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About IG Living
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Improving Patients’ Lives with IG Therapy

OVER THE PAST decade, health-related qualify of life (HRQOL) assessments have become an increasingly used tool by physicians for measuring patients’ physical, mental, emotional and social functioning to make treatment decisions. Recently, a team of researchers released the results of a study that measured HRQOL in patients with primary immunodeficiency disease (PI), which found that, compared with normal controls, PI patients “experience measurably lower general health with higher hospitalization rates and increased physical, school and social activity limitation.”

Can you imagine, then, what the findings would be without access to lifesaving immune globulin (IG) therapy? Both clinical and anecdotal studies show that IG greatly decreases the number of life-threatening infections for PI patients. Indeed, it has been found that with IG therapy, the proportion of PI patients surviving 10 years after diagnosis is 93.5 percent, a similar statistic to the population at large. But IG therapy is made possible only with the millions of plasma donations made each year. In our article “Saving Lives Through Plasma Donation,” we provide an update on the rising rates of plasma donation. We also take a look at how manufacturers are making improvements to increase plasma yield, including a dramatically improved fractionation process and expansions of U.S. fractionation and plasma donation centers. Alongside all this are increased safety measures employed at both the federal and industry levels that are greatly improving the outlook of IG therapy for patients.

In this issue, we highlight three different conditions that have been shown to be effectively treated with IG therapy. In our Immunology 101 column, Dr. Terry Harville discusses the characteristics of DiGeorge syndrome, a type of PI that occurs during fetal development. In our Clinical Brief column, we examine the ways in which IG therapy is being used to treat patients with neuromuscular diseases such as chronic inflammatory demyelinating polyneuropathy, multifocal motor neuropathy and dermatomyositis. While typically patients with neuromuscular diseases are administered IG intravenously, subcutaneous administration is now being examined as a way of improving HRQOL by diminishing side effects and providing patients more independence with their treatment regimens. And, in our article “Understanding Scleroderma,” a group of autoimmune diseases that affects the skin and, in many instances, major organs of the body, we look at new research being conducted that shows IG therapy shortens outbreaks of skin lesions and improves joint function with less pain. A current clinical trial is also examining the effects of IG on the gastrointestinal system and lung involvement.

Scleroderma and neuromuscular diseases are just several of the many diseases for which IG is prescribed “off-label,” meaning the drug has not been approved by the U.S. Food and Drug Administration for those indications. In our article “On-Label vs. Off-Label Drug Prescribing,” we discuss how drugs are approved, what the legal and ethical issues are of prescribing them off-label and, while there are risks associated with this type of prescribing, how the result is often better care for patients.

As always, I hope you gain insight from the information presented and enjoy this edition of IG Living.

Ronale Tucker Rhodes, MS
My Role as Patient Advocate

By Abbie Cornett

MOST PEOPLE UNDERSTAND what the job of a police officer, doctor or accountant generally entails. But when I say “I am a patient advocate,” I know I have to be ready for the next questions: “What is that?” and “What do you actually do?” It’s not surprising that not everyone has heard of this specialty profession. Usually, only people with serious life-threatening diseases or chronic illnesses and their family members have worked with one.

In today’s healthcare climate, patient advocacy is a developing field. Historically, patients themselves, along with their family members or caregivers, were primarily responsible for ensuring they received the care they needed. And while they can still fill this role, it is becoming increasingly difficult to do on their own. Therefore, the role of the professional patient advocate has evolved to help patients and their families navigate the increasingly complicated and often broken healthcare system. This includes coordinating care, health insurance benefits, reimbursement issues, and employment and legal problems, among other things, to help patients achieve the best outcomes possible.

Because patients are challenged by multiple issues affecting their access to care, the role of the patient advocate has grown to address many aspects of healthcare. For instance, many patient advocates work with state and federal lawmakers to change policies by enacting legislation that affects access to care. While this type of advocacy is typically removed from direct contact with patients, it is very important to the quality of care and access to care that a patient receives. Other patient advocates are hired by patients and their families to help them file insurance claims and appeals, attend doctor visits, help coordinate care and find legal services if needed. Such advocacy is important because it can relieve a great deal of stress from patients and their families.

My current role as patient advocate for IG Living focuses on:
• keeping you informed of policy and legislative issues that can affect your access to care;
• developing and maintaining good working relationships with multiple experts in the fields of healthcare, insurance and legal issues who can act as resources for providing guidance to answer your questions;
• always protecting your privacy.

While the role of patient advocate may take different forms, it is important to remember that anyone can be an advocate. The primary and most important quality of a good advocate is the desire to help those in need and a willingness to put the patient first.

As IG Living’s patient advocate, I am committed to working with you and your family to help you navigate our complicated healthcare system. Please contact me at any time if you have questions or need help.

ABBIE CORNETT, patient advocate for IG Living magazine, can be reached at (800) 843-7477 x1366 or patientadvocate@IGLiving.com.
New IVIG Product Gets Positive Results

I read with much excitement your article in the June-July issue about the ADMA Biologics RI-002 study. I had the great pleasure of being a participant in this study. The drug is phenomenal! I am a patient with common variable immunodeficiency and have been receiving intravenous immune globulin (IVIG) infusions for about 11 years. I have been on every IG medication during those years. When I participated in this study, I was able to take the infusions more than three times faster than usual, did not have any side effects and did not get sick. I was in the study for about 14 months. I am living for the day when the U.S. Food and Drug Administration (FDA) approves this drug. I am not the only one within this study who had such positive results. I have been told that several other patients, both adults and children, responded very favorably to this medication. I firmly believe that medications work differently for different people based upon their chemical makeup. I am currently being treated with Gammaked, which is the last drug I had not tried. I hope the FDA understands the importance of having RI-002 approved and what it would mean to many patients. If I could be of any help in advocating for RI-002, count me in!

— Rida Hernandez

Patient-Doctor Communication

What great tips for physicians and patients [in your October-November 2014 article “Improving Patient-Doctor Communication!”] Coincidentally, I have started writing about the special set of circumstances that patients with rare/chronic illness face when we work with young doctors (interns, residents). [My focus is] on how their stress level — which is high enough as it is — is compounded by their lack of experience dealing with patients [with rare chronic illnesses], and why a role reversal is oftentimes necessary to achieve good results. [We] as patients … have to become the educators and walk a fine line in doing so.

Spread the Word About HSCT

I just wanted to reach out and say thank you for doing an absolutely great article on hematopoietic stem cell transplantation (HSCT) in your October-November 2014 issue with Jamie Stewart (Let’s Talk, pp34-35). This is a good thing; let’s continue to get the word out about the benefits of HSCT. My husband also suffers from chronic inflammatory demyelinating polyneuropathy and, like Jamie, he will have an opportunity to go to Russia.

Reader Responds to IG Living Blog: Is an Infusion Port Right for You?

After three years of intravenous immune globulin (IVIG) and seeking to find a vein, I got a port. I’ve had it for 15 months and have had no problems. I only wish I had listened to the wonderful nurses who kept advising me to get a port sooner. These ladies and gentlemen really do have our best interests at heart.

The Editor replies:

This blog can be read at www.igliving.com/BlogEngine/post/Is-an-Infusion-Port-Right-for-You.aspx.

Today is That Dreaded Day!

Again that long day is here … The day I wish would disappear The day that is always a huge intrusion The day I get that necessary infusion.

Why do other days fly by But this one is met with a sigh Is it because of the long hours Over which I have no power?

I can watch TV and maybe even eat Or curl up in my chair and sleep But the hours still pass so slow Because I cannot tolerate a faster flow.

One drop follows another, It really seems like quite a bother But what often goes unsaid … Without it I’d be dead.

So I will continue to do this infusion That is an obvious conclusion And instead of being a moaner I should be thanking the donor … That allowed me to have this special day.

— Joy Kiser
Please thank those who donate!

If it weren’t for donors, I would not be walking today. Thank You!
— Brandy G

I want to write thank-you notes to each donor. [It’s] the best gift.
— Linda G

I’ve been a recipient now for 13-plus years. I am so appreciative and thankful!
— Maria FH

Would you try an experimental drug?

[I’m] not sure. I already feel like a lab experiment with most of my doctors. I have many side effects from all the medications I have to take. Treatments require three extra medications to counteract side effects, and I have awful side effects from those. It’s a vicious cycle, and I don’t want to risk adding to it.
— Deb K

Of course I would if there was a chance it could improve my quality of life!
— Thomas R

How do you choose your hospital?

I choose mine on [the] experience of others. I want to go where the doctors actually care; a lot of doctors don’t seem to care anymore.
— Terri S

I choose a hospital by where my doctor works and by my previous experience.
— Heather M

I was choosing my hospital [based] on where my doctor has privileges. Having a hospitalist and the endless stream of doctors that don’t know you is quite stressful and often frustrating.
— Judy S

[I chose the] location because of where we live; it is quite a drive to get to any other hospital.
— Robin P

[I chose mine] because the doctor I needed to see was there. He is one of the top lymphoma specialists in the world.
— Debbie K

[I chose mine because it is] local, where my specialist is practicing. I also look at out-of-town ratings of both the hospital and specialists. [I] love how Cleveland Clinic works together with many specialists as a team for accurate diagnosis and a treatment plan.
— Mindy P
**Michelle** » The Affordable Care Act has not changed the IVIG reimbursement policy for MG. In general, payers are reimbursing IVIG only for crises. It used to be that when MG patients had a crisis, the IVIG prescription was written for six months or one year, and payers would approve that. However, for the past year or more, payers stopped reimbursing for more than one course of IVIG because no literature states it is appropriate for maintenance therapy. Prednisone and immunosuppressants are supposed to work as maintenance therapy for patients in between crises, and most of the time, they do. But, in certain patients, those medications are not enough, or they are not tolerated. Unfortunately, those patients are bearing the brunt of the new policy that allows only one course for exacerbation of MG.

**Michelle** » It is my understanding that medications for MG are stopped before trying to conceive, and I’m not aware of routine use of IVIG instead of other disease modifiers before getting pregnant. Relapses decrease even without medications during pregnancy, and medications are started again postpartum unless you plan to breastfeed. I have seen a lot of requests for IVIG from nursing mothers with MS during the postpartum period to decrease the likelihood of relapsing. However, it is usually difficult to get this covered by a payer. And, because there isn’t much information about IVIG pre-pregnancy, getting it covered while you are trying to conceive would be even more challenging. Nevertheless, you should talk to your neurologist about this, and if it’s something you both want to pursue, an effort can be made to get it covered. Just know that it will be tough. 

**Abbie** » Here are links to some websites that can help you find financial assistance for your son:

- Financial assistance for private school tuition: [www.babycenter.com/0_how-to-get-financial-assistance-for-private-school-tuition_64647.bc](http://www.babycenter.com/0_how-to-get-financial-assistance-for-private-school-tuition_64647.bc)
- Grants to pay for private elementary, middle or high school tuition: [grantspace.org/Tools/Knowledge-Base/Individual-Grantseekers/Students/Funding-for-private-elementary-secondary-school](http://grantspace.org/Tools/Knowledge-Base/Individual-Grantseekers/Students/Funding-for-private-elementary-secondary-school)
- School and student services by the National Association of Independent Schools: [sss.nais.org/parents](http://sss.nais.org/parents)
HYQvia, the only once-a-month subQ Ig*1
For adults with primary immunodeficiency

Schedule an appointment with your physician to see if HYQvia is right for you.

* subQ Ig, also known as subcutaneous immune globulin.

Reference

Please see the Detailed Important Risk Information on the adjacent pages and the Brief Summary of HYQvia Prescribing Information, including Boxed Warning, on the reverse side.

To learn more about HYQvia, visit www.HYQVIA.com
A second site can be used at the discretion of the physician and patient based on tolerability and total volume.

**INDICATION AND USAGE**

**HYQVIA** ([Immune Globulin Infusion 10% (Human) with Recombinant Human Hyaluronidase]) is the only once-a-month subQ Ig with recombinant human hyaluronidase (hy• al•Rou•en•i•dase) and Ig. The hyaluronidase temporarily opens the subQ space, allowing a larger amount of Ig to reach the subQ tissue and be absorbed into the bloodstream to help fight infection. It’s the reason you can infuse your monthly dose of HYQVIA using 1 needle, 1 infusion site, 1 time a month.

These are not all the possible side effects with HYQVIA. Talk to your healthcare professional about any side effects that bother you or that don’t go away.

**What is the most important information that I should know about HYQVIA?**

- HYQVIA can cause blood clots.
- Call your healthcare professional if you have pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s), unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness or weakness on one side of the body.
- Your healthcare professional may perform blood tests regularly to check your IgG level.
- With your consent, your healthcare professional may provide blood samples to Baxter Healthcare Corporation to test for antibodies that may form against the hyaluronidase part of HYQVIA.
- Do not infuse HYQVIA into or around an infected or red swollen area because it can cause infection to spread.
- Talk to your healthcare professional if you become pregnant. Women who become pregnant during HYQVIA treatment are encouraged to enroll in the HYQVIA Pregnancy Registry by calling Medical Information at 1-866-424-6724.

**Detailed Important Risk Information**

**HYQVIA can cause serious side effects. Call your healthcare professional or go to your emergency department right away if you get:**

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of swelling in your brain.
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of swelling in your brain.
- Pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s). These could be signs of a blood clot.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.
- Chest pain or trouble breathing, blue lips or extremities. These could be signs of a lung problem.
You may be eligible to save up to $4,000 on HYQVIA

If you are starting or currently receiving treatment with HYQVIA (Immune Globulin Intravenous 10% (Human) with Recombinant Human Hyaluronidase) for PI, you may be eligible to save up to $4,000 on your deductible/co-payment/co-insurance costs over 12 months.

