Economics of Illness
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Proven efficacy in 3 FDA-approved indications

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All immune globulin (IG) formulations are not the same. Consider your patient's risk factors when choosing an IG therapy.

### Important Safety Information

**GAMUNEX®-C** (immune globulin injection [human], 10% caprylate/chromatography purified) is indicated for the treatment of primary humoral immunodeficiency disease (PIDD), idiopathic thrombocytopenic purpura (ITP), and chronic inflammatory demyelinating polyneuropathy (CIDP).

**Thrombosis** may occur with immune globulin products, including GAMUNEX-C. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling central vascular catheters, hyperviscosity, and cardiovascular risk factors. Thrombosis may occur in the absence of known risk factors. For patients at risk of thrombosis, administer GAMUNEX-C at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.

**Renal dysfunction**, acute renal failure, osmotic nephrosis, and death may occur with immune globulin intravenous (IVIG) products in predisposed patients. Patients predisposed to renal dysfunction include those with any degree of preexisting 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 IVIG 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 patients who have had an anaphylactic or severe systemic reaction to the administration of human immune globulin. It is contraindicated in IgA-deficient patients with antibodies against IgA and history of hypersensitivity.

Severe hypersensitivity reactions may occur with IVIG products, including GAMUNEX-C. In case of hypersensitivity, discontinue GAMUNEX-C infusion immediately and institute appropriate treatment.

Monitor renal function, including blood urea nitrogen (BUN), serum creatinine, and urine output in patients at risk of developing acute renal failure. Hyperproteinemia, increased serum viscosity, and hyponatremia may occur in patients receiving IVIG treatment, including GAMUNEX-C.

There have been reports of noncardiogenic pulmonary edema (transfusion-related acute lung injury [TRALI]), hemolytic anemia, and aseptic meningitis in patients administered with IVIG, including GAMUNEX-C.

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 GAMUNEX-C is made from human blood, it may carry a risk of transmitting infectious agents, eg, viruses, and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.

Do not administer GAMUNEX-C subcutaneously in patients with ITP because of the risk of hematoma formation.

Periodic monitoring of renal function and urine output is particularly important in patients judged to be at increased risk of developing acute renal failure. Assess renal function, including measurement of BUN and serum creatinine, before the initial infusion of GAMUNEX-C and at appropriate intervals thereafter. Consider baseline assessment of blood viscosity in patients at risk for hyperviscosity, including those with cryoglobulins, fasting chylomicronemia/markedly high triacylglycerols (triglycerides), or monoclonal gammopathies, because of the potentially increased risk of thrombosis.

If signs and/or symptoms of hemolysis are present after an infusion of GAMUNEX-C, perform appropriate laboratory testing for confirmation.

If TRALI is suspected, perform appropriate tests for the presence of antineutrophil antibodies and anti-HLA antibodies in both the product and patient's serum.

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 PIDD) and infusion-site reactions, headache, fatigue, arthralgia and pyrexia with subcutaneous use (in ITP); and headache, vomiting, fever, nausea, back pain, and rash (in ITP).

The most serious adverse reactions in clinical studies were pulmonary embolism (PE) in 1 subject with a history of PE (in CIDP), an exacerbation of autoimmune pure red cell aplasia in 1 subject (in PIDD), and myocarditis in 1 subject that occurred 50 days post-study drug infusion and was not considered drug related (in ITP).

**Please see brief summary of GAMUNEX-C full Prescribing Information on adjacent page.**

**Reference:** 1. GAMUNEX®-C (immune globulin injection [human], 10% caprylate/chromatography purified) Prescribing Information. Grifols; 2013.
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: THROMBOSIS, RENAL DYSFUNCTION and ACUTE RENAL FAILURE
See full prescribing information for complete boxed warning.

- Thrombosis may occur with immune globulin products, including GAMUNEX-C. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.

-For patients at risk of thrombosis, administer GAMUNEX-C at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.

- Renal dysfunction, acute renal failure, osmotic nephrosis, and death may occur with immune globulin intravenous (IGIV) products in predisposed patients.

- Renal dysfunction and acute renal failure occur more commonly in patients receiving IGIV products containing sucrose. GAMUNEX-C does not contain sucrose.

- For patients at risk of renal dysfunction or failure, administer GAMUNEX-C at the minimum concentration available and the minimum infusion rate practicable.

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

GAMUNEX-C is an immune globulin injection (human) 10% liquid indicated for treatment of:

- Primary Humoral Immunodeficiency (PI)
- Idiopathic Thrombocytopenic Purpura (ITP)
- Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

------------------------CONTRAINDICATIONS------------------------

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

------------------------WARNINGS AND PRECAUTIONS------------------------

- IgA deficient patients with antibodies against IgA are at greater risk of developing severe hypersensitivity and anaphylactic reactions. Have epinephrine available immediately to treat any acute severe hypersensitivity reactions.
- Monitor renal function, including blood urea nitrogen, serum creatinine, and urine output in patients at risk of developing acute renal failure.
- Hyperproteinemia, with resultant changes in serum viscosity and electrolyte imbalances may occur in patients receiving IGIV therapy.
- Thrombosis has occurred in patients receiving IGIV therapy. Monitor patients with known risk factors for thrombosis; 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.
- 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.
- Passive transfer of antibodies may confound serologic testing.

------------------------------ADVERSE REACTIONS------------------------------

Serious adverse reactions which occurred in the clinical trials were an exacerbation of autoimmune pure red cell aplasia in one subject and pulmonary embolism in one subject with a history of PE. The most common adverse reactions observed in ≥ 5% patients were:

PI: Intravenous: Headache, cough, injection site reaction, nausea, pharyngitis and urticaria.
- Subcutaneous: Infusion site reactions, headache, fatigue, arthralgia and pyrexia.

ITP: Headache, vomiting, fever, nausea, back pain and rash.

CIDP: Headache, fever, chills, hypertension, rash, nausea and asthenia.

To report SUSPECTED ADVERSE REACTIONS, contact Grifols Therapeutics 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.

------------------------------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.

GRIFOLS
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Research Triangle Park, NC 27709 USA
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Dona Darr
Mother of a Daughter with IgG Subclass Deficiency

IG Chronicles: Friendship: The Best Things in Life
“My daughter’s relationship with her friend can teach people many things.”

Terry O. Harville, MD, PhD
Medical Director, Special Immunology Laboratory, University of Arkansas for Medical Sciences

Diagnosing an Antibody Deficiency: Case 5, Part 3
“Some children take longer than others for their immune systems to fully mature, and become as active as the normal adult level.”

Annaben Kazemi
IG Living Patient Advocate

Working While Chronically Ill: Is it Possible?
“Finding a work environment that is accepting and conducive to the demands placed upon individuals with chronic illness is challenging.”

Medical Data Storage for Patients
“Medical imaging technology has improved, providing a less burdensome way of storing all that information.”

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IG Living isn’t just a magazine; it’s an interactive community of people with interesting stories. And, we want you to be a part of it!

Our blog, which has a new entry posted each Friday, provides interesting perspectives on topics such as living with IG, people to be admired, support and encouragement, current news of interest and things you just need to know. We encourage you to comment with your opinions and stories. And, better yet, if you have a blog you’d like to post, send it our way!

IG Living’s Facebook page has hundreds of fans who respond to our questions that are posted each Monday through Friday. Together, these fans share their life stories and thoughts. What’s more, they are making a connection with one another that otherwise wouldn’t be possible.

So, be a part of it now at www.igliving.com/blogengine and www.facebook.com/IGLivingMagazine.

Connect with Other IG Living Readers through Monthly Teleconferences!

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

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

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

Sign up for the Teleconferences now by emailing akazemi@IGLiving.com or calling (800) 843-7477, ext. 1366.
The Evolving Role of Immune Globulin Therapy

Despite the availability of immune globulin (IG) since the 1950s, how the drug is administered continues to evolve. This has certainly been the case during IG Living magazine’s tenure that began eight years ago with the goal of educating patients and their caregivers about this lifesaving therapy, as well as helping to improve patients’ lifestyles and to facilitate communication among those in the IG community. During these years, IG Living has expanded its coverage about the therapeutic aspects of IG for primary immunodeficiency diseases (PIDDs), as well as a host of other disease states for which there is a lack of standards pertaining to how it is prescribed.

The evolution of IG treatment begins with its use as intramuscular injections for the prevention of infectious diseases such as measles, hepatitis and polio. Then came its widespread use in the 1950s as replacement therapy for PIDDs. But it wasn’t until the 1960s that the first intravenous preparation of IG (IVIG) became available. Even so, IVIG was not a U.S. Food and Drug Administration-approved therapy until the 1980s, at which time, IVIG was also found beneficial for treating autoimmune idiopathic thrombocytopenic purpura. Since that time, more and more autoimmune and inflammatory diseases continue to join the ranks of accepted IG-treatable indications, many of which are prescribed IG as only an exploratory and secondary line of treatment.

In this issue, we look at one of those indications, chronic regional pain syndrome (CRPS), an autoimmune disease that causes prolonged and intense pain that can often be constant and for which IG is showing great promise as an emerging treatment. In recent years, there have been reported cases of CRPS patients who were subsequently diagnosed with common variable immune disease (CVID) — not surprising with the increased prevalence of autoimmune diseases in PIDD patients. This was what happened to Deborah Norris, who is featured in our article “Diagnosing and Treating Complex Regional Pain Syndrome.” While first diagnosed with CRPS, once Deborah was diagnosed with CVID and treated with IG, she discovered that it lessened her chronic pain.

Both PIDDs and autoimmune diseases can now be treated with an increasing number of IG products. Today, there are 11 IG products in the marketplace, which allows doctors the benefit of matching the best-suited product to each patient. What’s not so beneficial is that making the best match is sometimes not an easy feat because while all products have comparable efficacy, they are not pharmaceutically equivalent. How physicians determine which IG product to prescribe depends upon a number of patient and product considerations, which is discussed in our article “Immune Globulin: Each Product Is Unique.” Yet, while an understanding of these factors helps in the prescribing process, a successful match may often result only from trial and error, as well as from manipulating infusion rates and changing how the drug is administered.

I hope you benefit from the information presented and enjoy the many other articles in this eighth-anniversary edition of IG Living.

Ronale Tucker Rhodes, MS
Ask the Experts

**Reader:** Are there new guidelines for calculating the dosing of intravenous immune globulin (IVIG) infusions? I was told the new guidelines calculate dosing based on ideal body weight rather than actual body weight.

**Leslie:** Several countries (Canada and Australia) and some academic centers in the U.S. have established a protocol of using adjusted body weight for calculating IVIG dosing. Adjusted body weight is between actual body weight and ideal body weight. It is calculated like this: adjusted body weight = ideal body weight + 0.4 (actual body weight − ideal body weight).

The theory behind using adjusted body weight is as follows: Typically, weight-based drugs are dosed on actual body weight when they are a lipid (fat)-soluble drug. IVIG is not a lipid-soluble drug, so it doesn’t distribute into the fatty (adipose) tissue. Patients who are obese have a greater volume of body fluids than those who are at an ideal body weight. In pharmacokinetic terms, they would be said to have a larger volume of distribution. Using adjusted body weight accounts for the increased volume of distribution found in someone who is obese.

Currently, we are not aware of any clinical studies underway that evaluate the efficacy of treating conditions with dosing based on ideal, adjusted or actual body weight. Dosing based on adjusted body weight is a physician or institution preference at this time, rather than a published guideline or practice parameter.

**Reader:** Does intravenous immune globulin (IVIG) cause anemia?

**Leslie:** Hemolytic anemia is a very rare, but known, side effect of IVIG. Review of the literature shows the development of hemolytic anemia is limited to individuals with non-O-blood types. IVIG contains many different types of antibodies, including anti-A/B antibodies that may cause the development of hemolytic anemia. Each lot number of IVIG may have different levels of anti-A/B antibodies. Lot numbers with lower levels of anti-A/B antibodies may be less likely to cause problems. If you have a non-O-blood type, talk with your physician about whether or not you should develop a monitoring plan for anemia following infusions.

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**Reader:** I am considering switching my daughter from intravenous immune globulin (IVIG) infusions to subcutaneous IG (SCIG) infusions. I am hoping SCIG will reduce my daughter’s side effects from the infusions, and that the IG’s effects will last longer. Is that the case?

**Leslie:** SCIG may be a great alternative for your daughter and may address both her side effects and the wearing-off effect she experiences just prior to infusion.

In the studies conducted for U.S. Food and Drug Administration approval of SCIG, side effects such as headache, fatigue, body aches, etc., occurred less frequently than when receiving IVIG, although they can still occur. The main side effects from SCIG are typically related to the injection site and may include swelling, redness, pain or itching.

The wearing-off effect is related to the half-life of IVIG. Most IVIG products have a half-life in the 21-day to 28-day range, so at the four-week mark, half of the infusion your daughter has received is gone. The goal of SCIG is to infuse smaller doses more frequently to allow her to have a steady state of IG levels over time. The most common dosing protocol to get to the steady-state level quickly would be for her to receive her regular dose of IVIG and then start the smaller doses of SCIG a week later.

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**Have a question?**

Email us at editor@IGLiving.com. Your information will remain confidential unless permission is given.

**LESLIE J. VAUGHAN,** RPh, is senior vice president of clinical services at NuFACTOR Specialty Pharmacy.
FOR THE PAST two issues, we have been discussing a 7-and-a-half-year-old girl who is being evaluated for a possible immunodeficiency. Her main problem is chronic sinusitis that has not improved with antibiotic treatment, and she has a family history that would suggest she might have an antibody deficiency.

The most critical testing procedure for an antibody deficiency is a pre-/post-immunization comparison of antibody titers generated to pneumococcal polysaccharide vaccine. Blood was obtained at the girl’s initial visit to test for pre-immunization titers. She was then immunized with the pneumococcal vaccine, and in four weeks, post-immunization titers were obtained (see table). Fortunately, the functional antibody study revealed a normal ability to respond to vaccination with good specific antibody production.

