Inheriting PI
Understanding the Genetics

Strategies for Managing Your Healthcare ‘Team’

Negotiating Healthcare: Reduce Costs Before and After

Effects of Long-Term Corticosteroid Use

Understanding Myasthenia Gravis
Get Connected
Your Complete Resource for Advocacy, Education and Support

On our new IGLiving.com

Features an easy-to-navigate design

Updated and enhanced content

New Patient Advocate, Abbie Cornett

Blogs

Valuable Resources and more
with IG Living!

On Facebook
Find timely and relevant information posted daily and participate in one of our new Chats hosted by medical professionals.

On the Go
The new IG Living App allows you to connect Anytime, Anywhere! And it is FREE!
**Life’s Gambles**

I’m not a “gambler” in the true sense of the word, but in life, we are all forced to bet the odds. Take “Tom,” for instance, who has a rare condition called X-linked lymphoproliferative syndrome (XLP), an often-fatal disease that compromises the immune system. Although he carries the inherited mutant gene, making it likely he would pass the disease to offspring, he has three healthy children. Or, Jessica, a carrier of the gene that causes X-linked agammaglobulinemia (XLA), an immune disorder that typically occurs in one of every four births when the mother is a carrier. After her first two sons were born with XLA, she thought her family’s XLA quota was fulfilled. Instead, four of her five children inherited XLA.

As we discuss in our article “An Undesirable Inheritance,” the decision to have children when one or both parents have the potential to pass along a defective gene to their offspring is indeed a gamble. But, understanding how diseases like primary immunodeficiency (PI) are inherited can help parents better understand the risk factors and how to deal with the guilt that often accompanies that decision. For PIs, genetic counseling can help alleviate fear and shed light on treatment options and quality-of-life issues, allowing couples to fulfill their lifelong dream of having a family, despite the risks.

Unfortunately, as many chronically ill patients learn, treatment comes with its own set of risks. Some patients suffer years of illness before finding a physician who can diagnose them. Once they are diagnosed, often with multiple conditions, managing their own health while dealing with multiple specialists can be a challenge at best. But patients can help improve the odds of good outcomes by choosing and managing their medical team as outlined in our article “Managing Multiple Healthcare Specialists.” Strategies include looking at different sites of care, negotiating boundaries, keeping track of symptoms, medications and tests, and maintaining up-to-date medical records.

Similar strategies can be used to help increase the chances of reducing the cost of care. As our article “Negotiating Healthcare Costs” explains, patients can’t assume that treatment will always be covered by co-pays and deductibles listed on their policy, or that fees are set in stone. They must ensure that procedures are pre-approved and that referrals are given to in-network specialists. And, what patients often don’t realize is that some physicians will discount their rates for services in certain situations, and many may offer payment plans. In addition, there are resources that patients can turn to for payment assistance.

So many things in life are a gamble. In most cases, there is a 50-50 chance of having things turn out for the better or worse. But, as immunologist Terry Harville, MD, PhD, explains, what individuals need to factor in is the statistical phenomenon of “regression to the mean”: “It is the final moments of the basketball game, and the two best shooters on the team hit 50 percent of the baskets; one has made his last five shots, and the other has missed his last four shots. Do you give the ball to the one with the ‘hot hand’ (the last five made) or to the one with the ‘cold hand’ (the last four missed)? Initially, the odds would appear to be equal for either. But regression to the mean suggests that the person making the last five will have to miss five to get back to the mean, whereas the person missing the last four will have to make four to get back to the mean.” This phenomenon can apply to all things in life. “Due to the random nature of events, there appear to be ‘streaks’ and ‘slumps,’” adds Dr. Harville. “But these are there to allow for the mean to be the average that it is.” Patients, then, can find solace in the fact that when the odds seem against them, they will eventually rebound for the better.

Ronale Tucker Rhodes, MS
**Why Should I Worry?**

By Abbie Cornett

"WHY SHOULD I worry?" is a question I am constantly asking myself. I worry about things that 20 years ago I wouldn’t have known existed because, today, there is a constant bombardment of information in our lives.

Of course, advances in technology and communication are a good thing; they have made the world a much smaller place, and they have done much to bring people together. Today, it is easier to see the world from a global perspective.

Yet, while this unprecedented access to information has brought us closer together, it also has consequences. The constant stream of information about everything from the climate and economic projections to possible pandemics means we must constantly filter information. We are forced to continuously determine how current issues will impact us as individuals. But because of the nature of mass media, we are frequently given only sound bites of information — just enough to cause us stress, and not enough to allow us to make an educated decision about how this will impact us.