To enroll, call us.
We’ll take care of the rest.

Terms and Conditions

- To be eligible, patients must: 1) be starting or receiving treatment with HYQVIA with an IgG9 or IgG10, as applicable, for adult (≥16 years of age) Primary Immunodeficiency (PI); and 2) have commercial insurance that covers medication costs for HYQVIA treatment and allows for co-pay/coupon assistance.
- This manufacturer coupon program is not valid for prescriptions reimbursed, in whole or in part, by Medicaid, Medicare, Medicaid, VA, DoD, IRCAH, or any other federal or state healthcare programs, including state pharmaceutical assistance programs, and where prohibited by the health insurance provider or by law.
- The coupon program provides a maximum benefit of $4,000 for eligible out-of-pocket costs and expires 12 months from date of activation. Eligible costs include deductible, co-payment, and co-insurance costs for HYQVIA. Non-medication expenses, such as ancillary supplies or administration-related costs, are not eligible.
- Patients are eligible for a maximum benefit of $4,000 in total Baxalta support in any 12-month period, including any amount received as part of the GAMMAKARI ALUMINIUM SODIUM CO-Pay Program.
- Acceptance of this offer must be consistent with the terms of beneftis provided by patient’s health insurance provider.
- Offer limited to one card per person and expires 12 months from date of activation and may not be combined with any other coupon, discount, prescription savings card, rebate, free trial or other offer.
- This program is only valid for residents of the United States, excluding Puerto Rico and other U.S. territories.

What are the possible or reasonably likely side effects of HYQVIA?

After HYQVIA infusion a temporary, soft swelling may occur around the infusion site, which may last 1 to 3 days, due to the volume of fluid infused. Mild or moderate pain, redness, swelling, or itching may occur at the site of infusion and generally go away in a few hours.

Local reactions are less likely after the first few infusions. The most common side effects of HYQVIA are headache, fatigue, nausea, fever, and vomiting. Antibodies to the hyaluronidase component of HYQVIA were formed in some patients taking HYQVIA. It is not known whether there is any long-term effect. In theory, these antibodies could react with your body’s own PH20. PH20 is present in the male reproductive tract. So far, these antibodies have not been associated with increased or new side effects.

What is HYQVIA?

HYQVIA is a liquid medicine containing immune globulin and recombinant human hyaluronidase. HYQVIA contains IgG antibodies, collected from human plasma donated by healthy people. The antibodies help your body to fight off bacterial and viral infections. The hyaluronidase part of HYQVIA helps more of the Immune globulin get absorbed into the body to fight infection.

Before starting HYQVIA, tell your healthcare professional if you have or had any kidney, liver, or heart problems, a history of blood clots, because HYQVIA can make these problems worse. Also tell your doctor if you have IgA deficiency or a history of severe allergic reactions to immune globulin (IgG) or other blood products, or are pregnant, trying to become pregnant or are breast feeding.

How should I take HYQVIA?

HYQVIA is infused under the skin (subcutaneously) up to once every 4 weeks. You can get HYQVIA at your healthcare professional’s office, clinic, or hospital. You can use HYQVIA at home. You and your healthcare professional will decide if home self-infusion is right for you. Do not use HYQVIA at home until you get instructions and training from your healthcare professional.

Who should not take HYQVIA?

Do not take HYQVIA if you are allergic to IgG, hyaluronidase, or other blood products, or have IgA deficiency with antibodies to IgA.

To report suspected side effects, contact Baxalta US Inc. at 1-866-888-2472 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.
Please see Brief Summary of HYQVIA Prescribing Information on following page, including Boxed Warning.

Baxalta and Hyqvia are trademarks of Baxalta Incorporated
August 2015 US/BS/MSB8/I4-01613Q2
More free time with HYQVIA®

Infusing 1 time a month with HYQVIA doesn’t mean your infusions will take longer. Typically, infusions take less than 3 hours with HYQVIA [Immune Globulin Infusion 10% (Human) with Recombinant Human Hyaluronidase]. Instead, you’ll have more free time.

Please see the Detailed Important Risk Information on the adjacent pages and the Brief Summary of HYQVIA Prescribing Information, including Boxed Warning, on the reverse side.

INDICATION AND USAGE
HYQVIA [Immune Globulin Infusion 10% (Human) with Recombinant Human Hyaluronidase] is an immune globulin with a recombinant human hyaluronidase indicated for the treatment of Primary Immunodeficiency (PI) in adults. This includes, but is not limited to, common variable immunodeficiency (CVID), X-linked agammaglobulinemia, congenital agammaglobulinemia, Wiskott-Aldrich syndrome, and severe combined immunodeficiencies. Limitation of Use: Safety and efficacy of chronic use of recombinant human hyaluronidase in HYQVIA have not been established in conditions other than PI.

Selected Important Risk Information about HYQVIA

HYQVIA can cause blood clots. Call your healthcare professional or go to your emergency department right away if you have pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s), unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness or weakness on one side of the body. These could be signs of a blood clot.

Do not use HYQVIA if you are allergic to immune globulin (IgG), hyaluronidase, or other blood products, or have IgA deficiency.

These are not all the possible side effects with HYQVIA. Talk to your healthcare professional about any side effects that bother you or that don’t go away.
Brief Summary of Prescribing Information
HYQVIA [Immune Globulin Infusion 10% (Human) with Recombinant Human Hyaluronidase]

The following summarizes important information about HYQVIA (pronounced Hi-Q-via). Please read it carefully before using this medicine. This information does not take the place of talking with your healthcare professional. If you have any questions after reading this, ask your healthcare professional.

What is the most important information that I should know about HYQVIA?
• HYQVIA can cause blood clots.
• Call your healthcare professional if you have pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s), unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness or weakness on one side of the body.
• Your healthcare professional may perform blood tests regularly to check your IgG level.
• With your consent, your healthcare professional may provide blood samples to Baxalta Healthcare Corporation to test for antibodies that may form against the hyaluronidase part of HYQVIA.
• Do not infuse HYQVIA into or around an infected or red swollen area because it can cause infection to spread.
• Talk to your healthcare professional if you become pregnant. Women who become pregnant during HYQVIA treatment are encouraged to enroll in the HYQVIA Pregnancy Registry by calling Medical Information at 1-866-424-6724.

What should I tell my healthcare professional before I start using HYQVIA?
Before starting HYQVIA, tell your healthcare professional if you:
• Have or had any kidney, liver, or heart problems or history of blood clots because HYQVIA can make these problems worse.
• Have IgA deficiency or a history of severe allergic reactions to IgG or other blood products.
• Are pregnant, trying to become pregnant or are breast feeding.

What is HYQVIA?
HYQVIA is a liquid medicine containing immune globulin and recombinant human hyaluronidase. HYQVIA contains IgG antibodies, collected from human plasma donated by healthy people. The antibodies help your body to fight off bacterial and viral infections. The hyaluronidase part of HYQVIA helps more of the immune globulin get absorbed into the body to fight infection.

Who should not take HYQVIA?
• Do not take HYQVIA if you: Are allergic to IgG, hyaluronidase, or other blood products.
• Have IgA deficiency with antibodies to IgA.

How should I take HYQVIA?
• HYQVIA is infused under the skin (subcutaneously) up to once every 4 weeks.
• You can get HYQVIA at your healthcare professional’s office, clinic, or hospital.
• You can use HYQVIA at home. You and your healthcare professional will decide if home self-infusion is right for you.

What are the possible or reasonably likely side effects of HYQVIA?
After HYQVIA infusion a temporary, soft swelling may occur around the infusion site, which may last 1 to 3 days, due to the volume of fluid infused.

The following local reactions may occur at the site of infusion and generally go away in a few hours. Local reactions are less likely after the first few infusions: mild or moderate pain, redness, swelling, and itching.

The most common side effects of HYQVIA are headache, fatigue, nausea, fever, and vomiting.

Antibodies to the hyaluronidase component of HYQVIA were formed in some patients taking HYQVIA. It is not known if there is any long term effect. In theory, these antibodies could react with your body’s own PH20. PH20 is present in the male reproductive tract. So far, these antibodies have not been associated with increased or new side-effects.

Call your healthcare professional or go to your emergency department right away if you get:
• Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
• Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of swelling in your brain.
• Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
• Pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s). These could be signs of a blood clot.
• Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.
• Chest pain or trouble breathing, blue lips or extremities. These could be signs of a lung problem.

These are not all of the possible side effects for HYQVIA. For more information about HYQVIA, go to www.HYQVIA.com. For more information on patient resources and education, please visit www.immunedisease.com.
DiGeorge Syndrome

By Terry O. Harville, MD, PhD

O U R  N E X T  S E R I E S of columns will explore the immunodeficiency and other features of the condition known as DiGeorge syndrome (DGS), which has also been called DiGeorge’s syndrome, DiGeorge anomaly and DiGeorge anomalad (many believe the terms anomaly or anomalad are more correct due to the changes that have occurred in the involved tissues and organs, but most still use the term syndrome).

Angelo DiGeorge was a pediatric endocrinologist who in the 1950s and 1960s noted infants born with characteristic changes in their facial appearance, as well as cardiac abnormalities and low serum calcium levels. The low calcium levels were due to low parathyroid hormone activity caused by reduced-to-absent parathyroid glands, which was caused by reduced-to-absent development of the thymus. (The parathyroid glands form at the ends of the lobes of the thymus.) Thus, face abnormalities, low serum calcium levels, cardiac abnormalities and thymus abnormalities became known as DGS. In infants who survived the cardiac abnormalities, it was discovered they may have had an immunodeficiency, sometimes as serious as that of severe combined immunodeficiency (SCID). This further added to the recognition of the essential role of the thymus for T lymphocyte development — an important feature in DGS.

The complications that lead to DGS occur before the end of the first trimester, with the most problematic issues generally occurring between four and eight weeks of gestation, frequently before the mother knows she is pregnant. DGS is caused when the timing is off during developmental events that are supposed to occur in a very precise sequence at specified time points in the first trimester. Depending on when in the first trimester, and how long the specified order of development is affected, more or less severe changes in the affected tissues that cause the features of DGS may be found. And, while the need for treatment for each feature of DGS can become quite complicated, most agree that the immunodeficiency is ultimately the most relevant feature.

Based on the severity of the immune system involvement, DGS is typically divided into two categories: 1) complete DGS (CDGS) and partial DGS (PDGS). CDGS indicates an essentially complete disruption of thymic development and, as a consequence, absence of development of T lymphocytes. PDGS indicates some level of impairment of thymic development that delays the production of T lymphocytes, but production is typically expected to catch up by about 3 years of age when a complete immunologic repertoire has developed.

In infants who present with all the features of DGS, it is reported to occur in approximately one in 4,000 infants. However, a compilation of studies indicates that nearly one in 13 infants may have some form of congenital abnormality of the heart, most of which are very mild and may heal over time without specific intervention (for example, a pinhole-size ventricular septal defect that closes on its own). Only approximately 0.5 percent of infants have congenital cardiac conditions that may require more in the way of evaluation or treatment. Now, with newborn screening for SCID, many infants are being detected who otherwise do not have SCID but do appear to have fewer T lymphocytes than expected in newborns. Many of these appear to have features consistent with PDGS that were not previously detected since they may not have become ill and they outgrew any delayed T lymphocyte development by about 3 years of age. Thus, milder forms of PDGS may be relatively common but not necessarily with significant immune complications.

In the next issue, we will further describe the timing of the developmental events that cause DGS.

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

Author’s note: The information in these columns about DGS comes from the vast literature on the subject, as well as insight gained while working with Douglas Barrett, MD, who was part of a group from California that, early on, added a vast amount to our knowledge of DGS from the immunologic standpoint; Lodewyk HS (Bob) Van Mierop, MD, and Lynn Kutsche, MD, at the University of Florida, who cataloged the types of cardiac abnormalities along with the thymic abnormalities in DGS, and were able to determine the series of events that lead to DGS; and M. Louise Markert, MD, PhD, at Duke University, who developed the first successful program to use donor thymic tissue transplantation to treat and “cure” the most severe forms of DGS.
Subcutaneous Immune Globulin Therapy for Neuromuscular Diseases

By Michelle Greer, RN

**THE FIRST SUBCUTANEOUS immune globulin (SCIG) treatment** (Vivaglobin immune globulin subcutaneous [human] 16% liquid) was approved by the U.S. Food and Drug Administration (FDA) for the treatment of primary immune deficiency disease (PI) in 2006 (and removed from the market in 2011). Since then, several other SCIG preparations have been approved to treat PI (Table 1). Yet, while SCIG is not approved for conditions other than PI, it is frequently prescribed off-label, and it is being explored in clinical trials and prescribed in clinical practice by some neurologists for certain neuromuscular conditions.

**SCIG Dosing for Neuromuscular Conditions**

SCIG has many benefits, including lower incidence of systemic side effects, elimination of the requirement for venous access, and patient convenience and independence. When PI patients have problems infusing IG intravenously (IVIG), switching them to SCIG is usually the next step. However, for those with neuromuscular conditions, physicians typically take other steps first, such as a brand change, additional pre-medications, central venous access placement and/or adjusting the duration of the infusion.