In the table, titer values are in units of microgram/milliliter (μg/mL). By definition, her titer values represent a normal antibody immune response, with all of the values in the protective range post-vaccination. Yet, while there has been no significant increase in the titer values post-
immunization, this is largely because the values were so good to begin with that we are not seeing the normal increase in titer values. However, in this situation, lack of increase should not be construed as poor immune response.

This is a case that may be more typical for many children with recurrent respiratory symptoms. It is common for people to believe that many children with these symptoms will outgrow them, especially around the age of puberty. But, as this case illustrates, not all children mature at the same rate, whether it is physical, mental, social or their immune systems. Some children take longer than others for their immune systems to fully mature, and become as active as the normal adult level. Further, this case illustrates that common things are common; it’s most likely that allergic disease would result in respiratory symptoms in many children, but if the possibility of an immune problem is not diligently considered and tested appropriately, disease will be missed and not treated.

We will continue with more cases next time.

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

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

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Immune Globulin: Each Product Is Unique

While all IG products are comparably effective, they also have relevant differences that determine their tolerability by patients.

By Ronale Tucker Rhodes, MS

THE CURRENT AVAILABILITY of multiple immune globulin (IG) products gives providers many choices when prescribing this lifesaving therapy. With the approval of the latest IG product in December 2012, there are now 11 IG products that treat six U.S. Food and Drug Administration (FDA)-approved diseases (primary immunodeficiency disease, idiopathic thrombocytopenic purpura, multifocal motor neuropathy, chronic lymphocytic leukemia, Kawasaki disease and chronic inflammatory demyelinating polyneuropathy), as well as a host of non-FDA-approved conditions.

The benefit of product choice, of course, is that it allows providers to match the best-suited product to the patient. And, this is extremely important because, while all products contain IgG (the most common protein in the body that helps ward off infections) and they all have comparable efficacy, they are not pharmaceutically equivalent. There are relevant differences between the current products on the market, considered third and fourth generation, that have evolved in terms of composition, resulting in decreased risk of infusion-related reactions and other adverse events. Product variations in sodium content, stabilizers, osmolality, IgA content, concentration and pH can affect the tolerability of a product for one patient versus another, based on both clinical conditions and comorbidities.1-2

Adverse effects of IG therapy have been greatly reduced in the last two decades; however, with the new generation of products, there are other serious adverse events that have been observed, including acute renal failure, aseptic meningitis, hemolysis and thrombosis. Some of these events can be attributed to either the size of the dose administered for specific indications, the rate of infusion, the differences between IG products or to characteristics of the patient receiving the treatment.3

When choosing an IG product based on the differences between each, the key factors a clinician considers are the patient’s body type, weight, conditions presenting in addition to the one being treated with IG (such as diabetes, high blood pressure or heart disease), whether they are pregnant or post-menopausal, other medications taken, kidney function, and if there is patient history of blood clots or migraines. This information is particularly important for dosing recommendation and premedication selection, and it helps clinicians tailor patient-specific suggestions for tolerating therapy.4

Following is a review of the key differences among the products’ stabilizers, osmolality, IgA content and concentration, as well as a discussion of infusion rate and route of administration.

Stabilizers

When intravenous IG (IVIG) was originally approved by the FDA in 1981, it contained no stabilizers, and patients often experienced undesirable side effects such as fever, chills, fatigue and chest, hip, joint and back pain, which were believed to be due to the formation of immunoglobulin aggregates. To resolve this issue, stabilizers were added, primarily sugars such as sucrose, maltose, glucose and sorbitol, and in some cases, glycine and albumin.5

The specific stabilizer used can play an important role in a product’s tolerability.1 Today, most IG products are no longer stabilized with a sugar; however, a few still are, which can result in other adverse events. There is a strong association between renal failure and sucrose-containing products, rapid rates of infusion and
diabetes. This is rare, and the cause of renal failure is unknown, but it is believed that it could be due to the fact that sucrose has the highest osmotic activity of the stabilizers in IG products. In addition, since sucrose is metabolized by an enzyme, called sucrase, that is found only in the intestine, when administered intravenously, sucrose is eliminated unchanged in the urine, possibly resulting in osmotic nephrosis. And, while cautious use of IVIG is recommended in patients at increased risk for adverse renal events, including those with renal impairment, diabetes mellitus, age greater than 65 years, dehydration or hypovolemia, sepsis, paraproteinemia or concomitant use of nephrotoxic drugs, they are not contraindicated in patients with renal insufficiency. In fact, screening for IgA deficiency prior to IVIG infusion is not routinely recommended.

The amount of IgA in a given IG preparation may also influence the risk for common reactions that are milder such as fever, malaise, myalgia and headache.

**Concentration**

Today’s IG products come in 5%, 10% and 20% solutions. The solution percentage is the number of grams of IgG protein in an IG therapy solution. For instance, a 5% IG product contains 5 grams of IgG protein per 100 mL of solution, a 10% IG product contains 10 grams of IgG protein per 100 mL of solution, etc. The highest-concentration 20% solution can only be infused subcutaneously. Three of the higher-concentration 10% products can be infused subcutaneously, while all of them can be infused intravenously. The lowest-concentration 5% products are approved only for intravenous infusion.

Most of today’s products are available as a ready-to-use liquid formulation. However, there are two products that are lyophilized and require reconstitution and pooling into an evacuated container for administration to the patient.

**Routes of Administration**

IG can be administered intravenously through a vein (IVIG) approximately once every three to four weeks, or subcutaneously under the skin (SCIG) every other week, once weekly or twice weekly. SCIG is FDA-approved therapy for only primary immunodeficiency, although it has been prescribed to treat other conditions.

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**With advances in manufacturing processes, today’s IG products are safer than ever before.**

products stabilized with maltose, there is a possible interaction with strips that test for glucose in the blood. The maltose may cause an erroneous reading indicating glucose is high when it really isn’t. However, most test strips have been modified to prevent these erroneous readings when maltose is present.

**Osmolality**

Osmolality is the solute concentration contained in the IG solution; thus, the higher the osmolality, the higher the concentration of the IG solution. Higher osmolality solutions, also known as hyperosmolar, are typically seen with older lyophilized IVIG products. In contrast, today’s fourth-generation products have a more physiologic osmolality comparable to that of individuals’ blood because they have had amino acids glycine and L-proline added to them to help reduce the overall solute load, which can become elevated with sugars.

Hyperosmolar solutions tend to cause more local venous irritation at the infusion site. They also may be associated with an increased risk of thrombosis. Dehydration also can cause the blood to become hyperosmolar, which is one of the reasons people receiving IG therapy are encouraged to drink a lot of water before, during and after the infusion.

**IgA Content**

All IG products contain varying amounts of IgA (one of the five classes of antibodies found in the blood). IgA is not problematic for most people. However, in patients who are IgA deficient, IgA can cause the formation of anti-IgA antibodies that can cause anaphylactoid reactions upon infusion of IVIG, which would result from the IgE development against IgA. While the risk of anaphylactoid reaction in IgA-deficient patients is anticipated, the incidence is low given the total number of reactions reported compared with the overall number of patients.
Risk-assessment guidelines are important tools when dosing regimens and routes of administration are being considered. Guidelines include evaluation of patient history and physical examination, risk factors, comorbidities and tolerance to appropriately manage potential serious and nonserious adverse events.

SCIG is not appropriate for everyone. For those requiring IG for autoimmune disorders, the SCIG route of administration may not be possible due to the large volume of solution needed for a dose. However, for someone receiving a smaller dose, SCIG administration may be possible. Those with very thin skin don’t tolerate SCIG as well as those with normal or thicker skin. And, very thin patients may not have enough fatty tissue in the space between the skin and the muscle to tolerate the SCIG infusion. Conversely, patients with very small veins or who have difficulty getting an IV started may be great candidates for SCIG.

In the past several years, SCIG has become an alternative method of administration of IG because of its many advantages. For one, SCIG results in a reduction in anaphylactoid reactions due to its slower absorption from the subcutaneous tissue into the systemic circulation. SCIG also eliminates the need for vascular access, stabilizes immune globulin levels and increases patient autonomy. And, notably, it has been used by patients with IgA deficiency with antibodies against IgA without inducing hypersensitive reactions. There is, however, an increased incidence of local reactions such as swelling and redness at the site of infusion.

Infusion Rates
Each patient has a maximum tolerated rate of infusion based on his or her risk factors and infusion-related reactions. For all patients, and this is essential for those just beginning therapy, IVIG should be administered slowly initially and titrated as tolerated. In general, primary immunodeficiency patients can be administered IVIG in one day approximately once a month (the half-life of IVIG is approximately 30 to 40 days), and they can be administered SCIG in one day over a matter of hours once or twice a week, or every two weeks as specified in the product labeling. On the other hand, patients with autoimmune diseases are generally administered larger doses of IVIG that, in some instances, may be divided into daily infusions over two to five days.

Slower rates of infusion have been linked to a reduced risk of side effects, including common reactions, acute renal failure, aseptic meningitis and thromboembolic complications. One of the most commonly reported side effects of IG therapy is headache, which has been found to increase with larger doses infused over a shorter period of time. Cases of aseptic meningitis are rare, but when it occurs, it requires discontinuation of IG treatment, and the symptoms typically stop after three to five days. Several causes for this have been proposed, including hyperviscosity that may be the result of rapid infusions of high doses into a volume-depleted hyperviscous bloodstream.

Stopping an infusion at the first sign of reaction tends to be the best way to manage it. After symptoms abate, most patients tolerate continuing the infusion at a slower rate.

Highly Improved and Tolerated Products
With advances in manufacturing processes, today’s IG products are safer than ever before. However, every IG product has different pharmaceutical characteristics, and there is even variation from batch to batch of each product. It’s these differences that can influence patient tolerability. But with careful patient screening and understanding of the inherent differences in the products, clinicians can ensure that the most appropriate product is prescribed to the patient.

RONALE TUCKER RHODES, MS, is the editor of IG Living magazine.
Did You Know?

Legislation

Stakeholders Conference Held for IVIG Demonstration Project

In November, the Centers for Medicare and Medicaid Services (CMS) held an open-door forum for the Medicare Intravenous Immune Globulin (IVIG) demonstration project that was signed into law on Jan. 10, 2013. The project is mandated by the Medicare IVIG Access and Strengthening Medicare and Repaying Taxpayers Act of 2012. The forum was held to get input from key stakeholders on questions related to the design and implementation of the demonstration project, which will examine the benefits of providing coverage and payment for items and services necessary to administer IVIG in the home for patients with primary immunodeficiency disease (PIDD).

During the three-year demonstration period, $45 million has been authorized to conduct and evaluate the benefit costs and administrative costs of administering IVIG in the home. The project is limited to 4,000 beneficiaries who have been diagnosed with PIDD and who voluntarily enroll to participate. Of particular note is that participants must be enrolled in Medicare Part B and must be covered under traditional Medicare (also known as fee-for-service Medicare) rather than Medicare Advantage, and they must not currently be treated for a home health episode for which they are already receiving benefits under homebound status. During the demonstration, there will be a per-visit payment amount for items and services needed for the in-home administration of IVIG based on the national per visit low-utilization payment amount under the prospective payment system for home health services.

While the project was scheduled to begin Jan. 1, CMS has announced that it has been delayed, as numerous system changes and approval processes still need to be implemented. CMS plans to start taking applications later in 2014 and begin delivering services after an enrollment period of about one month.

The main questions under discussion during the forum were:

- Should billing for demonstration-covered nursing services and supplies be permitted by organizations that are not supplying the drug?
- What types of organizations should be eligible to participate in the demonstration?
- To whom should CMS reach out to inform about this demonstration?
- How can CMS best reach out to beneficiaries and their providers?
- Should CMS have an open enrollment period during which applications would be submitted on an equal basis for consideration, rolling enrollment or some combination?
- Should a patient’s physician be required to sign a beneficiary’s application to confirm the diagnosis and awareness of the demonstration and location of service?
- Should the beneficiary’s application specify a particular drug or supplier?

Other issues under discussion included whether the demonstration was likely to impact the supply of IVIG and whether there are other factors Medicare should consider in designing the demonstration.

More information can be obtained by emailing ivigdemo@cms.hhs.gov.

Research

New Drug Approved for Treating Fungal Infections

The U.S. Food and Drug Administration (FDA) has approved a new tablet form of an antifungal drug manufactured by Merck. Noxafil (posaconazole), 100-mg delayed release tablets, is approved for preventing invasive Aspergillus and Candida fungus infections in patients aged 13 and older who are at high risk of developing them due to depressed immune function resulting from hematopoietic stem cell transplants and low white blood cell counts caused by chemotherapy, a condition known as neutropenia. The new tablet formulation is designed to be taken in two 300-mg doses on the first day, followed by a 300-mg dose once per day. Merck also markets a 40-mg-per-milliliter oral suspension form of the drug.
Scientists at the National Institutes of Health have discovered that a gene called Bach2 may play a central role in the development of diverse allergic and autoimmune diseases such as multiple sclerosis, asthma, Crohn’s disease, celiac disease and type 1 diabetes. In the study conducted on mice, the Bach2 gene was found to be a critical regulator of the immune system’s reactivity, which may explain why people with allergic and autoimmune diseases commonly have alterations in the Bach2 gene. The researchers found that if mice lacked the Bach2 gene, their cells became inflammatory and the mice died of autoimmune diseases within the first few months of life. When they reinserted Bach2 gene cells using gene therapy into Bach2-deficient cells, the mice’s ability to produce regulatory cells was restored.

Genome-wide association studies that analyze genetic variants among people to determine whether specific variants are associated with particular traits were critical to the discovery. These studies showed that DNA from patients with diverse autoimmune disorders often had minor alterations in the Bach2 gene, which laid the foundation for this research.