While all people are susceptible to anxiety caused by partial or incomplete information, people with chronic illness are particularly susceptible. Our very lives may depend on what’s trending in the news.

As patient advocate for IG Living, I see this stress every day on a firsthand basis. Patients contact me because they have questions about what they have read or seen in the media. Examples of this are how outbreaks of diseases such as the Zika virus may affect the safety and availability of a plasma product like immune globulin (IG), or how changes in current laws will affect insurance coverage. Although these types of issues affect all people, they are frequently of greater concern to the chronically ill and their families.

When I receive questions from patients about trending issues, I look for the most current information available from reliable resources to find answers. If a question is about an issue that can affect all patients, such as guidelines for plasma collection from the Plasma Protein Therapeutics Association (PPTA), I make that information available through either our website at www.igliving.com or our Facebook page at www.facebook.com/IGLivingMagazine.

For instance, I recently posted a link to new guidelines from PPTA concerning the collection of plasma from people who have traveled to areas affected by the Zika virus and the risk the virus poses to the plasma supply. Through this post, patients were able to learn about the Zika virus and how it is spread, primarily through Aedes mosquitoes, and more rarely through sexual contact or transfusion. They were also able to learn why PPTA does not recommend deferral of donors because of Zika’s “relatively large size and lipid envelope [making] it highly susceptible to virus inactivation and removal through the current manufacturing process.” In other words, patients should be reassured that the plasma supply is safe from Zika!

This type of accurate information about issues that affect patients is invaluable to relieving stress. My job is to ensure patients and their families have the information they need to determine what may actually affect their health or their access to lifesaving medications by sorting the facts from the fiction. This allows people to make educated decisions regarding their care.

If you are interested in learning more about issues that may affect you or your family, I recommend you look at our website for news and upcoming teleconference schedules and our Facebook page for educational posts.

**ABBIE CORNETT** is the patient advocate for IG Living magazine. She can be reached at patient advocate@igliving.com or (800) 843-7477 x1366.
**ASK THE EXPERTS**

**Question** » Is a rash a common side effect of IVIG?

I have been prescribed three different brands of intravenous immune globulin (IVIG). The loading dose was Gammaked. I was then switched to Gamunex-C, and now I am treated with Privigen. All of these have given me an ugly rash. My doctor sent me to a dermatologist who conducted a biopsy that showed the rash was related to the IVIG. How common is this?

**Abbie** » Patients who have a normal IgG antibody count, but who are low in one or more IgG specific subclasses (IgG1, IgG2, IgG3, IgG4), may have what is called a selective antibody deficiency (SAD). This is determined by testing for a response to either the Prevnar 13 or Pneumovax 23 vaccines. Prevnar 13 tests for conjugate protein antigen response, whereas Pneumovax 23 tests for response to polysaccharide antigens. Many physicians commonly use Pneumovax as part of the workup, but each physician has a different methodology. Either way, the results suggest a diagnosis of SAD.

**Question** » Why would a person’s antibody levels test normal when the individual serotype titers test low?

I have had recurrent chest infections that have been treated with antibiotics. After testing, my total IgG antibody count was normal, but my individual serotype levels were very low for 10 of the 13 components of Prevnar 13, the pneumococcal 13-valent conjugate vaccine. Why would my total antibody count be “normal,” but my individual serotype titers be so low? And, why did my immunologist test against the Prevnar 13 vaccine rather than Pneumovax 23, the 23-valent pneumococcal polysaccharide vaccine?

**Abbie** » A rash is a very common side effect of IVIG, and some patients develop worse rashes than others. It’s possible that switching to another brand of IVIG could help, but since you have already tried three different brands, you may experience the same issue. Taking premedications such as steroids or Benadryl before treatments can reduce or eliminate some of the side effects. If the rashes persist, you may want to discuss switching from IVIG to subcutaneous IG (SCIG). However, your ability to switch to SCIG will depend upon your prescribed dosage of the medication.

**Question** » How can severe side effects from high-dose IVIG treatments be reduced?

I have dermatomyositis, and I am being treated with high-dose intravenous immune globulin (IVIG) once per month. I am prescribed 55 grams of Octagam over two days at a fairly slow infusion rate. I know IVIG is working to treat the disease because my rash is so much better, and I regain my muscle endurance in the second part of the four-week cycle. However, despite slowing the infusion and taking medications before, during and after, I still get a terrible migraine. Even more incapacitating is the body bloating, nausea and upper-gastrointestinal upsets that occur for at least 10 days after. I’ve never gained weight easily, and now I find I’m 5 to 10 pounds heavier. Some of it is bloating, but most of it sticks around. And, I’m eating much less because of the nausea and discomfort. Even my legs feel swollen for a week. Is there an explanation?