When the decision is made to prescribe SCIG or IVIG for neuromuscular conditions, volume and dosing are the main factors. Volume, or the amount of the IG preparation being infused, is a consideration whether administered intravenously or subcutaneously. With IVIG, any patient health condition that may involve volume restrictions must be considered. With SCIG, volume impacts site reactions and, in turn, the number of SC sites and needles necessary to successfully infuse.

IG is dosed by weight. For the treatment of PI, IG is considered a replacement therapy. When administered intravenously, it is usually dosed around 400 mg/kg and, on average, given monthly. For example, the dosing for an individual who weighs 75 kg would equate to 30 grams (or 300 milliliters) of fluid. When administered subcutaneously, however, a dose conversion is recommended that will result in additional volume (although, in some instances, one-to-one dosing can be used, which is determined by the physician and the severity of the condition).

For the treatment of autoimmune conditions, IG is considered immunomodulation therapy. With autoimmune conditions, the immune system malfunctions and creates an autoantibody that attacks some part of the body. In neuromuscular disease, the attack occurs on some part of the nerve, muscle or neuromuscular junction. Immunomodulation is used to prevent the immune system from making the autoantibody. How this works is complex and multifaceted, and in many cases, it is not clearly understood. Dosing for an immunomodulatory effect requires more grams than for PI. A typical dose may be as high as 2 grams/kg administered monthly. So, for a 75 kg individual, the dose would be 150 grams (or 1,500 milliliters) of fluid.

When SCIG is prescribed, the physician and/or dispensing pharmacist will determine the rate of infusion and the number of sites needed to infuse. The total volume to be infused determines the number of sites, because only a certain amount of fluid per minute can be infused into any one site. Frequency of infusion is typically weekly, and patients are taught to self-administer.

**Clinical Trials of SCIG for Neuromuscular Conditions**

Currently, there are several clinical trials underway that are examining the efficacy of SCIG treatment for neuromuscular conditions.

The PATH (polyneuropathy and treatment with Hizentra) study is currently active and recruiting in 98 centers in the U.S. and many other countries. This prospective three-arm study is investigating

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two different doses of subcutaneous IgPro20 compared with placebo for maintenance treatment of patients with chronic inflammatory demyelinating polyneuropathy (CIDP). During the trial, patients stabilized on IVIG will be randomized to receive weekly infusions of one or two SCIG doses (0.2 or 0.4 g/kg body weight) or placebo for 24 weeks to measure the proportion of patients who experience a relapse in their CIDP over a period of 52 weeks.2

Another active trial is evaluating the efficacy and safety of SCIG (Hizentra) in patients with dermatomyositis (DM). At present, patients with steroid-resistant DM can be treated only with IVIG. This study will look at whether SCIG is a suitable replacement by exerting an immunomodulatory effect on complement antibodies. It is being led by Dr. Marinos Dalakas, director of the neuromuscular disease program at Thomas Jefferson University in Philadelphia, who previously conducted studies that show the efficacy of IVIG treatment for DM via complement inhibition. Dr. Dalakas wants to explore how SCIG works in this disease state. In the trial, skin biopsies to look at complement and other inflammatory mediators are being performed to determine whether SCIG is as effective as IVIG, if it is preferred by patients and how it works. There are currently three patients enrolled.

There are many challenges to conducting clinical trials for rare conditions such as these, including getting patient participation and FDA approval for the headache, fever and nausea. In addition, some patients with clotting or renal problems may do better with SCIG than IVIG. However, larger studies to prove these differences still remain to be completed."

Currently, there are several clinical trials underway that are examining the efficacy of SCIG treatment for neuromuscular conditions.

**The SCIG Alternative**

SCIG appears to be a viable route of administration for some patients with neuromuscular conditions. While it is currently being prescribed off-label, many other therapies are also prescribed for conditions before being approved by FDA, including SCIG for PI. SCIG can offer alternatives when there are difficult side effects, challenges with venous access or simply the desire to be more independent with treatments. It is hoped, then, that the positive outcome of the clinical trials investigating the efficacy of SCIG for neuromuscular conditions will lead to FDA approval.  ■

MICHELLE GREER, RN, is senior vice president of sales for NuFACTOR Specialty Pharmacy.

**References**

In a small study of six patients, abatacept (Orencia) had a major positive impact on three patients with common variable immunodeficiency (CVID) who were treated for five or more years, and it appeared particularly effective at reversing the devastating lymphocytic interstitial lung disease caused by CVID.

The discovery of the use of abatacept, a drug already approved for treating rheumatoid arthritis, for treating CVID stemmed from an enhanced understanding of immune system protein regulation. The researchers’ key insight was that the CVID-associated deficiency in cytotoxic T lymphocyte antigen (CTLA4) was the result of a flaw in how CTLA4 is recycled between the cell surface and internal lysosomes in regulatory T cells. Normally, CTLA4 is expressed on the cell surface, where it blocks activation of responder T cells and thus prevents autoimmune activation. Periodically, CTLA4, along with other cell surface molecules, is removed from the T cell surface and swept into recycling endosomes, where the lipopolysaccharide-responsive and beige-like anchor (LRBA) protein grabs it and recycles it to the cell surface. However, the researchers discovered that patients with CVID are deficient in LRBA and, thus, lack the mechanism for replacing CTLA4 on the cell surface.

“We were quite surprised that it worked as well as it did in terms of improving interstitial lung disease (clearing chest [computed tomography] scans, improving lung function),” said Michael B. Jordan, MD, from the Cincinnati Children’s Hospital Medical Center, University of Cincinnati, Ohio. “We were also surprised by the apparent decrease of infections in the three patients treated longer term. We fully expected that they might experience increased infections (this is still certainly possible in other patients), but these frequently ill patients had an overall decrease in common infections after being on this medication. We don’t understand why, but perhaps this relates to the cessation of other medications such as corticosteroids that might have been contributing. Also, one patient looked as if she was developing hypogammaglobulinemia around the time we started this medication, but despite all indicators, she has not done so over the ensuing four to five years. Perhaps we averted the development of this complication seen in most (but not all) LRBA-deficient patients.”

The study was published in the July 24 issue of Science.
Research
New Genetic Immune Disorder Is Identified

An international team of researchers has identified a new immune disorder, DOCK2 deficiency, which they named after the mutated gene responsible for the disease. The disorder was discovered in a study of four boys and one girl from different ethnic backgrounds who had been diagnosed with combined immunodeficiency (CID) after experiencing debilitating infections early in life. CID refers to a group of inherited disorders distinguished by defects in T cells, although it may also affect other cells of the immune system, including B cells. When the researchers sequenced the children’s genomes, they discovered mutations in the DOCK2 gene that ultimately cause this particular CID. In lab tests, T cells and B cells from the five children had impaired ability to move in response to infection-related stimuli, and antiviral responses were impaired in many cells. Three of the children were successfully treated with bone marrow transplants, which replaced the defective immune cells with those of a healthy donor.

According to the researchers, this finding demonstrates the importance of understanding the role of DOCK2 in a healthy immune system. It also demonstrates that early screening for CID to identify patients with DOCK2 deficiency can potentially prevent life-threatening infection early in life. Further, identifying causative genes underlying CIDs such as DOCK2 may enable researchers to develop targeted therapies.


Research
Body’s “Safety Procedure” May Explain Autoimmune Disease

Researchers have found an important safety mechanism in the immune system that may malfunction in people with autoimmune diseases, potentially paving the way for innovative treatments. The research, conducted at Monash University in Australia, described for the first time how the body manages marginal zone (MZ) B cells that form a general first line of attack against germs but are potentially harmful. That’s because MZ B cells have the potential to turn against the body since some are capable of producing antibodies that attack healthy, rather than foreign, cells.

Bacteria trigger MZ B cells whether the cells are dangerous or benign, effectively placing anyone with a bacterial infection at risk of developing an autoimmune disease. “We found that while MZ B cells are rapidly activated, they have a very short life span,” said Professor Fabienne Mackay, head of the Monash Department of Immunology. “In fact, the very machinery which triggers a response leads to MZ B cells dying within 24 hours. This means that in a healthy person, the potentially harmful immune cells are not active for long enough to cause tissue damage. We now need to look at whether a malfunction in this safety feature is leading to some autoimmune diseases.”

When MZ B cells are activated by bacteria, they express greater amounts of a protein known as TACI. When TACI binds to another protein as part of the immune response, this triggers the activation of the “death machinery” inside MZ B cells. The detection of a pathogen sets off a chain reaction that both activates and then destroys MZ B cells. “This says something important about our environment; pathogens are not always the enemy,” said Mackay. “They can also work hand in hand with our immune system to protect us against some immune diseases.”

Autoimmune Corner

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The road to producing lifesaving plasma therapies is long and complicated, made possible only with the millions of donations collected each year.

By Ronale Tucker Rhodes, MS
NEARLY 30 MILLION PEOPLE in the U.S. have been diagnosed with a rare disease,1 a great number of whom have genetic, chronic conditions caused by an inability to produce plasma proteins in sufficient quantities or of sufficient quality. These individuals depend upon access to plasma protein therapies, including coagulation factors used to treat bleeding disorders and immune globulins to treat primary immune deficiencies, neurological disorders and autoimmune diseases. Generally, these therapies are infused or injected throughout life to replace missing or deficient proteins that allow individuals to lead healthy and more productive lives. But sustaining the need for these therapies requires millions of liters of plasma each year that must be donated and then manufactured into lifesaving medicines.

How Plasma Is Donated

Plasma is the pale yellow portion of blood that functions as an aid in the circulation of red and white blood cells and platelets.2 In the U.S., there were more than 32 million donations of plasma collected in 2014, according to the Plasma Protein Therapeutics Association, which is more than triple the increase of 12 million donations over the previous decade.3 Most of the world’s plasma is collected in about 400 plasma donation centers scattered throughout the U.S., with some of it exported to other countries.4

In U.S. plasma donation centers (owned exclusively by plasma therapy manufacturers), donors receive a small financial compensation (between $15 and $40)4 for their time to donate plasma and to incentivize them to return. To donate, individuals must meet certain requirements. They must be 18 years or older (19 years in Nebraska or 18 years with an authorized consent form), weigh at least 110 pounds and be in general good health. When arriving at a donation center, they are required to produce either a Social Security number or an INS number, as well as a valid picture ID (driver’s license or student or military ID) with their current address.5 And, they are checked against the National Donor Deferral Registry (NDDR) before being allowed to donate.6

Donors must also proceed through a process that ensures they are healthy enough to safely donate their plasma. Before the first donation and once a year thereafter, donors must receive a physical evaluation during which their pulse, blood pressure and temperature are taken. They are also given a hematocrit test via a small finger prick to confirm a healthy level of red blood cells, and they must give a urine sample. Questions are asked of them to ascertain if they have participated in any high-risk behaviors or have any medical conditions that may disqualify them from donating.6 For instance, those who have recently had tattoos or body piercing, who have lived for a prolonged period in Europe, and have cancer and/or other medical conditions are ineligible to donate.1 And, in some instances, donors may be asked for medical records from their personal physician.6

New donors are required to donate two times. The second donation provides two sets of test results and health screenings to assure the safety and reliability of the plasma supply. If new donors donate only one time, their plasma donation is discarded. Plasma can be donated two times within a seven-day period with at least 24 hours between donations because the body replenishes the donated plasma within 24 to 48 hours.5

A newly developed patented technique is expected to greatly improve upon the plasma extraction rate.

Once all the requirements are met, donors are ready to begin plasmapheresis, a process that separates each donor’s plasma from the red cells, collects the plasma and returns the red cells to the donor. Once the donation is made, the plasma is placed on a 60-day hold while it undergoes further testing for viral agents. If it is found to be unsafe, it is discarded. Any donors whose plasma tests positive for hepatitis B, hepatitis C or HIV will be automatically entered into the NDDR and will be permanently barred from donating blood or plasma in the U.S.6

How Therapies Are Made

Once the plasma is released from inventory (the 60-day hold), it is ready for fractionation. The Cohn cold fractionation process has been the most common protocol for plasma protein extraction since its inception in the 1940s. During the fractionation process, plasma is pooled from multiple donations, purified and processed in a specific order to extract specific plasma proteins that have a proven health benefit. The steps and regulations required to
collect donated plasma and complete the manufacturing process that ultimately results in the final therapies take between seven and nine months. Between weeks 0 and 4, the plasma is collected. Then, between weeks 4 and 12, it is batched and transported to the fractionation plant, where it is stored from weeks 12 through 16. During this period, a combination of time, temperature, pH and alcohol concentration allows the extraction of the specific therapeutic proteins. At that point, the plasma is inspected and released for production. Production occurs between weeks 20 and 24. Then, between weeks 24 and 28, internal testing of the therapeutic proteins takes place, and the therapies are then released by the U.S. Food and Drug Administration (FDA) and shipped between weeks 28 and 32 to the wholesalers and end users.7

The Cohn cold fractionation process is long and arduous with a moderately successful extraction rate of 7 percent from blood (with plasma concentration of 1.8 grams to 3.5 grams per liter). But, a newly developed patented technique is expected to greatly improve upon the plasma extraction rate. The Optimized Plasma Process from PlasmaTech Biopharmaceuticals increases the plasma extraction yield tenfold, recovering almost 70 percent of plasma.8 As of this writing, it is unknown when this new plasma fractionation process will be available for use.