“Although genes have been found that play specific roles in either inflammatory cells or regulatory cells, Bach2 regulates the choice between the two cell types, resulting in its critical role in maintaining the immune system’s healthy balance,” said principal investigator Nicholas P. Restifo, MD. “It’s apt that the gene shares its name with the famous composer Bach, since it orchestrates many components of the immune response, which, like the diverse instruments of an orchestra, must act in unison to achieve symphonic harmony.”

The study must still be replicated in humans before its findings can be applied to the clinical setting. The study was reported in the June 2 online edition of *Nature*.

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Researchers at the University of Minnesota have identified infection-fighting and inflammation-suppressing functions for a gene associated with human autoimmune disease. The discovery centered on a gene known as PTPN22.

Researchers discovered a decade ago that people carrying a variant form of the PTPN22 gene have an increased risk of becoming sick with certain autoimmune diseases. According to the study’s authors, previous research showed that PTPN22 works in immune cells, but few studies had specifically examined PTPN22’s function in infection-fighting cells called myeloid cells, which are among the body’s first responders to a challenge with a virus or bacterium. They found that the PTPN22 gene not only fights infection, but that it also suppresses inflammation. Furthermore, they found that the PTPN22 risk variant is defective in suppressing inflammatory arthritis.

“We anticipate that our findings will open new lines of investigation into how PTPN22 and other autoimmune disease risk genes could work in infection-fighting and anti-inflammatory processes,” said Erik J. Peterson, one of the study’s lead authors. “Ultimately, we hope that the research will accelerate the drive toward better treatments and cures for autoimmune disorders.”

More research is under way to determine the impact of the PTPN22 variant in the function of myeloid blood cells, particularly in patients suffering from lupus. Researchers also are comparing immune responses to influenza A vaccines between carriers and non-carriers of the PTPN22 variant in an effort to understand the role of the disease-associated variant in mounting a normal response to immunizations against viruses.
Did You Know?

Guidelines

**IDSA Issues New Vaccine Guideline for Immunocompromised Patients**

The Infectious Diseases Society of America (IDSA) has issued a new guideline titled “Clinical Practice Guideline for the Vaccination of the Immunocompromised Host” to recommend that individuals with compromised immune systems get the flu shot and other vaccinations. These individuals tend to have lower vaccination rates in part because their doctors may be concerned about vaccine effectiveness and safety.

The guideline includes recommendations for most available vaccines, including measles, mumps and rubella, hepatitis A, pneumococcus, herpes zoster and influenza. Published in the Dec. 5 edition of *Clinical Infectious Diseases*, the guideline was written to fill a void in comprehensive recommendations for vaccinations for many different types of patients who are immunocompromised. It is also intended to help primary care physicians and specialists who treat immunocompromised patients, as well as people who live with these patients. “The guideline provides ‘one-stop shopping’ for clinicians caring for children and adults with compromised immune systems and includes recommendations and evidence for most vaccinations, from influenza to chicken pox,” said Larry Rubin, the lead author of the guideline. “Previously, the recommendations were difficult to retrieve because, in most cases, information has to be accessed individually by vaccine rather than by the category of patient disease.”

The guideline will be available in mobile device, pocket-sized quick-reference editions, and on the IDSA website at www.idsociety.org.

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**Imagine**

Imagine caring for your child with hemophilia with no factor, refrigerator, running water, electricity, or transportation to a clinic. This is the reality for thousands of families in developing countries. For just $20 a month, you can help an impoverished child with hemophilia. Become a sponsor today!

www.saveonelife.net / contact@saveonelife.net

Caring for people with hemophilia around the world—one at a time.
Working While Chronically Ill: Is It Possible?

Many chronically ill patients struggle to manage both their job and illness at the same time.

By Annaben Kazemi
Juggling work demands and sick time is never easy, but a chronic illness can make sick days disappear quickly, and bosses can become impatient with the uncertainty. According to the Partnership for Solutions, a national policy research program funded by the Robert Wood Johnson Foundation and based at Johns Hopkins University, more than 133 million Americans now live with at least one chronic health condition, and many live with more than one. By 2030, that figure is expected to grow to more than 171 million. This means an ever-increasing number of Americans face the challenge of managing a chronic illness while working, and many are eventually faced with the difficult decision of having to leave their careers.

Finding a work environment that is accepting and conducive to the demands placed upon individuals with chronic illness is challenging. How to find the right fit depends on each patient’s health, skill set and their company’s organizational culture. Success is dependent on flexibility and the willingness to make adjustments. In searching for the right fit, patients need to take in account their:

- Current field of work: Is it sustainable and flexible, and can modifications be made?
- Ability: What are the patient’s talents, gifts, skills and physical limitations?
- Access to equipment: Are needed tools also available at home; can equipment be modified to meet any physical needs?
- Financial situation: Are funds available if there is change or a setback; can a break in work be absorbed financially?

Once all factors have been taken into consideration, a plan can be put into place. Options patients contemplate most often when building a new plan are telecommuting, modifying the work day or workload, changing careers, working from home and starting a new business.

**Telecommuting**

One alternative for those coping with ongoing illness is to transition from working in an office to working from home, known as telecommuting. The home work environment can offer chronically ill patients many positives, including a more comfortable setting, privacy and the comfort of a disease-free environment. In fact, telecommuting has become a viable option for nearly 3.3 million people (approximately 2.6 percent of the U.S. workforce), according to the latest American Community Survey data from 2012. And, there are more than 316,000 disabled employees who regularly work from home using the Americans with Disabilities Act “reasonable accommodation” clause.

Laura, a software engineer in Seattle who suffers the symptoms of Guillain-Barré syndrome, found that telecommuting was a perfect solution for her. “After eight years of being sick, I was exhausted,” she said. Like many patients with chronic illness, working outside of the home eventually became unrealistic. She finally conceded that she had no choice but to restructure her life and find a way to reduce her time away from home. Since Laura had maintained a positive relationship with her employer, she decided to ask if they were flexible. Her boss agreed to restructure her workload and to allow her to telecommute on a trial basis. She was able to work from home two to three days a week and go into the office on the other days.

**Finding a work environment that is accepting and conducive to the demands placed upon individuals with chronic illness is challenging.**

Laura admits her biggest concern was that co-workers would think she was slacking off rather than working. “There was a misperception that if they didn’t see me at my desk busily chugging away, then I was not working,” she explained. “I really had to increase communication when I worked from home. I constantly updated my team about projects and worked hard to stay in the loop.” This strategy seemed to pay off because her boss agreed to continue allowing her to telecommute, and Laura was able to get the rest she needed.

**Modifying the Workload or Work Day**

Some patients find they simply cannot telecommute because the nature of their job requires them to be physically present at the job site. In this case, when a patient’s illness interferes with their success to manage the workload, employees may want to approach employers
Strategies for Maintaining Your Current Job

Workers with chronic illnesses face uncertainty and are forced to worry not only about their health but about their jobs. And, protections afforded chronically ill workers in the United States are thin and somewhat vague. To protect their health and their jobs, workers must navigate employers’ policies, which may include short- and long-term disability plans, as well as a patchwork of federal laws and regulations. Here are a few strategies to help you maintain your current job.

Talk to your supervisor. If you have a condition that could interfere with your performance, tell your supervisor. Be honest. Explain what your condition is and how it might affect your work. “People are often afraid of being discriminated against,” said Rosalind Joffe, a career coach who counsels people with chronic illnesses.1 A supervisor who understands what is wrong is less likely to make false assumptions about what you can and cannot do.

Ask for modifications. If your illness meets the definition of a disability, your employer is required under the Americans with Disabilities Act (ADA) to make reasonable accommodations to your job or work environment. A disability is defined as a physical or mental impairment that substantially limits one or more major life activities. Although your illness may be episodic or controlled by medications, it is still a disability, according to a recent amendment to the ADA.

If you are not sure what type of accommodations you are entitled to or how to ask for them, contact the Job Accommodation Network at (800) 526-7234. This service is provided by the Department of Labor. In general, the network recommends that you put your request to your employer in writing. But, if you work in a small, informal setting, that may not be necessary.

Know policies. You can learn about your company’s time off and sick leave policies by going to your company’s intranet or speaking with its human resources department. If you need to take a few weeks or months off due to your illness, research your company’s short- and long-term disability plans. Disability policies usually allow you to take a specific time off at reduced pay.

The Family and Medical Leave Act allows employees to take up to 12 weeks off each year for medical or family emergencies — but without pay. You can use the 12 weeks at any time, and you may take the time intermittently or all at once.1

Explore options. If you can’t continue to work at your current job due to your illness, find out whether you could work part time or could even take a different job in your company. If neither is feasible, explore new career possibilities. Be flexible and open to change.

Sometimes patients don’t have the choice to work or not to work. If illness prohibits a patient from working altogether, Social Security disability insurance can offer some relief. The process is lengthy, and employees must prove that they cannot work at any job. The amount paid is based on lifetime earnings; the number can be found on the annual statement from the Social Security Administration. Payments are modest, but once a patient receives disability payments for two years, they automatically qualify for Medicare coverage.

References
For people with primary immunodeficiency

Hizentra now offers even more freedom and flexibility

Now approved for biweekly (every 2 weeks) dosing

- The first and only 20% SCIg—now with the flexibility of biweekly administration

Now available in a convenient 10 g (50 mL) vial

- May reduce the number of vials patients need to handle and administer

Important Safety Information

Hizentra is indicated as replacement therapy for patients with primary humoral immunodeficiency (PI), age 2 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.

WARNING: THROMBOSIS

Thrombosis may occur with immune globulin products, including Hizentra. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.

For patients at risk of thrombosis, administer Hizentra at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity. See full prescribing information for complete boxed warning.

Please see additional Important Safety Information on reverse side and brief summary of full prescribing information for Hizentra, including boxed warning, on adjacent page.
Hizentra—
the first and only 20% SCIg therapy

- Delivers steady-state Ig levels in half the volume of 10% Ig products*
- Offers the choice of weekly or biweekly (every 2 weeks) dosing
- Supplied in 4 convenient vial sizes: 1 g (5 mL), 2 g (10 mL), 4 g (20 mL), and 10 g (50 mL)
- Does not require refrigeration, so patients can infuse anytime, anywhere

Now available in a convenient 10 g (50 mL) vial

Important Safety Information (continued)
Hizentra is contraindicated in patients with a history of anaphylactic or severe systemic reaction to human immune globulin preparations or components of Hizentra, such as polysorbate 80. Because it contains the stabilizer L-proline, Hizentra is contraindicated in patients with hyperprolinemia. Hizentra is also contraindicated in patients with immunoglobulin A deficiency who have antibodies against IgA and a history of hypersensitivity.

Hizentra should be administered subcutaneously only. Do not administer intravenously.

IgA-deficient patients with anti-IgA antibodies may be at greater risk of developing potentially severe hypersensitivity and anaphylactic reactions with administration of Hizentra. If hypersensitivity occurs or anaphylactic reactions are suspected, discontinue administration immediately and treat as medically appropriate.

Monitor patients for aseptic meningitis syndrome (AMS), which has been reported with SCIg. In patients at risk of acute renal failure, monitor renal function, including blood urea nitrogen, serum creatinine and urine output. Also monitor patients for clinical signs of hemolysis or transfusion-related acute lung injury (TRALI).

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

The most common adverse reactions (observed in 5% or more of study subjects receiving Hizentra) were local reactions (ie, swelling, redness, heat, pain, and itching at the injection site), headache, diarrhea, fatigue, back pain, nausea, extremity pain, cough, rash, pruritus, vomiting, upper abdominal pain, migraine and pain.

Ig administration can transiently impair the efficacy of live attenuated virus vaccines, such as measles, mumps and rubella. It can also lead to misinterpretation of serologic testing.

Please see additional Important Safety Information on reverse side and brief summary of full prescribing information for Hizentra, including boxed warning, on adjacent page.

*Based on an equivalent dose in grams.
Hizentra is manufactured by CSL Behring AG and distributed by CSL Behring LLC.
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1020 First Avenue, PO Box 81501, King of Prussia, PA 19406-0901 USA
HIZENTRA®, Immune Globulin Subcutaneous (Human), 20% Liquid
Initial U.S. Approval: 2010

BRIEF SUMMARY OF PRESCRIBING INFORMATION
These highlights do not include all the information needed to use HIZENTRA safely and effectively. See full prescribing information for HIZENTRA.

WARNING: THROMBOSIS
See full prescribing information for complete boxed warning.

• Thrombosis may occur with immune globulin products, including Hizentra. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.

• For patients at risk of thrombosis, administer Hizentra at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.

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

DOSE AND ADMINISTRATION
For subcutaneous infusion only. Do not inject into a blood vessel.

Administer weekly or biweekly (every two weeks).

Dosage
Before switching to Hizentra, obtain the patient's serum IgG trough level to guide subsequent dose adjustments.

Weekly: Start Hizentra 1 week after last IGIV infusion

Initial weekly dose = Previous IGIV dose (in grams) x 1.53
No. of weeks between IGIV doses

• Biweekly: Start Hizentra 1 or 2 weeks after the last IGIV infusion or 1 week after the last weekly Hizentra infusion. Administer twice the calculated weekly dose.

• Adjust the dose based on clinical response and serum IgG trough levels (see Dose Adjustment).

Administration

• Infusion sites – 1 to 4 injection sites simultaneously, with at least 2 inches between sites.

• Infusion volume – First infusion, up to 15 mL per site. Fifth infusion, up to 20 mL per site, then to 25 mL per site as tolerated.

• Infusion rate – Up to 15 mL per hr per site. Increase to 25 mL per hr per site as tolerated.

DOSE FORMS AND STRENGTHS
0.2 g per mL (20%) protein solution for subcutaneous injection

CONTRAINDICATIONS
• Anaphylactic or severe systemic reaction to human immune globulin or components of Hizentra, such as polysorbate 80

• Hyperprolinemia (type I or II) (Hizentra contains the stabilizer L-proline)

• IgA-deficient patients with antibodies against IgA and a history of hypersensitivity

WARNINGS AND PRECAUTIONS
• IgA-deficient patients with anti-IgA antibodies are at greater risk of severe hypersensitivity and anaphylactic reactions.

