novaformgel.com

Helpguide  
Helpguide provides resources about healthy sleep habits to help individuals put a stop to nighttime problems and improve their quality of rest. A downloadable Weekly Sleep Diary is available for individuals to record their daily activities and pre-sleep ritual, including information about exercise, naps, alcohol and caffeine, feelings, food and drink, medications or sleep aids, bedtime routine and bedtime.  
www.helpguide.org/life/pdfs/sleep_diary.pdf

Contour  
The memory foam leg pillow is designed to support and align the lower body, relieving pressure on the lower back, hips and knees while sleeping. The pillow fits the curves of an individual’s legs, so it comfortably stays with them throughout the night. It is made of 100 percent visco-elastic memory foam, it has a soft removable poly-cotton velour cover, and it is available in three colors.  
(800) 950-0230; www.contourliving.com/p-22-contour-memory-foam-leg-pillow.aspx

Tempur-Pedic  
The Tempur-Pedic contoured pillow collection includes the Tempur-Neck Pillow designed to support the curve created by the head, neck and shoulders; the Tempur-Side Pillow designed for side sleepers; the Tempur-Classic Pillow, which combines an ergonomic contour with a standard pillow shape; the Temper-Curve Pillow, which has a curved edge to support the head; and the Tempur-Symphony Pillow with a dual-sided design that provides a gently arched side for back sleepers and a more traditional pillow side for side sleepers. The company also makes filled pillows and travel pillows.  
(888) 811-5053;  
www.tempurpedic.com/tempur-pedic/ Pillows.asp

CPAP.com  
CPAP.com offers a large selection of CPAP masks, including nasal pillow, nasal and full face masks. Nasal pillow masks feed air directly through the nasal passages and are designed for individuals who easily become claustrophobic. They are triangular in shape and cover the area from the bridge of the nose down to the upper lip. Full-face masks also extend from the bridge of the nose, but go down to the chin, making them an option for people who breathe through their mouth while sleeping.  
(800) 356-5221;  
www.cpap.com
Sources

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

General Resources

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

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

IG Manufacturer Websites

- Baxter: www.baxter.com
- Bio Products Laboratory: www.gammaplex.com
- CSL Behring: www.cslbehring.com
- Grifols: www.grifolsusa.com
- Kedrion: www.kedrionusa.com
- Octapharma: www.octapharma.com

Disease-State Resources

Ataxia Telangiectasia (A-T)

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

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

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

Evans Syndrome

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

Guillain-Barré Syndrome (GBS)

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

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

Idiopathic Thrombocytopenic Purpura (ITP)

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

Kawasaki Disease

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

Mitochondrial Disease

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

**WEBSITES**
- Neuromuscular Disease Center at Washington University: neuromuscular.wustl.edu
- 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
- Genetic Alliance: www.geneticalliance.org

**ONLINE PEER SUPPORT**

**Myositis**

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

**Peripheral Neuropathy (PN)**

**WEBSITES**
- Neuropathy Action Foundation: www.neuropathyaction.org
- Calvary Neuropathy Association: www.calvaryneuropathy.com
- Texas Chapter of the Neuropathy Association: www.handsfeetheart.org

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

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

**Pemphigus and Pemphigoid**

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

**Primary Immune Deficiency Disease (PIDD)**

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

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

ONLINE PEER SUPPORT
- International Scleroderma Network: www.sclero.org/support/forums/a-to-z.html

Stiff Person Syndrome (SPS)

WEBSITES
- American Autoimmune Related Diseases Association Inc.: www.aarda.org
- Genetic Alliance: www.geneticalliance.org
- Living with Stiff Person Syndrome (personal account): www.livingwithspss.com
- Stiff Person Syndrome: www.stiffpersons syndrome.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.jhtml?exp=4 This federal government website offers a parents section titled “My Child’s Special Needs.”
- IVIG/SCIG Gammagard Liquid: www.gammagardliquid.com
- IVIG Gammagard S/D: www.baxter.com/patients_and_caregivers/products/gammagard_sd_5.html
- IVIG/SCIG Gammaked: www.gammaked.com
- IVIG Gammaplex: www.gammaplex.com
- IVIG Privigen: www.privigen.com
- SCIG Hizentra: www.hizentra.com

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

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

FOOD ALLERGIES
- Influenza and the influenza vaccine: www.cdc.gov/flu or call (800) CDC-INFO: (800) 232-4636
- IVIG Flebogamma 5% DIF and 10% DIF: www.grifols.com/portal/en/US/bioscience/?div=8883

PUMP AND INFUSION SETS WEBSITES
- Norfolk Medical: www.norfolkmedical.com
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