Even before the development of this new technique, manufacturers have continued to expand their fractionation capacity. For instance, CSL Behring will spend $450 million over the next few years to expand its production facilities in the U.S. and Australia, with a $240 million investment into its Kankakee, Ill., facility, which produces albumin and immune globulin, and a $210 million investment into its Broadmeadows plant in Melbourne, Australia. Grifols has also expanded its three manufacturing sites in Clayton, N.C., Los Angeles and Barcelona, Spain, the company’s global headquarters. The expansions include a new, 185,000-square-foot fractionation facility in Clayton and a new facility in Los Angeles dedicated to the production of immune globulin therapies. These new facilities have increased the company’s capacity to fractionate plasma from 8 million liters per year to 12 million liters in 2015. And, according to a statement by Baxter Healthcare, the company is “in the process of building a new state-of-the-art fractionation and production facility in the U.S., with commercial production scheduled to begin in 2018. In addition, [the company has] established an agreement with Dutch company Sanguin to provide additional fractionation
Ensuring the Safety of Plasma

The U.S. plasma fractionation industry is regulated by FDA. Since the 1980s, FDA and international agencies have developed a comprehensive set of measures to ensure the viral safety of plasma products, including multiple levels of regulatory oversight to ensure overlapping safeguards against the risks of transmitting bloodborne infectious agents. And, during the past few decades, manufacturers have introduced new technologies to further improve the safety of plasma protein therapies. Indeed, donor screening and testing are only the first steps in the complex manufacturing process for plasma products. Each individual plasma product is subjected to multiple purification and removal processes. The type of viral inactivation and removal methods used depend on the plasma product, but common viral inactivation methods include:

- Solvent detergent treatment that consists of adding a soap-like chemical to the plasma that breaks down and destroys the fatty coating surrounding lipid-enveloped viruses. By destroying this fatty coating, the viruses are also destroyed.
- Heat treatment that involves heating each product vial to 80 degrees Centigrade for 72 hours. The temperature is carefully controlled to maintain it at a level that is effective against pathogens but not damaging to the therapeutic proteins.

- Nanofiltration that allows the wanted therapeutic proteins to pass through a specially designed membrane with a reduced pore size, while other particles or pathogens are trapped and discarded.

These procedures have proven to be effective at eliminating a wide array of potential contaminants such as bacteria and viruses, including hepatitis, HIV and many others. It should be noted that there have been no cases of HIV or hepatitis transmission via plasma medicines since the implementation of these validated viral inactivation methods in the early 1990s.

In addition to FDA standards and improved manufacturing processes, many manufacturers adhere to the International Quality Plasma Program (IQPP), a voluntary industry certification program for plasma collection centers to exceed government standards for safety. IQPP guidelines include the exclusive use of repeat donors, 60-day inventory holds and nucleic acid testing for each donation — procedures that have been incorporated industry-wide in an effort to maximize plasma product supply chain safety.

Each individual plasma product is subjected to multiple purification viral inactivation and removal processes.

A Promising Outlook

The outlook for plasma therapies looks very promising thanks to an increasing number of plasma donors each year, which is welcome news as the number of diseases treated with them grows and off-label (non-FDA-approved) prescribing increases. With more plasma, manufacturers are expanding their fractionation production facilities and new technologies are being developed to meet therapeutic demand. What’s more: The plasma supply is safer now than ever with extensive safety measures that eliminate infectious donations and various steps that eliminate and inactivate contaminated viruses, which has all but eliminated the risk of disease transmission by plasma products.

RONALE TUCKER RHODES is the editor of IG Living magazine.

References
First described in ancient Greece by Hippocrates (who called it thickened skin), scleroderma was not given its present name until 1836 by Giovambattista Fantonetti. And it was only toward the end of World War II, in 1945, that Dr. Robert H. Goetz described it as a systemic disease. Today, it is classified as a group of chronic connective tissue autoimmune diseases that affects the skin, joints and, all too often, major organs.

Yet, despite two millennia of awareness of scleroderma, its often disfiguring nature can still cause discomfort — as witnessed this past summer when Facebook declined an advertisement that featured a photograph of a patient’s face. Lisa Goodman-Helfand of Highland Park, Ill., tried to purchase a Facebook ad to promote a website that shares her story of dealing with scleroderma. Goodman-Helfand’s scleroderma has affected the skin on her face, and her photos — taken without the makeup she usually wears in public to hide the signs of the disease — showed her skin covered with bright red and purple splotches. After rejecting the advertisement a second time — and after Goodman-Helfand blogged about the situation — Facebook relented, but not without very effectively highlighting the struggles with which scleroderma patients often are confronted.

What Is Scleroderma?

Scleroderma is broken down into two different classifications; however, both are characterized by an excessive production of collagen that creates a thickening of tissue. It is believed this is caused due to a breakdown of the body’s immune system, with the body’s cells reacting as if there is tissue damage that needs repairing even though there is not.
Localized scleroderma affects the skin only, and usually just a small portion of the skin — often on the hands or face. It may be severe and may leave permanent damage to the skin, it can also go away on its own.

Systemic scleroderma affects not only the skin, but the circulatory system and, often, major organs. It has elements of both rheumatic disease and connective tissue disease. It may affect the heart, lungs or kidneys, with excessive collagen deposits impacting the ability of the body to function normally. In addition, if the muscles in the abdomen are impacted by fibrosis (the formation of excess connective tissue, often resulting in scarring) and unable to move food through the small intestine normally, the ability of the body to properly extract nutrients can be compromised.

What Causes Scleroderma?

The cause of scleroderma is unknown. It is not contagious and is not believed to be hereditary, although it may be at least partly genetic. It is classified as an autoimmune disease because doctors and researchers believe the immune system plays a role, but that is not definitively known. It is thought that a triggering event — an injury or infection — causes the onset of biological reactions that lead to scleroderma. Again, this process is not yet fully understood by doctors or researchers.

For unknown reasons, women are more prone to develop scleroderma than are men (researchers are studying whether hormonal differences between the sexes play a role). Localized scleroderma is more common in people of European ancestry, while systemic forms are seen more often among people of African descent.

In addition, localized scleroderma tends to manifest between the ages of 20 years and 40 years, while systemic scleroderma most often shows up between 30 years and 50 years. Still, the disease is seen in all age groups and all ethnicities.

Symptoms of Scleroderma

Scleroderma symptoms, and whether it is localized or systemic, vary from patient to patient but nearly every case involves a thickening or hardening of an area of skin. These patches vary in size and shape, as well as number and location. They are often marked by a white area in the middle, with purple around the edges, and might appear shiny from the tightening of the skin due to thickening. For those with localized scleroderma, these will likely be the only symptoms.

Those with systemic scleroderma may also experience Raynaud’s phenomenon, which is marked by numbness, pain or change of color in the toes or fingers in response to cold temperatures or extreme emotions. Hands and feet can also become stiff or swollen. As systemic scleroderma continues, it can manifest in other ways, including difficulty swallowing (from scarring of the esophagus due to fibrosis), tightening of the skin on fingers and toes, swollen blood vessels on the face and/or extremities, and even seizures and headaches. Fatigue and loss of appetite are other common symptoms.

For unknown reasons, women are more prone to develop scleroderma than are men.

Diagnosing Scleroderma

Because the above symptoms are also associated with other conditions, it can be difficult to make a diagnosis of scleroderma. In fact, just estimating the number of people who have scleroderma is apparently a challenge, with numbers ranging from 49,000 (the National Institutes of Health) to 300,000 (the Scleroderma Foundation).

A definitive diagnosis will generally be made only after the general practitioner has consulted with rheumatologists, pulmonologists, orthopedists and/or dermatologists. In addition to a full medical history and detailed physical examination, a doctor may also order blood tests and a skin biopsy. Calcium deposits on the joints, changes in the capillaries at the base of the fingernails or the appearance of certain antibodies in a blood sample can all be clues that a patient has a type of scleroderma.

Treating Scleroderma

There is no cure for scleroderma, nor a method to prevent it. Treatment will depend on the type of scleroderma and the severity of the case.
Localized scleroderma (the type confined to the skin) is further divided into two distinct types: morphea and linear. Morphea is characterized by waxy, shiny patches of thickened skin, which can grow in size or shrink, and often disappear with no treatment. Linear scleroderma manifests as a line of hardened, waxy skin, often resembling a scar from a severe cut. (These are sometimes referred to as en coup de sabre due to their resemblance to a sword wound.)

**There is no cure for scleroderma, nor a method to prevent it.**

While there is not currently a way to cure the thickened skin of localized scleroderma, doctors do recommend regular exercise to keep the skin as limber as possible. Nicotine is known to make scleroderma worse, so eliminating smoking or chewing tobacco is important. And moisturizers and analgesic creams can help relieve pain, swelling and stiffness in the affected areas. Sunscreen can help prevent further damage from exposure to the sun.

Systemic scleroderma, affecting more parts of the body than localized, often requires more detailed treatment plans to relieve symptoms and reduce physical damage from the disease.

Systemic scleroderma is also broken down into different subtypes: limited cutaneous systemic sclerosis and diffuse cutaneous systemic sclerosis. Limited cutaneous systemic sclerosis affects larger portions of the skin than does localized scleroderma, but it does not affect major internal organs (although the esophagus can still be affected). It tends to manifest with painful calcium deposits on the joints, swallowing difficulties, tightening of the skin on the hands and feet and swollen blood vessels. Diffuse cutaneous systemic sclerosis can have all the above symptoms, plus include excess collagen on the lungs (the most common complication), heart and/or kidneys.

Treatment for systemic scleroderma is administered with the aim of controlling symptoms and preventing more serious complications. It may include:

- Nonsteroidal anti-inflammatory drugs to control swelling and pain in the joints arising from calcium deposits
- Blood pressure medications, if the heart or lungs are affected
- Medications to reduce stomach acid if the esophagus is affected
- Antirheumatic drugs to reduce joint swelling and damage
- Immunosuppressants to try to slow the body’s creation of excess collagen, particularly in those with lung damage

Patient advocates also point out that any time someone is diagnosed with a chronic condition, emotional well-being is key to a successful treatment plan. A newly diagnosed patient will do better when surrounded by family and friends in a support network.

**Scleroderma Research**

Important advances are already being made in treating the symptoms of scleroderma. For instance, cyclophosphamide has recently been found effective in slowing lung fibrosis. And, this summer, the U.S. Food and Drug Administration approved a clinical trial of Actemra (tocilizumab), a drug being developed by Genentech that slows the progress of fibrosis. If successful, this could provide physicians another tool to help treat the symptoms of scleroderma.

Among the most promising research is the use of intravenous immune globulin (IVIG) in patients who have not responded to immunosuppressants. Researchers in several studies have reported that patients receiving a regimen of IVIG have shorter outbreaks of skin lesions and improved joint function with less pain. Studies in mice indicate that IVIG treatment lessens the production of collagen and lowers the incidence of fibrosis. Georgetown University is currently conducting a clinical trial of 24 subjects to test the effect of IVIG on the skin, muscles, joints, gastrointestinal system and lung involvement. That study will conclude at the end of 2015, with results to be published in 2016.

But research into a cure for scleroderma is proceeding more slowly. The recent discovery of a gene linked to scleroderma among members of the Choctaw Nation has geneticists re-examining the link between genes and the disease. Researchers are also examining the genetic blueprint to see if they can predict risk factors so doctors might know which type of scleroderma is most likely to develop after an initial diagnosis. Others are looking at how and why the immune system in women works differently than in men, as well as the biochemistry behind fibrosis.

Because the specific mechanisms that cause scleroderma are not fully understood, an immunization against scleroderma is not likely any time soon.

**Scleroderma Outlook**

Since there is no cure or any preventive treatment, the prognosis
for those diagnosed with sclerodema varies according to the severity of each case. Patients diagnosed with localized sclerodema should see no difference in their lifespan based on the disease, although their quality of life may be diminished depending on its severity and how they respond emotionally. Those with systemic sclerodermia may face more serious consequences, depending on what organs are affected and how severely.

The overall five-year survival rate for those with diffuse cutaneous disease is about 80 percent, and for those with limited cutaneous systemic sclerosis, it is roughly 90 percent. Those numbers can vary widely, however, based on whether there are cardiac, pulmonary or renal complications. Younger patients also tend to face more serious prognoses, as do those of African descent. In addition, the more widespread the skin lesions, the more severe the prognosis.

On an optimistic note, all survival rates associated with sclerodema have shown marked growth over the past few decades as our ability to treat the symptoms and reduce organ damage has improved.15  ■

References

JIM TRAGESER is a freelance journalist in the San Diego area.
On-Label vs. Off-Label Drug Prescribing

FDA approval of drugs is based on what they are going to be used to treat, cure or prevent. But physicians regularly prescribe medicines for conditions not specified on the label, and while that may come with some risks, in many cases, it provides patients with better care.

By Heather Claverie

When patients are prescribed a drug to treat an ailment, most assume the drug has been approved for that use. However, one-fifth of drugs prescribed in the U.S. are for a use not approved by the U.S. Food and Drug Administration (FDA), which is known as an “off-label” prescription. Yet, while off-label use of a drug such as immune globulin (IG) may cause concern for some, it is both legal and, in most instances, beneficial.

FD A Approval Process: How a Drug Is Designated On-Label

FDA was established by the Federal Food, Drug and Cosmetic Act of 1938 to tighten controls over drugs and food, protect consumers against unlawful cosmetics and medical devices and enhance the government’s ability to enforce the law. FDA’s authority was further expanded when President Kennedy signed the Kefauver-Harris Drug Amendment into law in 1962 to strengthen the drug provisions of the Federal Food, Drug and Cosmetic Act. Thanks to those pieces of legislation, all over-the-counter and prescription medications sold in the U.S. must undergo a rigorous approval process overseen by FDA.