• Thrombosis may occur following treatment with immune globulin products, including Hizentra.

• Aseptic meningitis syndrome has been reported with IGIV or IGSC treatment.

• Monitor renal function, including blood urea nitrogen, serum creatinine, and urine output in patients at risk of acute renal failure.

• Monitor for clinical signs and symptoms of hemolysis.

• Monitor for pulmonary adverse reactions (transfusion-related acute lung injury [TRALI]).

• May carry a risk of transmitting infectious agents, e.g., viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.

ADVERSE REACTIONS
The most common adverse reactions observed in ≥5% of study subjects were local reactions (i.e., swelling, redness, heat, pain, and itching at the injection site), headache, diarrhea, fatigue, back pain, nausea, pain in extremity, cough, rash, pruritus, vomiting, abdominal pain (upper), migraine, and pain.

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.

DRUG INTERACTIONS
The passive transfer of antibodies may interfere with the response to live virus vaccines, and lead to misinterpretation of the results of serological testing.

USE IN SPECIFIC POPULATIONS

• Pregnancy: No human or animal data. Use only if clearly needed.

• Pediatric: No specific dose requirements are necessary to achieve the desired serum IgG levels.

Based on September 2013 version
later and reduce her work hours each day, resulting in her working 65 percent of her full-time hours. The downside of this new schedule was that working part-time also resulted in part-time pay. Yet, Cindi said it’s the only way she could manage keeping her job and her health. She has managed to juggle the financial changes with the help and support of her family, and has moved in with her sister and brother-in-law.

**Retraining**

Sometimes the nature of a patient’s illness makes the current job unsustainable. In such cases, retraining within a current field or even changing careers can be an option. Some companies will provide retraining opportunities, but if that is not an option, patients can look into short-term vocational classes, certification or technical classes at community colleges, career-oriented classes in secondary schools, apprenticeship programs, and a variety of other programs and institutions that offer job-specific skills.

The federal government plays a major role in training the disadvantaged and displaced, funding many major programs through the Department of Labor and Department of Education; it also funds the task of providing reemployment services. Such was the case for Daniel, a self-employed carpenter from Georgia who needed their support services.

*Copies of the physical limitations of his chronic illness, Daniel found himself struggling to make ends meet as his disease state progressed. The chronic flare-ups were occurring more often, and Daniel was unable to meet the physical demands of his job, where he had to climb ladders, work in tight quarters and lift and manipulate heavy objects. As a self-employed contractor, when he was unable to work, there were no sick days or vacation days

*Sometimes the nature of a patient’s illness makes the current job unsustainable.*
to fall back on. Daniel found himself facing the reality that he would not be able to continue working in the only field he’d known since graduating high school. The pressure to provide for his family was enormous as the medical and household bills began to mount quickly.

Even though Daniel thought he’d never be in a classroom again, he enrolled in school and retrained for a new career in technology with fewer physical demands. “That year of going to back school, coping with debt and managing my illness was incredibly challenging,” Daniel said. “My wife had to take on more financial responsibility, and that was hard on our family, too. But, the doors of opportunity that opened have allowed me a new lifestyle.” He now works for the IT department of a local school district, repairing and updating classroom computers and laptops. “The environment and work schedule are more conducive to juggling my medical treatments, and I am not hindered by my physical limitations. And, the medical insurance is a huge plus!” he explained.

Opening Your Own Business

Statistics from the U.S. Corporations of America show that one in every 153,700 people owns their own business. And, in fact, more than half of all U.S. businesses are based out of an owner’s home.\(^3\) While some may see starting a business from home as a luxury, it is the only way to survive for many with chronic illness.

Rachel had always loved her job as a pediatric nurse, and her employers were accommodating about flexing her work schedule when she was ill. But, eventually, even working minimally became too much. “I had a feeling of exhaustion so profound I could hardly get through an hour of work, let alone a full day,” Rachel said. “I could no longer manage many of the simplest tasks of daily living, let alone working. I had gone from five days a week, to four, then three, but eventually two even became too much.” That’s when Rachel knew her job situation had to change. Rachel began to look at what she could do from home.

She had a small savings and decided to use her financial resources to start her own business. After assessing her skills and realistically looking at her abilities, Rachel opened a nanny service providing short-term care for sick kids of working parents. “I saw a need while I was working at the pediatric office and thought of how I could fill that need,” she explained. She turned the extra room in her home into a home office, and is able to conduct most of her business by phone. There is very little overhead, and referrals have come from the parents she met while nursing.

“The best part of owning my own business is having the flexibility to take care of myself,” she said. “I can sleep when I need to and schedule meetings on my time.” Rachel states that a big advantage is that it’s much easier to schedule her infusion therapy, without worrying about the disruption to work. She says that often she infuses while working. But, there is also a downside to working at home: She misses the social connections of being in an office with other professionals. To avoid the feelings of isolation that can come from working at home, Rachel is conscious to create an online network that enables her to stay connected professionally, as well as socially.

Finding the Right Fit

Some patients find their financial situations have changed little since they became ill; their symptoms are mild, and they can continue to work with a few adjustments. For others, however, financial pressures can be great, even overwhelming. Some patients live alone with little or no income and struggle to make ends meet because they can no longer work. Many patients are somewhere in between, stressed to some degree, but able to maintain a lifestyle more or less similar to the one they had before becoming ill. The bottom line: Each person’s illness is different, and each person’s ability to cope and work with illness is different.

Finding the right work environment that can accommodate the challenges of chronic illness is difficult. Success is dependent on the willingness to make adjustments. The key is to be flexible and find the solution that’s right for you. ■

ANNABEN KAZEMI is the patient advocate for IG Living magazine.

References


Resources

- Career One Stop: www.careeronestop.org/TridionMultimedia/mySkillsmyFuture_tcm24-7103.pdf
What causes a disease — especially a chronic or rare disease? How can clinicians be sure that the medications prescribed to treat a disease are the best ones? Are those medications always safe? How can patients or providers increase the chances that health insurance will cover the cost of medications? How can it be determined what the long-term health prognosis will be for patients diagnosed with a disease? These are but a sampling of questions to which both patients and healthcare providers seek answers when it comes to patient care.

For years, healthcare providers have based their care for patients upon clinical trials that provide data about treatment effects in controlled conditions. While these data are reliable, they often are not applicable to the diverse population, which can result in “evidence gaps that impede the ability of patients and providers to make informed treatment decisions and of payers to determine what kinds of coverage will be appropriate.”¹ Therefore, to care for their patients based on real-world results, healthcare professionals are now focusing on evidence-based medicine, which is what patient registries provide.

Patient registries are operated by many different entities, including the federal government, state governments, universities, hospitals, non-profit organizations and private groups.² No one knows exactly how many patient registries there are. For instance, ClinicalTrials.gov contains more than 800 patient registries, but according to Elise Berliner, who heads the U.S. Agency for Healthcare Research and Quality’s (AHRQ’s) technology assessment, those probably represent just the “tip of the iceberg.”³ But, it is known

Why Join a Patient Registry?

The number of patient registries continues to grow, especially in the chronic and rare diseases communities, due to the benefits they provide to both patients and the healthcare community.

By Ronale Tucker Rhodes, MS
that the number of registries is growing and that they are powerful tools that provide broader results about the course of disease, variations in treatment and outcomes, factors that influence prognosis and quality of life, patterns in the delivery of care, and the effectiveness, safety and quality of care.4

Who Is Eligible to Join a Patient Registry?

To participate in a patient registry, a patient must be either diagnosed with the disease or treated with the medicine for which the registry is tracking. For instance, USIDNET, a registry established and managed by the Immune Deficiency Foundation for patients diagnosed with primary immunodeficiency diseases (PiDDs), originally enrolled only patients diagnosed with the more widely known PiDDs, including severe combined immunodeficiency, X-linked agammaglobulinemia, common variable immune deficiency, DiGeorge syndrome, Hyper IgM syndrome, Wiskott-Aldrich syndrome and chronic granulomatous disease. However, the registry has now been enlarged to include more than 100 PiDDs.5

Patients can register for most registries on their own. However, it is more typical for healthcare providers to be the facilitator between their patients and the registries. Patients’ healthcare providers assign someone in their office to input patient data into the registry and keep it updated as is required by the registry. Each registry differs in the types of information collected on patients once they have joined. All registries require patients to sign a consent form. Other forms that may be required include an authorization for the release of health information, a participant intake questionnaire, a family history questionnaire, an affected questionnaire (more in-depth questions about various symptoms, treatments and experiences), a semi-annual or annual update form or others.

How Is the Information in Patient Registries Used?

Registries are intended to collect data for a variety of reasons, including estimating the magnitude of a disease, determining the incidence of disease, examining trends over time, assessing healthcare quality, conducting research, estimating survival analysis, investigating etiological hypotheses, serving as a source of potential participants in clinical trials, and the reasons go on.2 But, the four major purposes for patient registries are to evaluate the natural history of disease, determine the clinical effectiveness and cost-effectiveness of treatment, monitor safety of treatment and improve quality of treatment.1,4

When evaluating the natural history of disease, its characteristics, management and outcomes with or without treatment are tracked. The reasons for tracking this may be due to the natural history of a disease not being well-described, to the variation across different groups and geographic regions that often change over time, or to changes in the disease after the introduction of certain therapies.1,4 For instance, the CGD (chronic granulomatous disorder) Society established a registry of CGD patients within the UK and Ireland, and in 2011 after evaluating the collected data, it found that younger patients generally are doing fairly well, but older patients still suffer complications and earlier death than unaffected people.6

All information in a registry is protected by the HIPAA Privacy Rule.

In contrast to the information that can be gained from clinical trials, registries can determine the clinical effectiveness of a treatment or service by looking at broader population bases (e.g., older versus younger, men versus women, etc.) and longer periods of time (e.g., from childhood into adulthood).1,4 Using the CGD registry as an example, it found that many years ago, young boys presenting with what looked like Crohn’s disease (a granulomatous inflammation of the gut) would simply be treated as having inflammatory bowel disease, even if it was CGD. Now, however, gastroenterology colleagues always think about CGD. In addition, the CGD registry found that survival is better today than it was 10 or 15 years ago for CGD patients, but it is not normal in most cases, and the quality of life is not always normal.6 Another example of how registries can improve clinical effectiveness is the Improve Care Now registry that collects data on children and adolescents with Crohn’s disease and ulcerative colitis. Since the registry was started, the percentage of kids with Crohn’s disease and ulcerative colitis who are in remission has increased from 50 percent to more than 75 percent — all without new medicines.7

Registries also can be designed to collect cost data and effectiveness data to model the comparative value of a treatment’s or service’s ability to achieve a desired outcome, such as life expectancy or disease-free periods. For providers,
Registries for Patients Whose Treatment May Include Immune Globulin

Below is a short list of the many registries that exist in the U.S. for patients who rely on immune globulin products — patients who suffer from both immune deficiencies and autoimmune disorders. To find a registry for a specific disease that is not listed here, patients can contact their healthcare providers or the organizations listed in the Resources section in the back of this and every issue of IG Living magazine, as well as those on our website at www.IGLiving.com.

**USIDNET: [www.usidnet.org](http://www.usidnet.org)**

USIDNET is a clinical registry for residents of the U.S. who are affected by several different primary immunodeficiency disorders. Established and managed by the Immune Deficiency Foundation, the registry is an outgrowth of a National Institute of Allergy and Infectious Diseases-supported pilot project begun in 1982 to establish a similar registry for U.S. residents affected by chronic granulomatous disease (CGD), which provided considerable information to patients about CGD that had not been previously available. USIDNET’s goal is to provide improved access to patients by researchers conducting both basic and clinical studies; accurate and up-to-date profiles useful to clinicians and genetic counselors; and improved access by patients to information about the latest treatments. The registry also will include disease prevalence in the overall population and in various subgroups; the number and types of genetic defects that result in PIDDs, the clinical spectrum of these diseases; the correlation between the genetic defect and clinical picture; effects of current therapy on the course of PIDDs; and causes and incidences of morbidity and mortality. There are 33 enrollment sites in the U.S.

**IDEaL (Immunoglobulin, Diagnosis, Evaluation and Key Learnings): [www.idealpatientregistry.com](http://www.idealpatientregistry.com)**

IDEaL seeks physicians to participate as sub-investigators in this landmark patient registry. All physicians who prescribe immune globulin therapy are encouraged to participate, and physician and patient participation are completely voluntary and can be terminated at any time. The registry is observational in nature, requiring no change to a patient’s medical care. Subjects in the registry are contacted approximately every six months to complete various quality-of-life scales.

**Rare Diseases Clinical Research Network (RDCRN)**

**Patient Contact Registry: [rarediseasesnetwork.epi.usf.edu/registry](http://rarediseasesnetwork.epi.usf.edu/registry)**

This registry was created to inform patients and/or parents of patients about clinical research studies. Information contained within the registry is used for recruitment for research studies directed at improving the knowledge and treatment of rare diseases, making it possible for researchers to find new treatments, create new studies and work for the improvement of patients’ lives. Supported by the Office of Rare Diseases Research and National Center for Advancing Translational Sciences, RDCRN has more than 150 clinical sites.

**MYOVISION: [www.myositis.org](http://www.myositis.org)**

MYOVISION is a myositis patient registry designed to capture environmental exposures and other potential triggers of myositis. Utilizing funds from the Centers for Disease Control and Prevention, The Myositis Association will collect information from adults and children with myositis about their disease.

**Peripheral Neuropathy Research Registry (PNRR):**


Launched by the Foundation for Peripheral Neuropathy, the PNRR focuses on diabetic, chemotherapy-induced, HIV/AIDS and idiopathic peripheral neuropathies. PNRR will collect information from adults and children participating in clinical studies to provide a current and accurate database for researchers. The registry is observational, requiring no change to a patient’s medical care. Subjects in the registry are contacted approximately every six months to complete various quality-of-life scales.