A pharmaceutical drug’s journey to approval begins when the sponsor, usually the manufacturer or marketer, gathers all collected data, including the results of animal tests and proposed labeling, and submits a new drug application or biologics license application to FDA. The results of the animal and toxicology studies must show that the drug is reasonably safe before it can be tested on humans. The manufacturing and labeling information must illustrate that the company will be able to adequately produce the drug. And, the application must include proposed clinical studies.

Before beginning any trials, a 30-day waiting period is instigated. During that time, FDA reviews the application for any safety issues and ensures individuals participating in the trials aren’t subject to unreasonable risk. Once approved, the company may begin clinical trials. If FDA runs into any issues concerning the drug, it may interrupt clinical trials or delay investigation into the proposed medication.

When the company has completed testing, it sends all data to FDA’s Center for Drug Evaluation and Research (CDER). A team of CDER physicians, statisticians, chemists, pharmacologists
and other scientists reviews the data and proposed labeling and
then decides if the drug is safe and effective in its proposed use
and if its benefits outweigh its known risks. If all boxes are
checked, FDA will approve the drug, and the company may
begin marketing it to the public.4

Off-Label Prescribing

FDA’s role is to ensure the quality of products in the U.S.
marketplace so that physicians and patients can rely on them to
treat a given indication. Once a drug is approved and released
into the market, FDA doesn’t play a part in defining the drug’s
standard of care or how it is prescribed. For patients, that means
a drug approved to treat depression may end up being prescribed
to treat chronic pain. A physician prescribing a drug exactly as
approved by FDA is doing so on-label. When a physician veers
from that path, he or she is prescribing a drug off-label.

Just because an indication isn’t listed on a drug’s approved
label doesn’t mean FDA disapproves of an off-label use. Rather,
the agency just hasn’t reviewed that use. While prescribing off-
label can be challenging for physicians, they have the discretion
to do so; however, they are not free to promote the off-label use.5

Drugs that are commonly prescribed off-label include antide-
pressants, antipsychotics, immune globulin (IG), chemotherapy
and pediatric medications, among others. “Despite having many
on-label drugs available, so often our patients will exhaust treatment
options,” said Dr. Wade Smith, medical oncologist for Breastlink
Orange and Temecula Valley centers. “Sometimes, off-label use
is very effective, especially in the area of cancer care where existing
options may be limited. Cancer providers sometimes make clinical
decisions that are not based on FDA approval so long as there
exists a supporting study with necessary validation.”

For instance, when Rituxan (rituximab) first came on the market
in 1997, it was approved for the treatment of non-Hodgkin’s
lymphoma. Since then, the drug’s role in treating diseases has
increased to include a variety of conditions, from blood disorders
such as hemophilia, chronic diseases such as Sjögren’s syndrome
and rheumatoid arthritis and other B-cell lymphomas.6 “If you
had to pick one drug to say, ‘Look at all the directions this
biologic has gone,’ rituximab would be the one to choose,” said
Dr. Smith. But, rituximab is just one drug among a long list of medications often prescribed to treat conditions for which they’re not approved. Today, one in five prescriptions falls under these so-called off-label designated drugs. 7

**The Number of Off-label Uses for IVIG Far Exceeds Those Approved On-label.**

Many prescriptions in pediatrics are written off-label because children are less likely to be included in clinical trials. For instance, children afflicted with pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS) present with a whole host of symptoms, from obsessive compulsive disorder (OCD) and anxiety to tics, depression and behavioral issues. When physicians are faced with such a wide range of disorders, they will turn to studies showing that certain drugs have proven effective in treating one or more of them. Risperdal Consta (risperidone), which is approved to treat bipolar 1 disorder and schizophrenia, is one of the drugs physicians may prescribe for the tics associated with PANDAS. In addition, various antidepressants may be prescribed for OCD, and Ativan (lorazepam) may be prescribed to stabilize the mood swings that are often brought on by PANDAS. 8

Some PANDAS patients are prescribed intravenous IG (IVIG) because studies have revealed it is effective for treatment of OCD and tics associated with the disorder. 9 In fact, the number of off-label uses for IVIG far exceeds those approved on-label. When prescribed on-label, IVIG is approved to treat just a handful of ailments, including primary immunodeficiency, chronic lymphocytic leukemia, idiopathic thrombocytopenic purpura, Kawasaki disease, multifocal motor neuropathy and chronic inflammatory demyelinating polyneuropathy. Yet, more than 150 diseases are treated with IVIG. Some of the most common include myasthenia gravis, dermatomyositis, polymyositis, inflammatory necrotizing myopathy, pemphigus and pemphigoid.

**Controversies of Off-Label Prescribing**

When Terry Gross-Morton injured her foot at work, she had no idea the next six years would involve countless doctor visits and result in a cabinet chock-full of prescription medications. Initially, physicians weren’t sure what was causing the severe burning and stinging pain in her foot or why the area would turn bright reddish purple before spreading to other extremities. Although Gross-Morton wasn’t suffering from fibromyalgia or shingles, she brought home prescriptions for Lyrica (pregabalin) and Neurontin (gabapentin) in an attempt to treat the unknown affliction. “I was willing to try anything that would help [that] my doctor recommended,” she said.

Eventually, she was diagnosed with reflex sympathetic dystrophy (RSD), a disorder characterized by chronic arm or leg pain that develops after an injury, surgery, stroke or heart attack. These days, she treats her RSD with an extensive list of prescriptions for the pain and depression associated with it. From IVIG to Lexapro (escitalopram), OxyContin (oxycodone) and Elavil (amitriptyline), all her medications are prescribed off-label.

Doctors aren’t required to disclose to patients that they’re prescribing a medication off-label, and some physicians aren’t even aware they’re doing so. Although off-label prescribing does have its place, is often effective and, in some instances, can lead to a drug’s accelerated approval by FDA, especially in the area of cancer care, the practice does have its fair share of critics, according to Dr. Smith.

One of the most tragic aftermaths of off-label prescribing was in the 1960s when doctors began prescribing the sedative thalidomide to pregnant women to alleviate morning sickness. It was quickly discovered that the drug caused severe birth defects in the infants born to women who had taken thalidomide while pregnant. At the time, clinical trials in the U.S. did not require FDA approval or oversight. Therefore, thalidomide was distributed to more than 20,000 patients across the nation, approximately 207 of whom were pregnant women. 10 There were 17 births of deformed infants tied to thalidomide in the U.S., according to FDA. This is the incident that led to the passage of the Kefauver-Harris Drug Amendment.

Still, even with strict guidelines now in place, there are times when off-label prescribing raises serious health concerns. In the late 1990s, doctors began prescribing patients a combination of an appetite suppressant called fenfluramine and phentermune, a type of amphetamine. The diet drug combination, coined Fen-Phen, seemed like a miracle weight-loss recipe. Yet, it didn’t last long. In 1997, FDA pulled the drugs off the market after discovering that the combination caused heart valve issues. 11

The fact that FDA oversees the approval of a drug but not the actual prescribing once it hits the market may come as a surprise to some patients. And some may balk at the thought of taking a
medication for a disorder not listed on the drug’s label. But for patients like Gross-Morton who are desperate for a treatment, the pros outweigh any possible cons. In addition to the other medications she takes, she now receives in-home infusions every three weeks, and the treatment is helping. “I felt like I’ve tried every other option but the drastic ones,” she said. “This disease is not widely recognized.”

Prescribing in the U.S.

The control and quality of prescription medications in the United States have come a long way since the early days of FDA. Today, drugs must go through a rigorous process to receive on-label approval by FDA, and those strict guidelines ensure that medications are safe and effective. Even so, physicians are legally permitted to prescribe drugs off-label, and many patients would not receive the best available care without them.

HEATHER CLAVERIE is a contributing writer for IG Living magazine.

References
Depression and anxiety are common feelings, especially for those with chronic illness, but there are therapies that can help treat them.

By Erika Lawrence, PhD
AS IF DEALING with a chronic physical illness were not enough, many individuals also experience symptoms of depression and anxiety. These symptoms can be intermittent or chronic, mild to moderate to severe. Not only can chronic physical illness worsen symptoms of depression and anxiety, but depression and anxiety can worsen physical symptoms and immune functioning. Similarly, depression and anxiety make it harder to have healthy relationships with family, friends and coworkers, and stressful relationships often lead to depression and anxiety.

Am I Depressed or Anxious?
Depression can be a vicious cycle. When we’re depressed, some of us become more annoyed or hostile. Others become numb and withdrawn. Still others become more anxious. It can make us feel like we do not want to do anything, even get out of bed. We may withdraw from others and/or push away the people close to us. It becomes really hard to be there for other people when we’re consumed by depression. We may spend a lot of time focusing on depression-related thoughts — thoughts about ourselves (I am a loser, stupid, a failure), about others (they don’t like me) or about our future (this will never pass, I will always feel like this). Certain things make our depression worse, like ruminating about it (focusing on your negative thoughts) and isolating yourself from others.

Anxiety often goes hand in hand with depression, though not always. Anxiety can be challenging because of the thoughts that come with it (I can’t do this, I will make a fool of myself), the physical symptoms that come with it (heart racing, shallow breathing, muscle tension, chest tightening) and the desire to flee or avoid a situation.

Those who are suffering from depression or anxiety or both need to know they are not alone. Everyone experiences these symptoms at some point in their lives. The problems come about in how we deal with them. Many of us try to avoid these thoughts, feelings or physical symptoms, or avoid the situations in which they arise. Others of us try to control them by trying to “get rid of” negative thoughts, feelings or physical sensations. Unfortunately, it is impossible to get rid of thoughts or feelings. Thoughts and feelings are natural and normal. Trying to get rid of them or control them is like trying not to think about chocolate cake. The more you try to get rid of the thought or avoid the thought, the stronger it becomes. Similarly, the more you try to avoid certain feelings (sadness, anger, fear), the more power they have over you.

A Brief Look at Acceptance and Commitment Therapy (ACT)

ACT is a mindfulness-based, values-oriented behavioral therapy that teaches individuals to accept what is out of their personal control while committing to action to improve quality of life. The therapy uses mindfulness skills, which are a way that a person can deal with painful thoughts and feelings so that they have less impact and influence over them.

With ACT, mindfulness skills are broken down into three categories:

• Defusion: distancing from and letting go of unhelpful thoughts, beliefs and memories
• Acceptance: making room for painful feelings, urges and sensations and allowing them to come and go without a struggle
• Contact with the present moment: engaging fully with the here-and-now experience with an attitude of openness and curiosity

ACT has six core processes:
1) Connection — connecting fully with the present moment
2) Defusion — learning to step back or detach from unhelpful thoughts, worries and memories
3) Expansion — learning to stop struggling with painful feelings and sensations without being overwhelmed by them
4) Observing self — being aware of whatever is being thought of or felt at the moment
5) Values — identifying what matters in life (i.e., what one wants to do with one’s life, stand for, etc.)
6) Committed action — taking action based on one’s values even if it’s difficult or uncomfortable

Combining these six processes allows a person to develop “psychological flexibility,” which is the ability to be in the present moment with awareness and openness, and to take action guided by values.

What Can I Do About It?

So if you can’t control these unwanted thoughts and feelings, what can you control? You can control what you do about your depression or anxiety and whether and how you seek help.

There are two types of therapy that have been shown to be effective for depression. One is known as behavioral activation, which simply means engaging in activities that you enjoy and that you feel you are good at. It also includes spending time around other people who support you and make you happy. The other type of therapy is called acceptance and commitment therapy (ACT). Despite the name, this type of counseling does not mean you should accept that you have depression. Instead, it is designed to help you continue functioning and doing what you need to do despite having feelings of depression or unwelcome thoughts. It has also been shown to be highly effective for chronic pain. For patients who want to try to do something on their own, I recommend either *The Happiness Trap* by Russell Harris. It covers the same ACT material, but you can use it on your own. Reaching out to people online or by phone if you are too physically ill to see them in person can also be helpful.

The best type of counseling for most types of anxiety (phobias, panic attacks) is still cognitive behavioral therapy. However, for those who are chronic worriers, ACT might be a better fit. Mindfulness meditation and relaxation can be great places to start, and patients can learn how to do these exercises on their own at home. For those who want to try something on their own, I recommend either *The Relaxation and Stress Reduction Workbook* by Martha Davis, Elizabeth Robbins Eshelman and Matthew McKay or *A Mindfulness-Based Stress Reduction Workbook* by Bob Stahl and Elisha Goldstein.

**There are two types of therapy that have been shown to be effective for depression.**

**Depression and Anxiety Can Be Treated**

We all experience sadness and nervousness at times in our lives. However, those who are having these feelings and thoughts most of the time, making it more difficult to work or to interact with others, may be experiencing true depression or anxiety. If so, they should seek out help to get assessed and possibly get treatment. Depression and anxiety are both challenges that professionals can successfully treat if given a chance.

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**A Brief Look at Cognitive Behavioral Therapy (CBT)**

CBT is a combination of psychotherapy (emphasizing the importance of the personal meaning people place on things and the thinking patterns that begin in childhood) and behavioral therapy (the relationship between problems, behavior and thoughts). It is a short-term (four to seven months), goal-oriented therapy that takes a hands-on, practical approach to problem-solving.

CBT is based on the theory that it’s not events themselves that upset people, but the meanings people give those events. It differs from many other types of psychotherapies because the typically once-weekly sessions are structured, meaning the individual being treated doesn’t talk freely about whatever comes to mind. Clients meet with a therapist to describe specific problems and set goals that then become the basis for planning the content of sessions and discussing how to deal with them. Homework, a vital part of the therapy, is given between sessions. Homework will vary, but it could include keeping a journal or performing exercises to cope with a particular problem. CBT also differs from other therapies because it favors a more equal relationship between the client and therapist. Known as “collaborative empiricism,” it emphasizes the importance of client and therapist working together to test out how the ideas behind CBT might apply to a client’s individual problems.

CBT can be an effective therapy for a host of problems, including anxiety and panic attacks, chronic fatigue syndrome, chronic pain, depression and general health problems.

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THREE DAYS AGO, I took that long walk across the stage for the conferral of my master’s degree. I had worries similar to those of other students such as: Do I look OK? Will the speaker say my name right? What if I fall on stage? I also had some worries that only come with having a chronic illness: What if I get sick while waiting to go up? What if I get sick on stage? What if my vision gets funky during the long walk? Will I even be able to keep up with the pace of my cohorts? The list goes on and on. The result: I did not fall or get sick on stage.

It was tough, but I managed to keep up with the rest of the graduates. Was it rough? Sure. Was it worth it? Yes! Whenever I accomplish something, small or big, I am so happy and relieved. Accomplishing even the smallest tasks often brings on extra stress and worry, but it makes my accomplishments all the more special.

Getting through grad school is a daunting task for any student. Add a chronic illness to the mix, and the more daunting it gets. The good news is there are some steps to make the daunting doable.

1. **Determine your strengths and weaknesses.** What are you especially good at, and what do you struggle with? I found taking a strength-based approach when picking out a graduate program and school was the best option. I am structured, enjoy writing and am rather independent. My weaknesses included extreme fatigue, constant sickness, difficulty with reading comprehension and an extreme dislike toward mathematics. I needed the flexibility of an online school so I could rest and recover when needed and keep up with treatments. Since I am very structured and enjoy writing, this worked out well. Some students might benefit more from a traditional school depending on their strengths and weaknesses. A pros-and-cons list could
be helpful. A final note on picking out a program that is right for you is to ensure the program is actually something you will be able to make a career out of when you graduate. Depending on your disability, some fields might be more suitable than others.

2. Research, research, research. When you think you have researched a school or program enough, research it some more. This is grad school; it’s hard, it’s expensive and it’s going to take over your life for the next few years. Make sure it has the right accreditation and that the program fits your wants and needs, and check out the tuition and financial aid. Contact the disability office before you enroll to find out its policies. Practice your advocacy skills. Determine what the office will need to accommodate you. Ask about how it has accommodated other students. Perhaps the school has had a student with your same illness/disability in the past? Go through your list of trouble areas to see what the school can do. Get everything in writing. The school I attended required two out-of-state residencies. Before enrolling, my counselor told me that they could find me a local alternative or try to work something else out. When it came time for the residency, no such proof of that conversation existed, and the school would not waver. It was tough, it was long and it was painful, but I got through it. Lesson learned: Have everything in writing.

3. Learn self-discipline. Regardless of the school you choose, you will need to keep on top of the curriculum and keep up with grades. Many graduate schools require that you maintain a B average. You will have a lot of schoolwork and a lot of required readings. You must schedule in time to complete all of this work. With a chronic illness, you will need to put aside more time to complete the work and have plans in place for when you are sick. You must be flexible and willing to make some sacrifices.

4. Ask for help when you need it. Help might mean someone to watch the kids for an hour while you write a paper, or someone to help you when you are feeling stuck. With chronic illness, we need to ask for help from others when we are sick so we can take care of ourselves and get well enough to complete our responsibilities. This might also mean asking the professors for guidance. A friend to vent to is also a good idea for any grad student.

5. Be open and honest with your professors. By law, professors may not ask you about your disability. This is something I don’t really agree with. I found it was much easier to let my professors know what was going on so they could help me when I needed it. I explained to all of my professors about my multiple disability status and the issues it causes. I let them know when I might need some extra time to turn in an assignment and why. For example, I recently had to start infusions, which was very difficult to adjust to and I would feel sick for days after each one. I told my professors what was going on, and they were very understanding. At the same time, don’t abuse this. Use these accommodations only when you truly need them. Teachers are more apt to understand when they see that you are sincere and trying your best. If you are late with every assignment, not only will it undermine the professor’s understanding of your situation, but you will fall behind very quickly. As someone who believes that the individual impacts the community, those who abuse accommodations make it more difficult for the disabled population to get what they truly need.

6. Enjoy the experience! You will learn a lot of information in grad school that you will take with you for life. Use this time to learn as much as you can and to make friends and as a stepping-stone to the future you desire. I had some extra challenges, but all in all, I really enjoyed the experience.

Good luck to you as you start this exciting step in your life!

STEPHANIE LAUER is a 28-year-old from Long Island, N.Y., who recently graduated with a master’s degree in clinical mental health counseling. She has been living with common variable immuno-deficiency and other chronic illnesses since she was a child.
PROFILE: Evangeline Garibay

By Trudie Mitschang

Trudie: Tell us about your journey to
diagnosis.

Eva: After a lifetime of minor to major
illness and infections, my health took a
turn for the worse, and I suffered from
anemia, respiratory infections, asthma
and several malignancies in 2011 and
2012. That led to abnormal bleeding, a
blood transfusion, two strokes, double
vision, gait imbalance and several surgeries
and near death twice. It wasn’t until
after all of those events that I was diag-
nosed with an immune deficiency
disorder called hypogammaglobulinemia,
which basically means my immune
system is not producing the disease-
fighting antibodies necessary to protect
my body from infection, illness and
malignancies.

Trudie: What is your current treatm ent
plan?

Eva: In addition to intravenous
immune globulin (IVIG) therapy, I have
been prescribed a pharmacy worth of
prescription drugs. I do believe you have
to know your body and what makes you
feel complete. I’m naturally a very active
woman, accustomed to multitasking, and
I won’t accept any type of medication or
drug that makes me slow, unresponsive
or delayed in thoughts or actions. When
I had high cholesterol, for example, I
refused statins, niacin or other choles-
terol and triglyceride medications.
Instead, I realized that I’d felt my best in
my younger years when I ran marathons.
I decided I needed to create my own diet
plan and started my journey back to
good health. I started walking a mile a
day, and gradually increased it to three
to five miles per day until I reached 20
miles a week. Now, I hold steady at 20
miles minimum a week. Some people
won’t believe this, but the more you
exercise, the better you feel, and the
more active you become.

Trudie: How has exercise helped you?

Eva: I succeeded in reaching my goal
weight and bringing my cholesterol
levels back to normal, and I lost the
headaches and nausea and started
sleeping through the night again. I do
not suggest or recommend anyone going
against their medical regimen, but I do
believe no one knows your body better
than you do. There is no one miracle
drug or magic pill, and we can’t expect a
physician to know every small detail
about what makes us feel good or bad.

Trudie: Tell us about your nonprofit
organization.

Eva: I don’t think I’m dying anytime
soon because I am now 99.98 percent
healthy and feeling great, but we never
know what path life may take. When my
time comes, I want to know that I helped
as many people as possible through my
nonprofit organization called Immune2life
Corp. We offer counseling, group
sessions, medical referrals, tax and insur-
ance advocate services, fundraising events
to help people get the best care possible,
and compile research to share with the
federal government in an effort to
someday find a cure for our children
and grandchildren.

AFTER BEING diagnosed
with half a dozen life-
threatening illnesses and
undergoing several surgeries,
Evangeline Garibay decided
to take her life back one step
at a time by walking a mile
a day. Today, Eva walks 20
miles per week and has
reinvented herself as the
owner of a nonprofit titled
Immune2life Corp. Mindful
of her own challenges
following diagnosis, Eva is
determined to help others
successfully navigate the
path of chronic disease.
**Trudie:** What was your motivation for starting Immune2life Corp.?

**Eva:** When I discovered that my immune disease had no cure, I was very upset. You have to wonder why in a world with so much amazing technology and medicine there is no cure for this illness. I didn’t like being considered permanently disabled due to an immune disorder and hooked up to IVIG for the rest of my life. In respect to that, I wondered how a person with an immune disorder survives in this world with little to no help. For example, a person with an immune disorder might be diagnosed with breast cancer, cervical cancer, bronchitis, a sinus infection, fever, nausea, and migraine all in one week. After treatment, rest and recovery, the cycle begins again in a few weeks with a new set of recurring illnesses, infections or ailments. This would require this individual to receive constant medical care and follow-up, which would make it next to impossible to hold a full-time job.

There was a gap in the system, and I thought I could help fill it.

**Trudie:** What have you been surprised to learn since starting Immune2life?

**Eva:** Because there is no one specific diagnosis to some of these illnesses, they are difficult to detect and hard to prove in a Social Security disability evaluation. Most judges will deny the claims because they don’t understand how immune disease works. They just assume if you become ill and get better, then you’re good enough to go back to work. The Social Security Administration fails to educate their medical experts on these illnesses and, instead, just denies the claims. On top of that, state benefits don’t apply if you own your home, vehicle or have life insurance because you are required to sign a document giving the state the right to take your assets to recover any monies they helped you with. For some patients, that means depending on county hospitals for free medical care because they have no money to pay for their insurance, and they do not qualify for Medicaid unless they want to sign over their homes, vehicles or insurance policies. Where does that leave someone with immune disease?

**Trudie:** Since becoming ill, what has been your biggest challenge, and how have you overcome it?

**Eva:** I have had to make life adjustments, sell my businesses and learn to live and work with the immune disorder. I have to be mindful of preventive healthcare: I avoid crowded or closed-in facilities during cold and flu seasons because someone’s flu would result in pneumonia for me. I wear a mask in public places during the winter, and I wash my hands often. I also use soap and Germ-X like I invented it! In the process, I had to develop new business investment opportunities for myself to help meet my financial needs.

**Trudie:** What advice do you have for others?

**Eva:** There will always be negative individuals out there and people who will demean you by their actions. Don’t take offense to people’s lack of knowledge or ignorance; take the time to educate them. Help them understand that while these illnesses are not contagious, they are often hereditary, and while it may skip a generation, it might show up in the next generation. In my experience, once people begin to understand, they will learn to put the puzzle together on their own, and their criticism sometimes turns to admiration when they realize what you’ve gone through. In the end, I tell people to stay strong, enjoy life and appreciate every day as if there was no tomorrow.

**Trudie:** How do you keep a positive attitude?

**Eva:** I wake up every day thankful to see my family, and when they complain about how bored they are, I plan vacations and trips all over the world because I want to leave great memories behind. We collect a Christmas ornament from every state or country we travel to, and at Christmas, we decorate the tree with all those memories of good times.

**Trudie:** Do you ever feel like quitting?

**Eva:** Often when I have to do my monthly IVIG treatment, I become irritable and I just don’t want to do it. I talk about quitting, and I talk about not needing it anymore, but toward the end of the third week of the month, I start to feel weak and tired, and then I go in and do my treatment, and I feel fantastic and ready to run again.

TRUDIE MITSCHANG is a staff writer for IG Living magazine.

Pictured (from left) are Amanda Del Toro, vice president of Immune2life Corp., Isabel Lopez, grant director of Immune2life, Evangeline’s daughter Ashlee Garibay, age 16, and Jesse Arredondo, marketing director of Immune2life.
PATIENT PERSPECTIVE

Loving Us Is a Full-Time Job
By Ever Fecske Mazza

A WHILE BACK, I was thinking about how difficult it is for us to find people who truly understand all we go through. I have written many times about how lonely it is to be chronically ill. How to cope when not feeling well is often a burden that only we can understand. Our souls and minds are in a body that doesn’t function the way we want it to, and it is frustrating. It’s as if our minds say “go,” but our bodies say “stop.” When how we think and how we feel don’t align, it is isolating and maddening. Then, trying to explain how that feels can truly be an obstacle. I really dislike trying to explain how I feel about my health because of my own insecurities; I don’t want people to think I am complaining or whining. I don’t want people to get the wrong idea and think that I want them to feel bad for me or make excuses for me. I can make those all on my own.

But, then it came to me. I have a handful of people in my life who know exactly what I go through. It may not be on their minds every minute of every day (my health isn’t even on my mind that often). But, when I need something or I am feeling less than human, they get it. The people who get it are the ones who really matter to me, which makes me not care about anyone else understanding because my people have my back.

There are so many instances when my people have been there for me, and I love them for it. Since I started dating my husband 11 years ago, he has probably spent a year of his life waiting for me to come out of the bathroom. One year! No complaints; maybe a chuckle here or there, but he understands that there is often urgency, and he will put his game face on to help me find the nearest restroom. And, then, he waits. Sometimes he waits for a while, but when I emerge, he makes sure I am OK, and we continue on with whatever we were doing — like it’s no big deal. I love him for that, among countless other things.

My sister is one of my people. She is my baby sister; I am five years older. I remember when I took her and some of her friends out for pizza one night while I was in the thick of my prednisone puffiness. As I paid for our order with a credit card, the girl at the register asked to see my ID. I handed over my license with a picture of me when I was 16, thin and learning how to drive. The girl looked at the picture, then back at me, then back at the picture again, and laughed out loud. She said: “This isn’t you. You don’t look anything like this.” I froze. What could I say? I knew it didn’t look like me. In fact, I was unrecognizable. My sister, who couldn’t have been much older than I was in that picture, jumped right in and said: “Hey! That is her. You don’t know what she’s been through. You don’t know her. How about you slide the card, and stop being so rude!” My baby sister was my hero that day. Not everyone has the nerve to always stand up for themselves; I made that painfully obvious. So, the fact that she had my back without any hesitation proved she is one of my people.