Because registries provide data on a broad population, they can be valuable for monitoring patient safety. They can act as surveillance systems to monitor a population for any occurrence of an unexpected or harmful event, which can help to both quantify the risk and to properly attribute it. In fact, FDA noted that “through the creation of registries, a sponsor can evaluate safety signals identified from spontaneous case reports, literature reports or other sources, and evaluate the factors that affect the risk of adverse outcomes such as dose, timing of exposure or patient characteristics. Registries also can be used to monitor safety for products with a known risk factor, which can be acces-
neuropathies. It was created to help researchers better characterize clinical phenotypes of patients with the disorder and will facilitate both the basic and clinical research studies needed for an improved understanding of the etiology and pathogenesis of peripheral neuropathy.

**Inflammatory Bowel Disease (IBD) Registry:**
www.hasbrochildrenshospital.org/services/pediatric-astroenterology/research/inflammatory-bowel-disease-registry.html

The Pediatric IBD Collaborative Research Group (PIBD CRG) at the Hasbro Children’s Hospital formed the IBD Registry to gather demographics, clinical and laboratory information, as well as responsiveness to treatment data on children newly diagnosed with IBD. Twenty-one centers throughout the U.S. and Canada have enrolled more than 600 children to participate in this prospective, observational research program that examines treatments and quality-of-life outcomes to gain a better understanding of how these issues impact children newly diagnosed with IBD.

**Improve Care Now Registry:**
improvecarenow.org

Improve Care Now is a network designed to improve the care and health of children and adolescents with Crohn’s disease and ulcerative colitis. The network has developed Model IBD Care: Guidelines for Consistent Reliable Care based on carefully analyzed results of thousands of doctor-patient visits, as well as the latest studies and treatments worldwide. Clinicians and patients apply this information, experiences are tracked and studied, results are shared openly and further refinements are made to the guidelines to continually improve care for patients.

How Do Patient Registries Protect Patients’ Privacy?

A concern among many patients when participating in a registry is whether their personal information will be revealed to those who are using the data. How personal information is protected is governed in several ways.

First, for an institution to receive U.S. Department of Health and Human Services (HHS) support for research involving human subjects, it must designate at least one international review board (IRB) registered with the HHS Office for Human Research Protections (OHRP). (Protection of human subjects arose from the Belmont Report in the 1970s to be sure all human research follows the same code, which is known as the “common rule” that is under the jurisdiction of the HHS.) An IRB, also known as an independent ethics committee or ethical review board, is a committee that has been formally designated to approve, monitor and review biomedical and behavioral research involving humans. The board often conducts some form of risk-benefit analysis in an attempt to determine whether or not research should be done, with its No. 1 priority to protect human subjects from physical or psychological harm. There is no federal IRB; typically, each institution has its own IRB, which is known as an “internal” IRB. However, it is also possible for an institution to designate an already registered IRB operated by another organization (an “external” IRB) after establishing a written agreement with that organization. Also, to receive support by HHS for research involving human subjects, the institution must have received a federalwide assurance by OHRP that commits to HHS that the institution will comply with the requirements in the HHS Protection of Human Subjects regulations.

Second, all information in a registry is protected by the HIPAA Privacy Rule. HIPAA provides federal protections for individually identifiable health information held by covered entities and their business associates. It also gives patients an array of rights with respect to that information, while at the same time balancing those rights so that the disclosure of health information needed for patient care and other important purposes is permitted.

Last, all patients in a registry are assigned a random code, and all personal identifying information for each of those patients is removed and replaced with that code, which is the only identifying item that is ever used when accessing the data. The registry manager is the only person who has access to the personal data if it is contained in the registry database. In addition, representatives of organizations who are providing patient data to the
registry can only input and retrieve their own organization’s data. (Registries typically work with healthcare organizations or doctors’ offices that designate one or more individuals to enter their patients’ data into the registry.) Should they wish to access other organizations’ data, they would have to formally request it and, if approved, the data still would only include the code; no patients’ identifying information in that data would be available.

When patients decide to enroll in a registry, they often are given different options. An example of this is USIDNET. Each patient participating in the USIDNET registry is provided with four options when enrolling. Under option one, the patient’s identity is not stored in the registry and, therefore, it is not accessible by the registry administrator, so that administrator will not be able to determine which information is the patient’s. Instead, a code number is assigned to the patient’s information, and only the patient’s doctor knows it. In addition, the patient’s doctor is the facilitator of all communication between the registry and the patient. Under option two, the patient’s name, date of birth and mailing and/or email address are recorded by the registry administrator. However, that personal information is kept in a separate database from the patient’s medical information that contains an assigned code number. Therefore, investigators reviewing the patient’s medical information do not have access to the patient’s identifying information. Patients under this second option also have two other options: 1) to have all communication between the registry and the patient go through the patient’s doctor (unless the patient has moved or has ended contact with his or her doctor); or 2) to allow the registry staff to directly inform the patient about research results based on information gathered on the patient’s disease. The doctor would also receive the information.5

What is particularly important for patients to understand is that registries are considered “minimal risk” because they are merely sharing information about human subjects; they are not actually testing medical procedures or products on human subjects.

**Succeeding with Patient Registries**

Due to the growth in the number of patient registries, in December 2012, the Registry of Patient Registries (RoPR) was established by AHRQ. Known as a “metaregistry,” the goal of RoPR is to create a one-stop shop where physicians, patients and researchers can find lists of individuals who have made themselves available for observational medical studies. In essence, the database would serve patients and physicians looking for specific disease registries, researchers investigating a particular disease, and drug developers. The metaregistry also could be used to monitor outcomes and study best practices. RoPR has particular potential for rare diseases about which little is known by aggregating data and information, speeding up research on the diseases and ensuring that research projects are not redundant.3

But, the benefits of patient registries to patients and healthcare providers depend upon patients’ participation because these registries must contain enough data to be useful. Some of the benefits of sufficient data include helping to determine the causes of disease, the best therapies, how to prevent adverse events and how patients can be provided better quality of care. Patient registries can be found through an Internet search, through patients’ healthcare providers or by contacting patient organizations. ■

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

**References**


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When Deborah Norris was a young mother, she and her husband and two kids were active; they went “biking, hiking, to the beach, skating at the local roller rink and played chase.” But, in 1992, after she had arthroscopic surgery on her knee, Deborah says she felt “the worst pain I’ve ever experienced.” It began after a second surgery to clean out what her surgeon thought was an infection that occurred after the first. While recovering, she was unable to tolerate anyone touching her knee or that side of her leg. “I was swollen from my knee to my ankle, and the skin was so shiny, you could see your reflection it,” explains Deborah. “It was either a bright red, or at times, it changed to a purplish blue. Physical therapy kept my mobility to a point, but I still needed a cane to walk. Gone were my carefree hiking days! At that point, I was spending most of my days flat on my back with my leg raised to help the swelling or doing exercises for mobility.” Fortunately, after only three months, a doctor diagnosed Deborah with reflex sympathetic dystrophy, known today as complex regional pain syndrome (CRPS) — an early diagnosis for this disease. Unfortunately, there is no cure.

What Is CRPS?

CRPS is an uncommon form of chronic pain in which high levels of nerve impulses are sent to an affected site, usually an arm or a leg. What causes CRPS isn’t clearly understood, but it typically develops after a forceful trauma to an arm or a leg (such as a crush injury, fracture or amputation), surgery, stroke or heart attacks, infections and even emotional stress. And, it is believed that CRPS does not have a single cause, but instead results from multiple causes that produce similar symptoms. Why these injuries can trigger CRPS is also not well understood. One theory is that it may be due to a dysfunctional interaction between an individual’s central and peripheral
nervous systems and inappropriate inflammatory responses (an immune system correlation), thus disrupting the healing process. What is known is that the pain is out of proportion to the severity of the initial injury, if any. Knowing this, it is believed there are two ways to reduce the risk of developing CRPS. First, studies have shown that those taking vitamin C after a wrist fracture have a lower risk of CRPS than those who don’t take vitamin C. Second, some research suggests that people who get out of bed and walk around soon after a stroke (early mobilization) lower their risk of CRPS.

There are two similar types of CRPS, called CRPS-I and CRPS-II (previously known as causalgia), both of which have the same symptoms and treatments. Individuals with confirmed nerve injuries have CRPS-II, while individuals without confirmed nerve injury are classified as having CRPS-I. About 90 percent of patients with CRPS have CRPS-I.

CRPS can strike anyone at any age, and while it affects both men and women, it is more common in women. The average age of individuals with the disease is approximately 40. And, while CRPS is rare in the elderly, it is not uncommon in children, although not before age 5 and only very rarely before age 10.

The jury is out on how common CRPS is in the general population. To date, there have been only two studies conducted. The first, conducted by Sandroni et al. in 2003, reported that CRPS-I is rare with only 5.46 new cases per 100,000 annually that were associated with frequent spontaneous resolution. The conclusions of this study were controversial, however, because 90 percent of the individuals in the study received physical therapy and nearly half received sympathetic blocks and pharmacological intervention. The second study was conducted by de Mos et al. in 2007. When based on clinical diagnoses confirmed by the original treating physicians, this study reported the incidence was 26.2 new cases per 100,000 annually, a figure that is 4.2 times higher than the first study. And, even when restricted to those cases in which detailed specialist evaluation data were available to make independent diagnoses, this second study reported an incidence of 16.8 new cases per 100,000 annually, nearly three times higher than the first study.

Symptoms of CRPS

The main symptom of CRPS is prolonged and intense pain that, in some cases, may be constant, and in others, may come and go. The pain may consist of continuous burning or throbbing; sensitivity to touch or cold; swelling; changes in skin temperature, color and texture; changes in hair and nail growth; joint stiffness, swelling and damage; muscle spasms, weakness and loss; and decreased ability to move an affected body part. CRPS also can spread from one part of the body to another. This is the case for Deborah. Her pain began in her left leg on which she had knee surgery, but now it has spread to her right leg and hand and to the right side of her face.

Fortunately for Deborah, she was diagnosed in three short months. This is not the case for most, which can cause complications. Not diagnosing and treating CRPS early may cause the disease to progress to more disabling signs and symptoms, including tissue wasting (atrophy) and muscle tightening (contracture).

CRPS can strike anyone at any age, and while it affects both men and women, it is more common in women.

Diagnosing CRPS

CRPS can’t be diagnosed with any test. However, testing is important to rule out the cause of pain from other conditions such as arthritis syndromes, Lyme disease, generalized muscle diseases, a clotted vein or small nerve fiber polyneuropathies, all of which require different treatment. Generally, doctors will diagnose CRPS based on a physical examination that reveals a higher-than-expected amount of pain from an injury, a change in appearance of an affected area and no other cause of pain or altered appearance. Doctors will also look at the patient’s medical history of earlier injury to affected areas. In addition, some tests can provide clues as to whether a person has CRPS. These include a bone scan that can help detect bone changes, sympathetic nervous system tests such as thermography that measures the skin temperature and blood flow of affected limbs, an MRI that can show a number of tissue changes, and X-rays that can reveal a loss of minerals from bones that often occurs later in the stages of this disease. CRPS is often associated with excess bone resorption, a process in which certain cells break down the bone and release calcium into the blood.
Treating CRPS

There are many different treatments for CRPS that work for some and don’t work for others. Rehabilitation therapy is often used to help keep a patient’s painful body part moving to improve blood flow and lessen the circulatory symptoms, as well as to improve flexibility, strength and function. It can also help to treat the secondary profound psychological symptoms, including depression, anxiety or post-traumatic stress disorder, that are associated with CRPS and that heighten the perception of pain. Before Deborah found a physician who prescribed medications to lessen her pain, she said there were times when she felt she understood why people commit suicide. “I knew I couldn’t because I love my husband and children too much to leave them like that, but when you’re the only one awake at 2:00 a.m., and your pain is intolerable, you get quite depressed,” she explained.

While there are no drugs that are specifically approved to treat CRPS, there are several that have been shown to be effective, particularly when used in the early stage of the disease. These include non-steroidal anti-inflammatory drugs (aspirin, ibuprofen and naproxen), corticosteroids (prednisolone and methylprednisolone) in the early stages, drugs initially developed to treat seizures or depression, botulinum toxin injections, opioids (oxycontin, morphine, hydrocodone, fentanyl and vicodin), N-methyl-D-aspartate receptor antagonists (dextromethorphan and ketamine), nasal calcitonin for deep bone pain and topical local anesthetic creams and patches. Deborah lived in extreme pain from CRPS for five years before she located a doctor who was willing to prescribe pain medication, which allowed her to go back to teaching elementary school full time for 10 years. Of course, there were still bad times for which she had to find coping mechanisms. But, her doctor also prescribed an antidepressant, which greatly helped her.

Other treatments that can help to relieve the pain associated with CRPS include sympathetic nerve blocks that provide temporary pain relief; spinal cord stimulation to provide a tingling sensation in the painful area; neurostimulation delivered near injured nerves, outside the membranes of the brain and within the parts of the brain that control pain; intrathecal drug pumps that deliver pain-relieving medications, typically opioids and local anesthetic agents such as clonidine and baclofen, directly into the fluid that bathes the spinal cord; and surgical sympathectomy to destroy some of the nerves. While surgical sympathectomy is somewhat controversial, Deborah undergoes the procedure approximately every six months, which eliminates the pain in her face. “It has to be done about every six months because nerves have a tendency to grow back,” explains Deborah. “But, those months with no facial pain are like heaven.”

There also are emerging treatments for CRPS, including low doses of ketamine given intravenously for several days to either reduce substantially or eliminate the chronic pain, hyperbaric oxygen to deliver more oxygen to the body’s organs and tissues, and immune globulin (IG) infusions. In a small trial in Great Britain, 13 patients with CRPS who did not respond well to other treatments and who were given low-dose intravenous IG for six months to 30 months had a greater decrease in pain scores than those receiving saline during the following 14 days after infusion. Deborah, who is also diagnosed with common variable immune deficiency, was prescribed subcutaneous IG infusions weekly to see if it would reduce her pain. “At first I thought it did,” said Deborah. “But, now, I’m not so sure. The one thing it does do, though, is make the rest of my body feel better and eliminate all the infections I get so easily.”