It’s easy to think about all those we come in contact with who don’t have any idea what we are going through, and the truth is, they probably won’t even try to understand. It’s not because they are bad people. There are many reasons why people can’t understand what it’s like to be chronically ill. And that’s OK. Over time, I have realized that thinking about those people and dwelling on their lack of understanding isn’t going to change them. I now try to focus on everyone around me who does get it. I choose to surround myself with supportive, kind and accepting people. But, here is the best part: I may have chosen my people, but my people also chose me!

EVER FECESKE MAZZA was diagnosed with CVID and interstitial lung disease in 2004. She is a new mom of a sweet little boy named Boston, and loves every minute of it! She lives in Los Angeles, Calif., with her husband, and when she isn’t changing diapers and playing with her son, she enjoys wedding planning, baking, flower arranging, cooking, shopping and anything that sparkles!
Traveling with an Immune Deficiency? Be Prepared!

By Ilana Jacqueline

WALKING FROM THE couch to the bedroom with needles attached to your thighs or arms is no easy task. So, the thought of traveling across the country with all your medical supplies — IV poles, bags and bottles of medications — may seem overwhelming. It’s difficult getting the nebulizer, Neti pots, tubing and infusion supplies out of the house and into the car, let alone through airport security! Sooner or later, though, you’ll need to drive or fly to new adventures. Follow these tips to help you navigate new terrain:

1. Call the airline’s help line, and ask to speak with someone in security and regulations. Let them know what kind of products you need to take on board with you (and which ones you might need to check). They’re able to accommodate most patients, and they can prepare you ahead of time so you’re not scrambling in the airport security line.

2. Keep all your medications and their labels on your person when traveling. Never check them. If you usually have medications in a pill holder, that’s fine. Just make sure you also take the bottles with you. You’ll want to have proof of prescriptions in case you run into any trouble at airport security. This is also a precaution if you run out of a medication and need to get an extra pill through an unfamiliar pharmacy.

3. Research hotels, cabs and parks. If you’re wheelchair enabled, get tired easily or need grab bars in restrooms, it will decrease some of your anxiety to know if the places you’ll visit will be able to meet your needs. If they can’t, the time to make new arrangements is before you leave.

4. Protect yourself. Traveling usually involves many people in very small spaces, so take precautions to stay germ-free. Carry hand sanitizer, wear a mask if you feel you need it, and wash your hands thoroughly and often. Ask your doctor for additional ways to help protect your immune system before you leave.

5. Use a medical ID card and bracelet. ID bracelets are a good backup in case you have a medical episode in an unfamiliar place. Your ID bracelet should tell people to search your wallet for an ID card with more information. You may never need it, but knowing it is there will relieve some stress.

6. Research the local ER. With Google maps, you can easily search your hotel’s address and nearby hospitals, pharmacies and urgent care clinics. This may help you narrow down your hotel search. Then, if you do need to go to the doctor, you can get back to your vacation as soon as possible.

7. Use motion illness remedies and tools. Many medications and diseases can really amplify plane turbulence, the swaying of a cruise ship or even the motion sickness of riding in a cab. Your doctor can prescribe medication just in case. You can also rely on remedies like peppermints, a pressure-point bracelet and ginger ale.

8. Travel with someone whenever possible. Doing so will give you the peace of mind of knowing you have someone to lean on in case of flares or fatigue. Travel alone only when you feel you’re at your strongest.

9. Pack backup food. If you’re going somewhere exotic and aren’t sure if your body can handle the cuisine, you may want to pack some non-perishable food staples. Granola bars, formula drinks and even cereals or packaged soup mix are easy to pack in a suitcase. Even when traveling locally, bringing some of your own food is a good idea if you wind up without any energy to go out to eat.

10. Enjoy! Everyone deserves an escape once in a while. Even though traveling might be a little more complicated for you than for others, at the right time and with enough precautions, you’ll be on your way.

ILANA JACQUELINE is a 24-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.
We are a dog family. When our son Caleb started immune globulin therapy at age 3, our intuitive and exasperated nurse suggested we look into getting a dog because, “trying to place an IV in Caleb’s body can be compared to stuffing a giant octopus into a sleeping bag.” One call to our local Lab rescue, and enter George: a fur-infested, four-legged stomach looking for something or someone to embrace his heart and his incredible amount of slobber. After one night of Caleb recovering from a sinus infection, George seemed to know his calling: to take care of his primary immune deficiency disease kids Calvin, Caleb and Molly. George paved an easier path for the medically necessary and physically uncomfortable IV starts.

Eight years later, we adopted Jackson, a jet-black handsome fella with a gift for the goof. Jackson became Infusion Buddy. He was the pillow Caleb lay on while Nurse Nancy accessed his port. Jackson was right at Molly’s feet being a faithful friend and source of comfort when we were teaching her how to de-access her subcutaneous needles on her own. Even I got to bond with Jackson. A few years ago, I was diagnosed with an aggressive and degenerative form of arthritis. Three failed oral medications and one well-written prescription later, I found myself on the other end of infusion day. Jackson meant more to me than ever as patient instead of caregiver. A few months shy of Jackson turning 7 years old, our family made the gut-wrenching decision to free him from the bone cancer that took us all by surprise. That day, we got to take care of Jackson for the first, only and last time.

One Week Later
“I don’t think we should wait to get another dog,” college-bound Calvin said.
“This coming from the one leaving in two weeks’ time,” I reminded him.
“Yeah, we’ll be the ones cleaning up after it, feeding it and walking it, while you’re off having fun at togo parties,” Molly added with sisterly sass.
“It’s toga, not togo,” Caleb corrected her while drenching his taco in catsup.
“And Calvin won’t be going to any parties anyway. I’m not sending him off to college just to learn how to wear a sheet, but also how to wash one!” Mark joked. We were still so intensely raw from letting Jackson go, nothing tasted good, even on Taco Tuesday.
“Well, I agree with Calvin,” I added, while I dangled a piece of lettuce on my fork. “I know, for one, I have no desire to come home to an empty house after my infusion this coming week.”

We blame Sampson, the 14-week-old St. Berdoodle, on Calvin. He wanted to bond with the new family dog before he left for college, so we had to take the plunge. Sampson has quickly left his mark not just on our carpet but on our hearts. His floppy, moppy and shedless existence keeps us on our toes and our lives focused on the positive. Sampson has certainly eased the pain of losing Jackson, not to mention having to adjust to Calvin’s absence.

I must admit that while I was writing a reasonable check to Sampson’s breeder, many questions entered my mind. Would Sampson “dog-up” when it came to infusion days? Did he inherit that special instinct to bring comfort to my kids’ sick bodies, not shying away from the occasional migraine or latest vomit virus? Will Sampson avoid or embrace durable medical equipment and understand tubing isn’t tug-of-war approved and pumps aren’t for playtime? Will he really embrace the service dog he’s expected to be, or did I just adopt a Muppet?

By Cheryl L. Haggard
The Morning After Calvin
Goes to College

I had made a brunch date with a very close friend, Kori, who understood exactly how to adjust to a household with a human hole. Like clockwork, I was running late. About the third text I sent to Kori explaining where I was in my getting-ready routine, I tripped over Sampson’s enormous yeti paws. This was a call to play in his canine existence, and I didn’t have time to offer up a ball or a rope. So, I pushed a cardboard box in his direction, and he went to town on it.

Seconds later, Caleb and his best friend, Asher, who spent the night Sampson-sitting, emerged from slumber (or lack thereof).

“Hey guys, I gotta get going. I’m running late to meet Kori,” I hollered from within the depths of my room. I was attempting a two-handed mascara move when I noticed Sampson had, what we call in my family, the rips. Dodging the couch, diving under the coffee table and dashing through the office, Sampson was not to be denied. It was all fun and games until Sampson got hurt.

“Caleb, Asher! Come quick! He’s hurt!” I cried. Whimpering and shaking, Sampson tried to dislodge himself from a fishing pole that attached to him by way of a three-pronged lure!

The boys did all they could to get Sampson, and me, to calm down. With their combined 32 years of experience and Asher thankfully being raised on a farm, they took the situation by the scruff of the neck.

“We need to cut off the barbs; then we’ll just slip the lure out,” Asher instructed while Caleb held on to an impossible Sampson.

Despite the conundrum, Asher calmly asked me to “go have fun” and that he “had it all under control.”

After one marathon brunch, I came home to two 16-year-old boys trying to take a three-pronged lure out of our 40-pound puppy’s nares. Come to find out, Sampson got all excited when I left for my brunch with Kori, shook his head and lodged the lure deep into his sinuses. We rushed Sampson to our vet, where they had to perform emergency surgery to remove the lure.

While at the dinner table that night, Sampson’s fate came first course.

“So, you know how worried I’ve been that Sampson would fit in and understand his important role in our family,” I started. “Well, I’ve come to the conclusion that he is going to do just fine and will be the perfect Haggard helpmate.”

“What makes you believe all that?” Molly asked.

“Well, think about it! He’s endured a medical crisis that required Caleb’s assistance to keep him calm, similar to what Caleb’s nurses had to do to him when getting an IV.”

“Go on,” Caleb said with delight.

“And, he had to have sinus surgery, similar to what you guys have gone through!” Mark interjected.

“And, he’s on antibiotics so he doesn’t get an infection!” Molly finished.

“He’s a Haggard, that’s for sure!” Mark said with relief.

“He’s definitely a momma’s boy,” Caleb added.

I welled up with a sense of pride as our Sampson walked back into the room with my favorite pair of shoes dangling wistfully from his mouth. “Sniff!” I joked. “He’s gonna be my shoe boy!”

CHERYL L. HAGGARD is a stay-at-home mom and has three children with PI, two of whom have CVID.
Overcoming Medication/Treatment Refusal

By Jessica Leigh Johnson

Being a Parent to a child who suffers from a primary immunodeficiency (PI) comes with enough struggles without adding medication refusal to the list. Yet it’s an issue many parents deal with on a regular basis. Considering the number of medications our children take, or the frequency of their treatments, it isn’t surprising when these children grow weary of their health regimens. Some act out and rebel, refusing to take their medications or receive their infusions altogether.

After 11 years of administering subcutaneous immune globulin (SCIG) infusions, along with countless prescription medications, to three PI boys, I’ve dealt with my fair share of medication refusal. At age 6, my youngest son should be used to his SCIG infusions; he’s been getting them since he was 6 weeks old. Yet, week after week, when I pull out the tube of lidocaine cream and the Tegaderm patch, he runs away from me screaming: “No! Not again!”

On a recent infusion day, he said: “Mommy doesn’t like me. That’s why she gives me my infusions.” That couldn’t be further from the truth! It’s because I love my children that I do this painful thing to them week after week. It’s tempting to lose my temper and launch into an impassioned lecture: “I know firsthand what will happen if I don’t do this to you. I’ve already lost one child to PI, and I’m bound and determined not to lose another one. So get over here and let me stick you with this needle!” OK, maybe that’s not the best way to handle this already tense situation. So how does a parent get his or her kids to take their medicines or treatments without a nightly or weekly battle?

Get to the Root of the Problem

The first step in eliminating treatment refusal is to be sure there is no physical reason for the child’s aversion to his or her treatment. Yes, oral medications taste bad, and infusions can be painful. But there is a chance the child is experiencing unnecessary discomfort that makes his treatment even more undesirable than expected. A child may have innate reactions such as an allergy to an ingredient in a medication that causes bothersome symptoms. Certain medications have known side effects that could be behind a child’s aversion to treatment. For example, if an antibiotic causes stomach upset, a child will likely refuse it. Likewise, infusions can sometimes cause adverse reactions like headaches or pain and swelling at the needle insertion site. No child wants to take something they know will make them feel bad. So, it might be beneficial to ask the doctor if a different brand of medication would reduce these unwanted side effects.

Overcoming the obstacle of pain and discomfort will help treatments go more smoothly for your child.

In young children who cannot verbalize how they feel other than to kick and scream when they see the medicine dropper or needle coming toward them, getting to the root of the problem can be difficult. A parent knows his or her child best. If a child isn’t prone to tantrums, it might be wise to dig deeper to be sure there isn’t something going on other than stubborn refusal.

Tailor Your Approach to the Age of the Child

Medicine refusal takes on many forms depending upon the age of the child. While a baby may kick and scream before an infusion, an adolescent may flatly refuse or act out and rebel in other ways.

For babies and toddlers who can’t understand what is happening to them and why, physical force may be necessary to get the task done. While administering treatment or medicine, the child should be held in the parent’s arms.

This may make him feel more comfortable. Speak to the child in a calming voice while gently stroking his head or back until the medicine is given. If the child struggles, someone should gently hold his
arms and keep his head still.

For a school-age child, parents should calmly explain what they’re going to do. Any surprises will probably make things worse and cause more fear and reluctance the next time treatment is given. Some kids respond well when rewards are offered for good behavior. Unlike with babies, physically forcing an older child to take medicine should be done only as a last resort. If a child absolutely won’t take an important medicine, parents have no choice but to force the medicine down, and the child will almost certainly hate it. He may cry or feel betrayed. However, because the child’s health is at stake, it’s a necessary evil.