CRPS can’t be diagnosed with any test.

Living with CRPS

Living day to day with pain is difficult enough, often affecting the mental health of patients with CRPS, which is why it’s often a good idea for those with CRPS to see a psychologist or other professional to help them put things in perspective. Support groups also can help by sharing experiences and feelings with other people with CRPS. Add to that the difficulty of making family and friends understand what they’re experiencing, and living with CRPS can seem almost unbearable. Deborah knows this all too well. Her limbs are no longer swollen, so she looks normal, but she is still in a lot of pain. “Usually when people ask how I am, I lie and say ‘fine’ because they won’t understand the truth,” explains Deborah. “And, it’s hard to blame them. You might be screaming inside, but if people don’t have the experience of a debilitating disease in their own life, they can’t relate.” Her advice: It helps to have a
significant person in your life who understands your illness and believes you. I’m very fortunate to have a great, compassionate husband who’s also my best friend. My children (now 31 and 34) get it, but still at times underestimate the amount of pain I’m in. But, my husband knows just by reading my body signs — the curl of my fingers, the constant wiggling of my feet. That person makes the biggest difference in how you’re able to cope. Being understood is so important!”

Deborah also offers some additional advice. First, patients should find a doctor who is not suspicious of their pain. Instead, they should look for a doctor who is understanding and willing to be on their team. Second, they should find an outside interest or a hobby that they can focus on. “I’ve been very lucky because I’m a watercolor artist, and when I paint, I can ‘lose’ myself and set my pain aside,” she says. “It’s sort of like meditating.” Deborah also has other interests, including a puppy to train and care for. “I got a puppy when I found it necessary to retire from teaching six years ago,” adds Deborah. “My dad had just died, and I felt sorry for myself sitting at home with no job to make demands on me and command my focus, and [I was] in pain. My Maltipoo puppy, Daisy, took time to train, to care for and has become a therapy dog. Now, she and I are able to go to facilities for the elderly and other places to visit people, or to the library where children read to her. Being active not only takes your mind off of your pain, it helps you to personally feel like you are valuable.”

Research in Progress

While there are significant discrepancies in CRPS incidence, applying the most conservative incidence figures in the study by de Mos et al. to current U.S. census bureau population estimates, it is expected that there will be more than 50,000 new cases of CRPS-I annually. And, although that number on a percentage basis does not indicate that CRPS is common in the general population, it does represent a substantial number of patients who will develop CRPS every year with significant quality of life consequences. This, then, demonstrates the importance of continued epidemiological investigations, as well as continued research to more aggressively treat the disease and lessen its symptoms.

Researchers at the National Institute of Neurological Disorders and Stroke, part of the National Institutes of Health, are involved in many different studies concerning CRPS. Using an animal model of the disorder, they hope to better understand the neuroinflammatory basis of CRPS and to identify the relevant inflammatory signaling pathways that lead to the development of post-traumatic CRPS such as what occurs following limb trauma when having the limb placed in a cast. They also hope to identify specific cellular and molecular changes in sensory neurons following peripheral nerve injury to better understand the processes that underlie neuroplasticity (the brain’s ability to reorganize or form new nerve connections and pathways following injury or death of nerve cells). This could provide targets for new drug therapies that could improve recovery following regeneration. Last, they are studying children with CRPS to investigate neuroplasticity and the biological processes that cause CRPS to occur, with the hope of developing more effective therapies and accelerated recoveries for both adults and children.5

For now, however, life has to go on for CRPS patients despite the pain. “I’m 21 years older than when I found out about my disease,” says Deborah. “But, I’m also 21 years wiser. I use my time better even though it takes me longer to do everything. Although I know life would be great if I didn’t have pain or need to do weekly three-plus-hour infusions, I’m perfectly happy being me. And, for those times that get me down, I have skills to help me climb right back out!”

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

References
As a registered nurse (RN), Laura Brinton is accustomed to dealing with illness every day. But, when she was diagnosed with common variable immune deficiency (CVID), the tables were turned. Rather than allow the diagnosis to derail her career, Laura has embraced it as an opportunity to provide more compassionate care.

Laura Brinton was diagnosed with common variable immune deficiency in her mid-20s, shortly after finishing nursing school.

Laura was referred to a hematologist due to a very low platelet count. At the time, I also suffered from severe anemia, which was bad enough to warrant a bone marrow biopsy, and that is where the journey began.

Trudie: When were you diagnosed with CVID?
Laura: I was officially diagnosed with CVID in October 2012, but I had always been sick as a teenager into my mid-20s (I'm currently 25). I was referred to a hematologist due to a very low platelet count. At the time, I also suffered from severe anemia, which was bad enough to warrant a bone marrow biopsy, and that is where the journey began.

Trudie: How long have you been an RN?
Laura: I have been an RN for two years. I started with the adult population in an ICU stepdown unit, but later got moved into my passion: working with children. My decision to become a nurse was because I have a passion for people, and healthcare has always been of great interest to me.

Trudie: How did your diagnosis impact your career?
Laura: I was not diagnosed with CVID until after I finished nursing school. After I started working, I began getting very sick with respiratory infections and gastrointestinal (GI) bugs. At one point, I had to take a break and step down from my position to get back on track with my health. Then, I was hired as a pediatric solid organ transplant/renal/GI/endocrine nurse at a major Children’s Hospital. Unfortunately, that was the month when my diagnosis came to light. During a series of tests, my hematologist drew my antibody levels (IgG and all subclasses) and discovered all of them were severely low and that my best chance of staying well would be with monthly intravenous immune globulin (IVIG) infusions. I honestly had never heard of CVID until I was diagnosed.

Trudie: What challenges have you faced as a nurse with CVID?
Laura: Where to begin with this one! I have been told by different doctors that I should leave the healthcare field. Call me stubborn, but I was not put on this earth to give up on my dreams. I worked too hard in school and graduated nursing school with honors. I was asked at one point: “What is your back-up plan?” My answer was simply: “I do not have one, because this is what I have been called to do, CVID or not.” Of course, infection-wise, it has been a challenge. I do work with children, and sometimes the risk of infection is greater, but I try to take extra precautions regardless of what kind of patient I am taking care of.

Trudie: What happens when you become ill on the job?
Laura: It really hurts me to my core that I have to miss work more often than others. I have an intermittent FMLA (Family Medical and Leave Act) in place (a plan approved for my sick days that legally allows me to keep my job, even with excessive absences), but I try not to use it. I want to be there for my co-workers, and I want to be there for my patients, but sometimes it’s hard for others to understand because CVID is still quite unknown, even in the healthcare profession. I have been trying to advocate and educate as much as possible.
How does living with chronic illness make you a better nurse?

Laura: I may not know the answers to everything, but I can honestly relate to my patients. Working with transplant patients who are immunosuppressed, I know the risks they face. Also, CVID has taken a massive toll on my GI system, and when we see GI patients, I feel like I can listen, give advice, hold their hand and provide better support; I am living in that same situation.

Trudie: What have you learned through this journey?

Laura: Definitely, patience. I do have my bad days when I just wish CVID would go away and let me live without being extra cautious or without pain, but I have to keep going. I not only owe it to my patients, my co-workers and my family, but I owe it to myself. I have also learned that this will be a lifelong journey, and with the use of coping skills and support, it is possible to live a quality life.

Trudie: What advice do you have for other patients?

Laura: My advice for other patients is to be your own advocate. If you are not doing well with your current care, or have a suggestion, speak up! It is so very important. Because I work with doctors and other nurses, I can tell you that sometimes we are not aware that something is not working for you and we may not know unless you tell us. I also encourage patients to do their homework about their diagnosis and ask lots of questions. Do not overwhelm yourself, however, because CVID and everything that comes with it is overwhelming as is.

Trudie: What would you say to patients who feel limited by their diagnosis?

Laura: I say: Go for your dreams! Do not let your diagnosis hold you back. I have had to face many obstacles working in the healthcare field, along with criticism by others who think I should do something else. I say: Turn “I can’t” into “I can, and I will.” Do not let what others believe affect your decisions and dreams — being a nurse is amazing, and I love it!
AFTER 208 SUCCESSFUL intravenous immune globulin (IVIG) infusions, 105 blood draws, intravenous antibiotics, steroids, countless hours of charting, including three insurance changes, two late-night infusions that translated to dinner with our family, one football game, one hockey match (pop and a hotdog included) and two subcutaneous IG training sessions that have ultimately ended her every-four-week encounters with my kids, Caleb and Molly, our beloved home care nurse, Nancy, has officially closed out our kids’ cases. We blame this necessary unpleasantness on the reality that 1) Molly’s violent reactions to IVIG came to an end with the switch to SCIG, and 2) Caleb’s port-a-cath couldn’t undo itself from the wall of his subclavian vein. Two ports out plus twice-weekly SCIG infusions times two, minus a home care nurse equals one very sad family.

Nancy didn’t just leave us broken-hearted with a sharps container ready for shipping. Having a homecare nurse isn’t just about teaching you how to change out your loved ones’ colostomy bag or administer heparin through a PICC line; it’s the real life lessons they teach. So, in order to honor the 10 years of blood, sweat and many, many tears (along with a few other misguided body fluids) nurse Nancy has endured with our family, please allow me to reminisce about some of our more stellar moments recorded in the pages of IG Living. Just maybe, you and your nurses will see yourselves in the paragraphs to follow!

Slobberin’ Succotash

It’s no secret that we are voracious dog lovers in the Haggard household. In fact, our dogs are an integral part of our family’s ability to bounce back from illness, keeping our sense of humor intact and, for sure, our humility in check. So, when our neighbors came to us asking if we’d be willing to watch their bassett hound, Emmie, for the weekend, we jumped at the chance. Emmie was the perfect house guest until it came to IVIG day.

“So, who is joining us today?” nurse Nancy asked in her best “poochie” voice, while rubbing Emmie’s massive floppy ears. I couldn’t believe what I was seeing because Nancy didn’t normally baby talk to anyone or anything for that matter, let alone allow the dogs to be around while accessing the kids (something about an IV start and a border collie gone terribly wrong).

Minutes later, we found ourselves in the living room ready for Caleb’s access, with Emmie in a spot on the couch ready to comfort him like she was born to do. Like a well-oiled machine, Caleb’s shirt came off exposing his port, next the Tegaderm removal, which can be worse than the needle poke, and finally the screaming, but this time it came from me and Nancy, not from Caleb!

“Emmie! Stop licking that!” and “Oh my gosh, Emmie, STOP!!!!” and “Emmie, get away from him!”

But, it was too late. The damage was done and the battle won. Emmie...
had successfully licked Caleb’s numbing cream (aka EMLA) clean. It seemed that not only did basset hounds have a strong sense of hearing, but tasting, as well.

The battle of “Emmie and EMLA” might have been over, but the war had just begun; Emmie began drooling uncontrollably — everywhere and on everything. A minefield of dog drool-soaked paper towels littered our house, and after about an hour of nonstop dog spit, Nancy and I decided to call the vet.

Come to find out, Emmie was going to be OK; a little numb, but no lasting damage. As far as Caleb, his infusion went off without any more sloppy interruptions, not to mention he’d made peace with being poked. Nancy and I made out unscathed, as well — a little emotionally disturbed, but, all in all, we chalked up the (very wet) experience as another day in the trenches of warfare against the sworn enemy: immune deficiency.

Life lesson learned No. 1: Don’t underestimate the healing power in a (slobberly) kiss from a four-legged friend. After all, all’s fair in love and “drool.”

Vitavitaveggi-what?

I wish I had a dollar for every well-meaning person who offered advice about how to keep our family healthy. Some advice pays for itself in the humor it provides. Some ideas are generic and tolerable like Gummi vitamins. And, some are outrageously hilarious, like mixing high-quality soil/dirt for the runs. After 16 years of IVIG for my three loved ones diagnosed with PIDD, I am growing very weary of unsolicited advice. Except when it comes from my nurse, of course.

The best part of having Nancy is the conversations we have over the five to six hours during the kids’ infusions. We solve all the world’s problems and even some of our own. Nancy has been like a second mom to me — keeping me grounded while the kids have been seriously ill, calling me out on my personal poop, and encouraging me during a difficult battle with our insurance company. In the past 10 years, we’ve talked about the loves in our lives and the devastating blows of loved ones’ deaths. On our infusion-day chats, we laugh, cry, eat yummy snacks and drink “frou-frou” coffee. And in 10 years, we have become close friends. I can tell her anything, except my true feelings about vitamins.

“So, I was hoping we could talk about this new thing I’m trying out,” Nancy said, trying to distract me from baking cookies.

“Oh, yeah?” I added, cracking an egg into the batter. “Whatcha up to now?”

Before I knew it, Nancy had laid out on the kitchen table all these materials, glossy photographs and computer-generated bar graphs and charts. While I was readying one cookie sheet for the oven, Nancy started to share about the cottage business she just started, selling … wait for it … vitamins. I acted like I was giving her my undivided attention. Little did she know that I may have looked calm on top of the water, but below I was paddling awfully fast.

One of the things I like to do for Nancy is have a fresh IG Living ready for her to flip through; I like to hear about her favorite writer who shall remain anonymous. The only problem plaguing me at the moment wasn’t a lack of chocolate chips for my cookies. No, I had bigger issues on my hands called my IG Living column ragging on people peddling pills in multilevel marketing schemes.

“Snacks are ready!” I announced. I had miraculously manipulated the situation enough to distract Nancy with chocolate and butter. Infusion time was over, and I’d successfully avoided the unavoidable!

Come to think of it, though, she never did tell me about the multilevel marketing thing she got involved in some eight or nine years ago. Hmmm…?

Lesson No. 2: Contrary to popular belief, nurses cannot be bought, bribed or bullied; it’s the one with the most needles who wins.

In conclusion, I thought I’d keep it nice and simple and to the point because that’s the way nurse Nancy likes to run things. It’s a good game plan, because in homecare nursing, less really is more. Well, maybe except for one very important thing, and that’s the love homecare nurses inherently bring with them every time they step past the threshold of not just our homes, but our hearts.

Nurse Nancy, life lesson No. 3 I dedicate to you: Homecare nurses may administer medicine, but they also give us their hearts. Ten years ago, you became our homecare nurse with orders for IVIG. Now, you move on and leave us not just as a friend or nurse, but as part of the Haggard family.

P.S.: Don’t forget our lunch date next Tuesday. See you then!

**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.
Parenting:
Day Care for PIDD Kids

Because of the rapid spread of germs in day care centers, placing PIDD kids in them is a difficult decision.

By Mark T. Haggard

THINGS ARE DIFFERENT today than when I grew up in the 1970s. I was fortunate to have a father who made enough money so my mother didn’t work outside the home, and I could usually count on her being there for me after school. I was not privy to the life of day care, and neither were many of my friends. Today, it takes two incomes to raise a family, and the number of parents enrolling their children in day care, either for financial survival or for personal growth, has increased significantly. But, for parents of children with a primary immune deficiency disease (PIDD), enrolling them in day care is a difficult proposition. Many day care facilities are germ factories, placing PIDD kids at increased risk of infection.

Why Infections Spread in Day Care

Young children have immature and inexperienced immune systems, so they acquire nearly every pathogen to which they are exposed. As a result, when an infection is introduced into the day care setting, close physical contact among both children and adults allow it to make the rounds of the entire room. Infections that spread by the oral route easily pass from child to child as toddlers suck their thumbs after touching contaminated surfaces or as babies teethe on the same toys. Most day care centers have strict standards for proper handwashing and diaper-changing hygiene, but situations can occur that may interrupt this preferred technique and result in pathogens reaching children.

Once an infection appears, transmission is extremely rapid. According to Dennis Clements, MD, PhD, chief of primary care pediatrics at Duke Children’s Hospital, studies have shown that when a marked virus is introduced on a toy in a day care room of toddlers in the morning, it can be cultured from 80 percent of the children by the end of the day and 50 percent of their parents by the next morning. Generally, infants and toddlers in day care have a new viral infection about every three weeks to four weeks and have symptoms of illness about every two months. These repeated infections may not allow these children’s physiology to return to its normal stasis, making some children prone to chronic infections.

The frequency of illness and the potential of secondary bacterial complications make children in day care more likely to require antibiotic treatment. This raises the chances that antibiotic-resistant organisms will emerge, which complicates treatment. Fortunately, infant immunizations protect against the most dangerous bacterial infections. And, healthy kids’ immune systems will strengthen more quickly as they come into contact with more and different types of pathogens. This is not true, however, for those with compromised immune systems, which is the case with PIDD kids.

Protecting PIDD Kids in Day Care

My family’s first experience in a day care-like setting was when we placed our first-born in the nursery at our church. The sweet ladies who ran the nursery called our son “mulcos” (booger boy) because of the green gunk that discharged from his nose.
And, this occurred in only the course of a one-and-a-half-hour church service. The money we paid for preschool was essentially wasted since he was only there for half of the year. Once he was diagnosed with PIDD and prescribed intravenous immune globulin (IVIG), things began running more smoothly. Because our daughter was diagnosed earlier, her preschool experience was much better.

PIDD kids receiving monthly IVIG infusions are actually better off than healthy children in day care centers because the therapy provides greater protection from pathogens. This doesn’t mean, however, that they won’t be susceptible to infections. And, they will likely be most susceptible to infections during the week prior to their next monthly infusion, because the amount of IG circulating in their bodies is significantly decreased, causing their immune systems to be weaker.

Fortunately, there are ways to better protect PIDD kids from acquiring infections. Dr. Clements and other immunologists advise parents to choose the smallest day care group possible. With a smaller number of children, there are fewer sources from which PIDD kids can acquire an infection. Immunologists also advise making sure that day care employees practice frequent hand-washing techniques such as before and after diapering and after helping children blow their noses. The onus is also on parents to teach their children to wash their own hands and use hand sanitizer between activities.

Parents should also look for day care facilities with strict policies concerning allowing sick children to attend. Or, they can consider facilities that are specifically for kids with illnesses. For instance, Morton Plant Mease Hospital, part of the Baycare Health System in Clearwater, Fla., operates Rainbow Recovery, a sick-childcare program. Get Well Place at Rainbow Station is a franchise with nine sick-childcare programs in Virginia, North Carolina and Texas. While these types of facilities are rare, some hospitals offer similar programs, but they don’t advertise them.

The Cost of Staying Home

My wife and I chose to live on a single income, even before our kids were diagnosed with PIDD. It was in our best interest for her to stay home; we are an immune deficient “Leave It to Beaver.” That doesn’t mean my wife doesn’t work. When the kids are sick, she is their nurse, pharmacist, chauffeur, agent and advocate. When we occasionally fall behind on bills, she is our liaison to our creditors. Her work at home is more difficult than my work as a high school teacher.

A Difficult Decision

The decision to place PIDD kids into day care is a difficult one for parents. While PIDD kids who are receiving IG will fare much better in terms of combatting the bevy of pathogens in a day care center, sending immune deficient children into a germ factory is a recipe for disaster. Ultimately, the doctor is one of the best sources for making this decision. If day care is not an option, life on a single income is difficult, but not impossible. And, just think: Clipping, sorting and organizing coupons is a great way to teach youngsters how to use scissors and practice critical thinking skills while saving a few bucks. Then, add graham crackers and ice-cold milk, and you’ve just spent a few hours in day care without acquiring the creeping crud.

Parents should also look for day care facilities with strict policies concerning allowing sick children to attend.

Five people living on the income of one teacher is not easy. We have had to forego numerous things in life. I am still driving a car from the last century, hoping that it reaches 300,000 miles. We eat a lot of peanut butter and jelly sandwiches. We don’t eat out much. We don’t go to ballgames. We don’t travel, except to doctor appointments and an occasional Immune Deficiency Foundation conference. We are grateful for each windfall. Our kids have had the opportunity to play in the school band, in the school orchestra and in high school sports. Beyond that, they have not had the same experiences as other adolescents. But, as they have matured into young adults, we have concluded that they are better people for it.

MARK T. HAGGARD is a high school teacher and football coach, and has three children, two of whom have CVID. He and his wife, Cheryl, also operate Under the Hood Ministries at www.underthehoodministries.org.
I FOUND OUT early on that the shower had the best acoustics for singing. I had many a great performance in the shower. I would belt out everything from show tunes to the latest pop song. Shower time was my special time to be a star. Stepping into the shower stall was like stepping into a magic chamber where I could dream. One of my favorite songs was the version of “Blue Bayou” by Linda Ronstadt. At the tender age of 10, I tried to convey all that deep yearning that Ronstadt portrayed.

The recent news of Ronstadt being diagnosed with Parkinson’s disease is what triggered those shower memories. It also made me wonder why I had stopped singing in the shower. I couldn’t remember when showering became just functional, but it saddened me that I had lost that part of me that could be so free. Then it occurred to me: I had never really lost that part of me at all. I sing full blast while I’m driving! The location has just changed. All these years, the car has been my concert hall. The idea people might be watching has never come into my head (truth is, I wouldn’t care if they did see me). The important thing is I haven’t lost that uninhibited part of me to express myself.

Sometimes I feel my chronic illnesses have quieted certain aspects of my personality and sense of self. Lupus makes me feel anxious and nervous, and I’ve become very conscious of what I say. My multifocal motor neuropathy and immune globulin treatments make me want to retreat and hide from my body. I want to escape from all that’s physically going on that breaks me down or gives me pain. At times, I feel held back and not able to capture that playful, loose feeling of letting go.

In my 20s, I used to sing at bars, clubs and coffeehouses. Other creative endeavors called to me, and I moved on to pursue them. Recently, I got the itch to sing in front of a live audience again. Like Ronstadt, I sing other people’s works. I find great joy from interpreting others lyrics and making their poetry my own. This past fall, I put a little band together to do a show. After over a decade of not performing for audiences, it was heaven to feel everyone’s energy again. It only seemed right to honor Ronstadt, now a fellow neurological and autoimmune sister, so I sang “Blue Bayou” for the first time to an audience.

Maybe I really took to heart the lessons I learned from watching Sesame Street all those years ago such as when I heard the song lyrics: “Sing, sing a song, sing out loud, sing out strong, make it simple to last your whole life long. Don’t worry that it’s not good enough for anyone else to hear, sing, sing a song.” We all need to bring that lightness back into our lives. What did you do freely and lovingly as a child or younger adult that you might miss doing today in your life? It’s never too late to reconnect with that part of yourself. I may never perform live again, but I’ll always have my car concerts. That’s music to my ears!

STACY OLIVER was diagnosed in 2008 with multifocal motor neuropathy (MMN). She is the assistant director of the Center for the Writing Arts at Northwestern University, and she is working on her supersecret identity as Neuropathy Girl, who will one day save the world after her infusion and a nap.
IG Chronicles

Friendship: The Best Thing in Life

By Dona Darr

THIS IS A STORY of friendship. True friendship. One that has never wavered no matter the circumstances. One that has endured many illnesses and cancellation or rescheduling of activities. A friendship that when one is hurting, be it physical or emotional, the other is hurting. This is about my daughter and her best friend and what they have taught me through the years.

They were only about 3 years old without a care in the world when they met in preschool and became the best of friends. They couldn’t wait to get to school so they could play together. They were inseparable.

As luck would have it, they were in the same kindergarten class. Their friendship blossomed so much so that it was determined going forward that it might not be a good idea to have them in the same class together. It was heartbreaking to them at the time, but we laugh about it now. It seems they were a bit too chatty.

Now, they are both 10 years old, fifth-graders and beautiful young ladies. Believe it or not, they are still as close as they were then. They are more alike now than they were when they were little. They enjoy many of the same things, mostly computers and Nintendo DS. They admire each other’s individual abilities. They watch out for one another and take care of one another. When one is having an issue, the other is always there to help. When shopping, “Do you think she will like it, too?” is often the question. Another common question is: “Can she go with us?” or “Can we have a sleepover?” As you can probably already tell, I don’t just have one daughter, I have two, and I love every minute of it.

My daughter’s relationship with her friend can teach people many things. Through my daughter’s journey with primary immune deficiency disease (PIDD), their friendship has endured. Although disappointment does happen when plans need to be changed, there is understanding and forgiveness. When sickness is at hand, there is compassion. When there are delays due to doctor appointments, there is patience. Most of all, there is empathy for the disease that is a part of not just one, but both of their lives.

We should all learn what these girls have been modeling since they were 3 years old:

• Be accepting of one another.
• Be caring to one another — not only in words but also in actions.
• Be considerate of another person’s situation.
• Be understanding and not judgmental.
• Most of all, love one another.

God made us all; He just made some of us a little different than others.

PIDD doesn’t just affect the individual who is diagnosed; it affects their friends and family as well. My daughter and her friend have learned that it is better to accept one another for who they are and to love each other in spite of the disease that makes them different. If all people could be as understanding as these two are with one another, the whole world would be a better place.

DONA DARR is the mother of Emily who was diagnosed with IgG subclass deficiency and complement deficiency. Dona and Emily have been dealing together with this disease since 2004, when Emily was initially diagnosed. Dona and her support system of family and friends will continue to care for and encourage Emily for the rest her life.

This blog is reprinted with permission from Dona Darr’s blog at donadarr.blogspot.com. This blog also appeared on IG Living’s blog on December 5, 2013, at www.IGLiving.com/blogengine.

Patients who rely on IG therapy have unique life experiences. If you have a story you’d like to share about your adventures, experiences, relationships, reminiscences, self-portrayals, etc., for publication in this column, submit it to editor@igliving.com. All submissions must be 600 words or fewer and can be accompanied by high-resolution photos.
Life as a 20-Something

Five Rules for Revealing Your Chronic Illness

By Ilana Jacqueline

IN MY MIND, I have a secret identity. When I meet new people, I try to stay cool and act normal. But, underneath my dress and cardigan, there lies a ridiculously good-looking body — wrapped up in a Holter monitor and a hospital gown.

When getting to know new people in your life, it’s important to remember not to take off your dress and cardigan at the dinner table — even if you do feel antsy about getting your secret identity as a patient off your chest. Breaking the news about chronic illness is a delicate process — one that takes thought, precision and almost no partial nudity. Check out these five rules for the big reveal:

1. Reveal that you deal with an illness on a need-to-know basis. Don’t slap a classified tag on the summary of your medical woes. But, understand, that telling some people in your life about the tumultuous issues related to your chronic illness won’t benefit you.

Before revealing your situation, ask yourself: “Is the topic of my disease bound to come up between us eventually?” “Do I have any obvious signs of this disease that they might start to build their own (and possibly incorrect) assumptions about?” “Will this person be affected by how this disease affects me?” “Last, ask yourself: “Do they care?” You don’t need a pat on the back and someone to call you their personal hero every time you say “I have a chronic illness.” But, if someone in your life has already expressed some kind of prejudice against someone with disabilities or seems to otherwise be incredibly tactless, it might just be better to let that person figure it out (or not) on his or her own.

2. Prepare an elevator speech. Close your eyes, and imagine that nobody wants to sit and listen to you drone on about your aches and pains for an hour. Now, open your eyes. Surprise! That’s the truth! Between surgeries, symptoms, medications and flare-ups, chronic illness can seem like it’s hungrily taking over 98 percent of your life. By going on and on about it, you’re effectively handing over that remaining 2 percent of non-disease-related moments. If you have to talk about the disease, do it in the form of an elevator pitch, or a short summary used to quickly define a situation. By memorizing and reciting this speech, you’ll help minimize the impact of your troubles, while helping to educate new people. Having a speech defining the disease can also help you personally separate your disease from your personality. Once the elevator pitch is over, you can get off on the right floor and start talking about who you really are.