Educating a child about his condition when he’s young goes a long way in preventing non-compliance at an older age. Michelle Eakin, PhD, a health psychologist with the Johns Hopkins Cystic Fibrosis Center, recommends that parents and doctors educate young patients about the consequences of treatment refusal before they reach adolescence. Parents should reason with their school-age child, explaining why the medication is important and what will happen if he doesn’t take it.

Even though the treatment itself is non-negotiable, giving the child some of the responsibility such as putting the numbing cream on by himself will make him feel more involved in the process and less like it is simply being done to him. According to Dr. Eakin, “Children who know more about their medications and what they do, and who feel more confident in doing certain steps of the treatment, are going to be more likely to take their medications when they get older.”

When it comes to adolescents, medication compliance often becomes one more source of conflict between parents and teens. What can be done to prevent refusal? “One thing is certain,” Dr. Eakin says, “nagging doesn’t help. [And,] yelling and screaming are not going to work.” Parents who have a cooperative working relationship with their teens generally have more success getting them to take their medications than those who are more rigid and force the issue.

A written contract between parents of chronically ill teens can improve medication compliance. In the contract, the teen will agree to take the required medicine or treatment, or they’ll lose privileges such as use of a cell phone or the family car on Friday night. The parents, on the other hand, must promise not to nag their child about the treatment.

Helping Liquid Medicine Go Down

Because of frequent infections, PI kids have to take oral medicines such as antibiotics more often than most children. While there are a few liquid medications that taste like candy, most of them taste like chalk. Getting kids to take these medications two or three times a day for a week or more can be a struggle, but there are a few ways to make the medicine go down more easily.

The best way to combat the taste issue is to stop using liquid medication or chewables. The older a child gets, the bigger his dose will be, and if it tastes bad, this can cause a daily struggle. As soon as a child is able to swallow pills (as early as 4 years old), switch from liquid medicine and chewables to tablets or capsules. Have the child practice swallowing mini M&Ms to get used to the sensation of swallowing without chewing. (Never break, crush or dissolve a capsule or tablet without talking to your pharmacist. New medications are often in a delayed-release form, and they can become unstable if the structure of the pill is changed.)

If pill swallowing isn’t going well and liquid medication is the only option, many pharmacies will add kid-friendly flavors to the bottle. For a child over the age of 1, a spoonful of Hershey’s syrup on a spoon can be used as a chaser following bad-tasting medicine. The chocolate syrup is thick enough to coat the mouth and hide the bitterness of certain medicines.

Giving liquid medicine in small bursts is also helpful. While it may seem that it’s best to get things over with quickly, shoving a syringe full of medicine into a child’s mouth all at once can make him gag, which sometimes leads to vomiting. Be sure to aim the medicine at the cheek instead of the back of the throat to prevent choking.

Do the Best You Can

Although it can be disheartening when your best efforts to keep your children healthy are met with rejection, remember that you are doing your best as a parent. Keep in mind that just as a spoonful of sugar (or chocolate syrup) helps the medicine go down, a few loving words don’t hurt, either. “Come over here, little boy, and let me squirt this saline solution up your nostrils. Oh, and remember, Mommy loves you.”

Jessica Leigh Johnson is a stay-at-home mom and mother of four kids, three of whom have X-linked agammaglobulinemia. She is a member of American Christian Fiction Writers and has written one book about the loss of her son to a primary immunodeficiency.

References
LIVING WITH PRIMARY immunodeficiency (PI) is often described as a “journey,” perhaps because the path one travels following diagnosis is full of unknowns, detours and even U-turns. Symptoms flare and recede, and many patients experience a roller-coaster ride of good versus bad days in terms of energy, stamina and mood. Getting comfortable — physically and emotionally — is a challenge that is universal for patients with PI and all types of chronic illness. That’s why stocking your home with products that provide relief and relaxation can help make your journey a bit less stressful.

Infusion-Time Comfort

Immune globulin (IG) therapy can take anywhere from four to six hours depending on the patient, dosage and type of infusion. Finding creative ways to pass the time is no small feat, but healthcare providers agree that finding “positive distractions” to ward off boredom can help patients maintain a positive outlook. Sometimes, a positive mental shift can occur simply by referring to the scheduled infusion as “me time.” Consider using it as an opportunity to focus on the things you enjoy while unplugging from life’s daily demands. Suggestions include:

• Stockpile articles, books and magazines you’ve been meaning to read, and keep them in a bin to peruse during infusions.
• Start a journal and use it to catalog your feelings, pen some poetry or simply reflect on life’s events.
• Create a movie marathon by streaming a popular series or film trilogy. Consider tried-and-true comedies like “I Love Lucy” to boost your mood.
• Learn something new by streaming educational or motivational podcasts. Content is readily available from popular speakers like John Maxwell, Jack Canfield and more.
• Try an audiobook; you can listen to short stories or finish an entire novel. Check out services by Audible, Simply Audiobooks and iTunes.

Side Effect Relief

When it comes to physical comfort during an infusion, plush pillows, cozy blankets and sound-blocking headphones are mainstays for the chronically ill. Of course, for PI patients, physical discomfort is not limited to the time spent waiting for an infusion to end. Although IG therapy is a lifesaver for those living with PI, it can also present with uncomfortable physical side effects. Patients report battling headaches that run the gamut from mild to migraine strength, body aches, flu-like symptoms and chronic respiratory infections. While having over-the-counter treatments like Tylenol and Benadryl on hand is a good idea, stocking your home with symptom-relieving devices like humidifiers, cooling eye masks and body-contour pillows can also provide immediate relief from some of the mild but bothersome symptoms associated with IG therapy.

Fighting Fatigue

Exhaustion and fatigue are perhaps the most common complaints among patients with chronic illness. In her blog “The Spoon Theory,” patient advocate Christine Miserandino explains what that constant energy drain feels like. “The hardest thing I ever had to learn is to slow down and not do everything. I need to think about the weather, my temperature that day, and the whole day’s plans before I can attack any given thing,” she says. “When other people can simply do things, I have to make a plan like I am strategizing a war.”

As Christine states on her popular website, But You Don’t Look Sick, learning to live with the challenges associated with PI is not easy, but it is possible. With so many aspects of chronic illness simply out of the realm of individual control, seeking comfort in even small ways can help ease some of the physical and emotional stress patients live with daily. Using infusion time to pursue enjoyable activities and investing in comfort-producing products is a small step toward personal empowerment that can make the chronic illness journey that much easier to navigate.

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.
**Sweet Dreams**

How does a massage to the nervous system sound? The Dreampad sleep and relaxation aid incorporates music that is transmitted to your body as a vibration. The vibrations are said to stimulate the parasympathetic nervous system inducing relaxation and sleep. Dreampad can be used with your own MP3 player, or you can purchase a preloaded Sony device with your order. From $159; [dreampadsleep.com](http://dreampadsleep.com)

**Go Fish**

Soothe aches and pains with these fun and functional microwavable corn bags. Fishbellies Pain Pads can be used as a heating pad or freezer cool pack. They are made of 100 percent cotton, filled with the largest surface area corn kernels to hold heat for hours and are available in an array of colors, sizes and designs. $30 and up; [fishbellies.com](http://fishbellies.com)

**Cooling Comfort**

The IMAK Pain Relief Mask conforms to your face and eyes, blocking out light. Breathable cotton material is comfortable to wear, and the smooth ergo beads inside create a gentle massaging effect around your eyes. For added stress relief, place Pain Relief Mask in the freezer to provide cool relief without the shock of ice or gel packs. $12.99; [amazon.com](http://amazon.com)

**Migraines Be Gone!**

Developed by a neurologist and lifelong migraine sufferer, Ausanil claims to work rapidly to relieve the pain of severe tension headaches, migraines, cluster headache, rebound headaches and medication overuse headaches. This popular homeopathic spray is made with chili pepper extract and promises fast relief. .27 fluid ounces $35; [ausanil.com](http://ausanil.com)

**Cheaper than Therapy**

This unique product’s description kind of sums it up: Whenever things don’t go so well, and you want to hit the wall and yell, here’s a little Dammit doll that you can’t do without. Just grasp it firmly by the legs and find a place to slam it. And as you whack the stuffing out, yell “Dammit! Dammit! Dammit!” From $12.99; [amazon.com](http://amazon.com) and various retailers
**BOOK CORNER**

**Slow Medicine**
Author: Michael Finkelstein, MD  
Publisher: William Morrow Paperbacks

Those who are chronically sick, tired or depressed need a medical examination that includes, but goes beyond, the exact location of symptoms, according to Dr. Finkelstein. He has helped tens of thousands of patients achieve extraordinary health with his slow medicine prescription of skillful living. In the book, he guides readers through the essential questions for understanding various symptoms and their causes. Each chapter includes the key components of a successful consultation — from revealing lessons to practical prescriptions — along with illustrative anecdotes from real patients. Written to take readers beyond conventional medicine to examine the intricate network of factors that lie behind many common illnesses, Dr. Finkelstein empowers individuals to take their health back and walk down the slow medicine path — one where the answers are in the questions.

**The Marvelous Transformation: Living Well with Autoimmune Disease**
Author: Emily A. Filmore  
Publisher: Footprint Books

More than 80 health conditions are caused by autoimmune disease, with symptoms ranging from occasionally uncomfortable to debilitating or life-threatening. Emily Filmore, a fellow sufferer, writes about her struggle with autoimmune disease and offers tips on how to thrive while living with physical limitations. She uses humor and spirituality to provide practical coping mechanisms to ease physical, mental and emotional discomfort.

**Live Well with Chronic Pain: A Journey of Discovery**
Author: Liza Leal, MD  
Publisher: iUniversity Inc.

Written by a medical expert with firsthand knowledge as a chronic pain thriver, Dr. Leal describes how the “Four Foundations for Living Well with Chronic Pain” can provide a powerful motivational tool to help those living with chronic pain more effectively manage their pain, health and future. Dr. Leal’s personal experience with rheumatoid arthritis and chronic pain gives her a unique perspective both in her practice and for her patients’ needs. The book demonstrates and outlines effective ways to manage chronic pain while living life to the fullest.

**How to Live Well with Chronic Pain and Illness: A Mindful Guide**
Author: Toni Bernhard  
Publisher: Wisdom Publications

The book is written to provide comfort, understanding and advice for those who are suffering and those who care for them. Author Toni Bernhard addresses these challenges and more, using practical examples to illustrate how mindfulness, equanimity and compassion can help readers make peace with a life turned upside down. She shows how to cope and make the most of life despite the challenges of chronic illness. Specifically, she describes how readers can benefit from mindfulness exercises to mitigate physical and emotional pain; concrete advice for negotiating the everyday hurdles of medical appointments, household chores and social obligations; and tools for navigating the strains illness can place on relationships. Several chapters are directed toward family and friends of the chronically ill, helping them to understand what their loved one is going through and how they can help.
For a more comprehensive list of resources, visit the Resources page at IGLiving.com.

**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

**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_306777_Article.jsp#1112boefW6E0
  - Kawasaki Disease Foundation: www.kdfoundation.org
  - KidsHealth: kidshealth.org/parent/medical/heart/kawasaki.html

**Mitochondrial Disease**

- WEBSITES
  - United Mitochondrial Disease Foundation: www.umdf.org
  - MitoAction: www.mitaction.org

**Multifocal Motor Neuropathy (MMN)**

- WEBSITES
  - The Neuropathy Association: www.neuropathy.org

**Multiple Sclerosis (MS)**

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

**Peripheral Neuropathy (PN)**

- WEBSITES
  - Neuropathy Action Foundation: www.neuropathyafrica.org
  - Western Neuropathy Association: www.pnhelp.org
  - Texas Chapter of the Neuropathy Association: www.handsfeetheart.org

**Primary Immune Deficiency Disease (PI)**

- WEBSITES
  - Immune Deficiency Foundation: www.primaryimmune.org
  - Jeffrey Modell Foundation: www.info4pi.org
  - American Academy of Allergy, Asthma & Immunology: www.aaaai.org
  - International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: www.ipopi.org
  - New England Primary Immunodeficiency Network: www.nepin.org
  - Rainbow Allergy-Immunology: www.uhospitals.org/rainbow/services/allergy-immunology

**Scleroderma**

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

**Stiff Person Syndrome (SPS)**

- WEBSITES
  - American Autoimmune Related Diseases Association Inc.: www.aarda.org
  - Genetic Alliance: www.geneticalliance.org
  - Living with Stiff Person Syndrome (personal account): www.livingwithspss.com
  - Stiff Person Syndrome: www.stiffpersonsand.org
my Ig source

ANSWERS AND ADVOCATES

Nancy
Travel enthusiast
living with PI
Support for living hands-on

Life is full of beginnings, and a primary immunodeficiency (PI) diagnosis is one of them. MyIgSource is here to support you throughout your PI journey.

Reach out for support today.

Call 1-855-250-5111 or visit MyIgSource.com to enroll in the program.

This program is available to all patients and caregivers regardless of treatment.

Insurance & Financial Support

Get help understanding health insurance and navigating your coverage, and find out if you qualify for copay assistance.

Nurse Advocates

Experienced nurses are available to answer your Baxalta Ig product and administration questions.

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Patient Advocates

Connect with other patients and caregivers living with PI. Your Patient Advocate will also be your guide to all things MyIgSource.

Educational Resources & Tools

Access educational books, magazines, emails, and more about managing your PI journey.
Take Control of your flu vaccine supply
with MyFluVaccine.com easy online ordering

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YOU PICK THE QUANTITY » Choose from a broad portfolio of products
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