3. Don’t overshare. You don’t need to reveal the details of your sensitive stomach, the current color of your snot or your barely-healed laparoscopy scars. All of these nice stories may have your listener internally screaming “Waaaayyyy too much information!” Save it for your friends in med school who are no longer shocked by any function of the human body.

4. Be the reaction you want to have. People are going to follow your lead when it comes to the state of your disease. If you’re frustrated about it, they’re frustrated about it. You’re sad about it, they’re sad about it. But, if you’re cool about it, they’ll be cool about it. If you haven’t come to terms with your condition yet and are still in a phase of mourning your old life, you may not be ready to reveal your experiences (and open yourself up to the judgment of others) just yet. You’ve got to love yourself — with or without the disease — if you’re going to expect someone new in your life to do the same.

5. Make it common knowledge. Most adult conversations don’t cover the topics of why the sky is blue or the grass is green. It’s just common knowledge (or, we don’t care.) What’s easier than having to explain to everyone you know now, have known or will ever get to know that you have a chronic illness? Having the Internet do it! Don’t be “that guy” who twitters about his medications all day long, but mentioning here and there on your social network how you’re fighting, managing or coping with your disease can help break the ice when you see old friends in person, and it can help lower the shock factor with new acquaintances.

ILANA JACQUELINE is a 23-year-old dysautonomia and primary immune deficiency disease patient from South Florida. She’s been writing professionally since 2004 on everything from health and wellness to celebrities and beauty. Her blog www.letsfeelbetter.com is both a personal collection of anecdotes about life with chronic illness, as well as a resource for patients of all ages.
Chronic Resilience is the author’s journey to become a more compassionate caregiver to herself. In the book, she offers everything she’s got about becoming an owner of one’s own diagnosis. She writes about her triumphs, embarrassments and insights about navigating illness, and she provides practical tips to help readers stop pushing themselves so hard, use research to empower — not frighten — themselves, let themselves be pissed, train their troops in how to care for them, cultivate focus and flexibility, find things to be grateful for and focus on what they can do, not what they can’t. The book also includes stories of nine amazing women who exemplify resilience as they each gracefully manage their own chronic illness.

Patient & Family Handbook for Primary Immunodeficiency Disease (5th edition)

This new edition of the Patient & Family Handbook for Primary Immunodeficiency Disease expands the content by another 50 percent to 33 chapters with descriptions of nearly 100 different primary immunodeficiencies (PIDDs). It includes an overview of the immune system and PIDDs to provide a basic description of the components of the immune system and how its defects lead to disease. There are 18 chapters covering the specific details of many of the individual PIDDs themselves. Additional chapters provide general information relevant to the inheritance, laboratory diagnosis, general care and specific medical treatments of PIIDDs, as well as chapters on life management issues for patients of different ages. Important new chapters discuss subjects such as autoimmunity, allergies and infections, stem cell and gene therapy, and innate immune defects. And, there is an enlarged section on phagocytic cell disorders within the chronic granulomatous disease chapter. In addition to the new chapters, all of the existing chapters have been revised and updated with new information, and many have been rewritten.

Honest Medicine introduces four lifesaving treatments that have been effectively treating — and in some cases curing — people for 25 to 90 years: low-dose naltrexone for autoimmune diseases (e.g., multiple sclerosis, lupus, rheumatoid arthritis, Crohn’s disease, etc.), HIV/AIDS and some cancers; the Ketogenic Diet for pediatric epilepsy; intravenous alpha lipoic acid for terminal liver disease and some cancers; and Silverlon for nonhealing wounds. In the book, nine patients whose lives were changed with these lifesaving treatments are profiled. The book’s author is the creator of the blog HonestMedicine.com. She has been a published writer and a public relations consultant for more than 20 years. When her husband, Tim Fisher, became ill in 1990 with a cancerous brain tumor, Schopick became his medical advocate and began writing on topics relating to health and medicine.
EVER HEARD THE joke: “You know you’re chronically ill when … your medical records take up several boxes and have to be brought in on a cart?” We laugh because there’s truth in that statement for chronically ill patients. Yet, coordinating and keeping track of all those medical records can be cumbersome.

It’s now estimated that 80 percent of health data has become unstructured, meaning it doesn’t fit into nice rows and columns. Add to that the volume of records from several specialists and hospitals, and most patients with chronic illness have an enormous load of medical material to juggle. Fortunately, medical imaging technology has improved, providing a less burdensome way of storing all that information.

Electronic Medical Records vs. Electronic Health Records

Many physicians and clinics have begun offering electronic health records (EMRs) for their patients. EMRs contain the standard medical and clinical data gathered in one provider’s office. However, the data stored in EMRs are not easily shared with providers outside of a practice. Should a member of patients’ care teams need access to the data, EMRs oftentimes have to be printed out and delivered by mail.

Electronic Health Records (EHRs), on the other hand, offer a more comprehensive patient history and focus on the total health of the patient. Unlike EMRs, EHRs allow patients’ health records to move with them — to other healthcare providers, specialists, hospitals, nursing homes and even across states.

Better Information Equals Improved Care

Healthcare is a team effort, and shared information supports that. In fact, research has shown that patients who are actively involved with providers in their healthcare delivery process discovered better results when they had electronic access to their medical histories. Because EHRs are single records that include complete, up-to-date and accurate health information, they can help improve the quality and safety of patient care by placing patients in a better position to work with providers to make good decisions about their care, whether during a routine office visit or in a medical emergency. For providers, having instant access to information about patients’ medical histories, allergies and medications enables them to make better, faster and more-informed decisions, which is especially important if patients have serious or chronic medical conditions.

Confidentiality and Security

While there are many benefits of both EMRs and EHRs, there is a valid concern regarding safety. The Health Insurance Portability and Accountability Act (HIPAA) made safeguarding the confidentiality, integrity and availability of patient information a legal requirement. However, operational inefficiencies, compliance issues, identity theft and cyberattacks are still real concerns for both patients and providers. The healthcare industry faces a number of challenges in monitoring and implementing the safety of EHRs. These include combatting issues caused by bad recordkeeping and other technology-related problems that can have serious, devastating results.

Keeping Track of It All at Home

For patients who are uneasy about the safety of EHRs, there are many tools available to help them to both organize their records at home, as well as easily carry those records with them and have access to them any time. A few options include phone apps, cloud storage, remote servers and thumb drives. Patients should explore the many choices and determine which solution works best for them.

ANNABEN KAZEMI is the patient advocate for IG Living magazine.

Sources

**MedXKey**
Developed and reviewed by medical professionals, the MedXKey is a medical alert emergency medical record contained in a small flash drive. It stores up to 2 gigabytes of information such as a living will, medical power of attorney, X-rays and any other documents. The device is waterproof, can be used with a Mac or Windows computer and is readable without passwords.

*MedInfo911, www.medinfo911.com*

**Medi-Chips**
USB Medi-Chips are personal portable emergency medical records that store any type of medical files, including X-rays, CAT scans, EKGs, medications and more. The database software included helps to manage health records and can also hold a copy of a driver license, passport and social security card. It can be worn as a necklace, keychain, bracelet or credit card style. It plugs into any USB port on any operating system and is password-protected with encryption capabilities. It does require a Windows Interface, Mac dual boot or Windows emulation software. The company provides live chat and telephone support 24/7/365.

*PPEMR Inc., www.personalportableelectronicmedicalrecords.com/Home.html*

**iPHER**
The iPHER, an Individual Personal Health Electronic Record, is a small device that plugs into a computer through a USB drive. Each iPHER carries inside the unique Patient Practitioner’s program, a self-contained medical recordkeeping database system that will store the medical records of a single individual, including EKG graphs, echocardiograms, X-ray images, dental records and dental X-rays. Its unique barcode system ensures that the records for one medical facility exactly match the records and definitions for any other facility. The iPHER comes in many storage sizes up to more than 4 gigabytes. Most iPHERs are delivered as "open" devices to allow first responders and healthcare professionals easy access to records in case of emergency or when an individual may be unable to communicate. However, they can be sold with biometric or dual password security.

*Patient Practitioners, www.patientpractitioners.com/ipher.html*

**MyMedicalRecords PHR**
MMRGlobal Inc. provides secure, easy-to-use multilingual personal health records (PHRs) and electronic safe deposit box storage solutions. The MyMedicalRecords PHR enables individuals and families to safely maintain their medical records and other important documents in one central location and instantly access them any time from anywhere in the world using the Internet. Documents, images and voicemail messages can be transmitted and stored using a variety of methods. In the event of an emergency or disaster, medical personnel and first responders can retrieve potentially lifesaving information accessible via a separate emergency login.

*MMRGlobal Inc., www.mymedicalrecords.com/login.jsp*

**Medical Data Alert Talking Watch**
The Medical Data Alert Talking Watch comes with a built-in USB to store medical data. With the medical alert symbol and the "medical data on USB, press play for message" on the watchband, emergency personnel will be alerted to play and listen to a patient’s own 12-second message, as well as to check the USB device for other important information. The USB can store a photo, documents such as a DNR, living will, medical tests, as well as information about medical condition(s), medication(s), physician(s) and more. The watch also features optional hourly announcements and an alarm. It is approximately 9.5 inches long with a rubberized band and a stainless-steel back.

*www.amazon.com/Medical-Alert-Talking-Built-In-Optional/dp/B00GDBCEQS*

**My Cloud**
My Cloud allows patients to store, organize and back up photos, videos, music and important documents all in one place. Personal cloud access with the My Cloud app requires a My Cloud, My Book Live, My Book Live Duo or My Net N900 Central with the most recent firmware. Access to cloud services requires the My Cloud app and an active Dropbox, Google Drive or SkyDrive account. My Cloud works on an iOS iPhone or iPad running versions 5.0 or later software, or an Android smartphone or tablet running versions 2.3 or later software.

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

- Advocacy for Patients with Chronic Illness: www.advocacyforpatients.org
- American Autoimmune Related Diseases Association (AARDA): www.aarda.org
- American Chronic Pain Association (ACPA): www.theacpa.org
- Band-Aides and Blackboards: www.lehman.cuny.edu/faculty/jfleitas/bandaides
- eMedicine from WebMD: emedicine.medscape.com
- FamilyDoctor.org: www.familydoctor.org
- Johns Hopkins Medicine: www.hopkinsmedicine.org
- KeepKidsHealthy.com (pediatrician’s guide to children health and safety): www.keepkidshealthy.com
- Mayo Clinic: www.mayoclinic.com
- National Committee for Quality Assurance (detailed report cards on health plans, clinical performance, member satisfaction and access to care): www.ncqa.org
- National 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.csblehring.com
- Grifols: www.grifolsusa.com
- Kedrion: www.kedrionusa.com
- Octapharma: www.octapharma.com

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

G eneral Resources

Ataxia Telangiectasia (A-T)

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

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

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

Evans Syndrome

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

Guillain-Barré Syndrome (GBS)

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

Idiopathic Thrombocytopenic Purpura (ITP)

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

Kawasaki Disease

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

Mitochondrial Disease

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

WEBSITES
- The Neuropathy Association: www.neuropathy.org

Multiple Sclerosis (MS)

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

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

Myasthenia Gravis (MG)

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

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

Myositis

WEBSITES

- International Myositis Assessment and Clinical Studies Group: www.niehs.nih.gov/research/resources/collab/imacs/main.cfm
- Michigan Immunodeficiency Foundation: www.facebook.com/groups/108048062584350
- Myositis Association Community Forum: tmacommunityforum.ning.com
- Myositis Support Group: www.myositisupportgroup.org
- Myositis Support Group – UK: www.myositis.org.uk

Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)

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

Pemphigus and Pemphigoid

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

Peripheral Neuropathy (PN)

WEBSITES
- Neuropathy Action Foundation: www.neuropathyaction.org

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

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

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

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

Education and Disability Resources

- Rainbow Allergy-Immunology: www.uhhospitals.org/rainbow/services/allergy-immunology
- Team Hope (for families and patients in New England): www.teamhope.info

Online Peer Support

- IDF Common Ground: www.idfcommonground.org
- IDF Discussion Forum: http://idffriends.org/forum
- IDF Friends: http://idffriends.org
- Jeffrey Modell Foundation Message Board: www.info4pi.org

Scleroderma

WEBSITES

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

ONLINE PEER SUPPORT


Medical Research Studies

- ClinicalTrials.com: www.clinicaltrials.com
- ClinicalTrials.gov: www.clinicaltrials.gov
- World Allergy Organization: www.worldallergy.org

Product Information

- Influenza and the influenza vaccine: www.cdc.gov/flu or call (800) CDC-INFO: (800) 232-4636

Medical Research Studies

- IVIG Flebogamma 5% DIF and 10% DIF: www.grifols.com/portal/en/US/bioscience?div=8883
- IVIG/SCIG Gammmagard Liquid: www.gammmagardliquid.com
- IVIG Gammmagard S/D: www.baxter.com/patients_and_caregivers/products/gammmagard_sd_5.html
- IVIG/SCIG Gammmaked: www.gammaked.com
- IVIG Gammaplex: www.gammaplex.com
- IVIG Privigen: www.privigen.com
- SCIG Hizentra: www.hizentra.com

Pump and Infusion Sets Websites

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

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.stiffpersionsyndrome.net

Other Resources

- IVIG/SCIG Gammmagard: www.gammmagard.com
- IVIG/SCIG Gamunex-C: www.gamunex-c.com
- IVIG Gammmagard: www.baxter.com/patients_and_caregivers/products/gammmagard_sd_5.html
- IVIG Privigen: www.privigen.com
- SCIG Hizentra: www.hizentra.com

Pump and Infusion Sets Websites

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

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

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- Coagulation Products
- Hyperimmunes
- Albumin
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