**Abbie** » I spoke with one of our experts who said it appears you are being treated correctly. As part of the premedications, we suggest asking your physician for one for nausea. While you state that your infusions are fairly slow, infusing 55 grams should take about 5-1/2 hours. However, you could try slowing the infusion rate even more or breaking it up over three days.

**Have a question?** Email us at editor@IGLiving.com. Or, submit your question on our Ask the Experts page on the IG Living website at www.igliving.com/life-with-ig/ask-the-experts.html. Your information will remain confidential unless permission is given.

**ABBIE CORNETT** is the patient advocate for IG Living magazine.
FACES OF IG

Join the conversation! Connect with other immune globulin patients through IG Living’s Facebook page at www.facebook.com/IGLivingMagazine. Each day, we post interesting articles and facts, as well as thought-provoking questions that you can weigh in on. These are some snapshots of what’s being discussed.

Plasma donations save lives!
Yes, thank you! Your gift touches many lives. We have moms who can be mommies. Dads who can be daddies. Children alive to give hope and beauty. You touch so many lives by your gift. Without you, many would be so very ill and cease to be. So may I touch lives through the life you help me sustain! Don’t stop. You are so appreciated by us all!
— J S-DiNottia

Thank you to all who donate! Your generosity allows me to receive the medicine I need every three weeks!
— T L Forbes

Thank you, and I hope you do realize just how many lives you save and sustain every time you give blood. If not for your plasma donations, my IV treatments would cease, and I would not be able to function or go to work every day.
— K J Pace

What you don’t want to hear!
The other day someone asked me if I am cured. I really do get tired of explaining it over and over again. But, in all fairness, how do we expect a nonmedical person to understand when our doctors often have trouble figuring it out?
— Debbie K

I have had cancer ... and due to my illness, my chances of getting cancer again are great, because my immune system isn’t working.... I don’t have the time to explain all this to someone. Nor do I sit and worry when it will raise its ugly head again. I am happy to live in the moment.
— J S-DiNottia

My favorite is: “Can’t you take vitamins?”
— C Moore

Why do we say that we are OK?
Because it gets old after a while. When you have a chronic illness that will not go away, you take your better days and make the best out of them. Then, some people think you are OK. For the most part, at that “moment,” I am OK. But that can change in an instant.
— J Sonkin

Because those who “used” to be our friends don’t really want to know. If they did, they would stay around long enough to listen. I have a few friends from church — I mean really few — who are true friends who stay to listen. But the rest don’t want to be bothered with me any longer.
— S B Rhodes

Because my so-called “friends” don’t want to be bothered. I have one male friend who’s supposed to be my best friend in the world, and when I say I’m not feeling well, he lies and says his messenger is down, doesn’t talk to me for days and then ignores my medical condition completely. I’ve unfortunately learned to live in isolation that has affected me in so many negative ways.
— K Peterson

By Terry O. Harville, MD, PhD

IN PREVIOUS issues, we have been discussing how disruption of the timing of developmental events during the first trimester results in the features of DiGeorge syndrome (DGS). The specific facial characteristics are typically noticeable, the hypocalcemia (low serum calcium levels in the blood due to malformation of the parathyroid glands) may have clinical and laboratory detection, and the cardiac malformations will have clinically detectable features; however, the immunodeficiency due to malformation of the thymus may not be evident early on. Further, other clinically relevant issues may not be readily apparent but can have significant impact on the child’s overall well-being.

As the human embryo begins to grow and develop, a remarkably ingenious process occurs to help ensure that the steps that result in the correct developmental stages happen in the proper order. Very early, specialized cells known as “neural crest cells” leave the area that will become the backbone and migrate to specific predetermined areas throughout the embryo to help induce the cells in those areas to become specific tissues, and in turn, specific organs. For instance, in DGS, failure of the correct neural crest cells migrating to the chest to direct the formation of the heart results in cardiac malformations. As the neural crest cells migrate, they leave a trail that helps the developing embryo know where to lay out nerves and blood vessels. Further, at the sites of specific organ development, the neural crest cells form a “scaffold” to direct the cells and tissues in the vicinity to where they need to go to build the organ. Thus, neural crest cells are analogous to trailblazers, surveyors, architects, contractors and master craftsmen, preparing and inducing the correct formations of an embryo’s tissues and organs in the appropriate sequence.

Most creatures on Earth develop in bilateral symmetry, meaning they have a left half and a right half that more or less mirror each other. Humans have two eyes, two lungs, two kidneys, etc., and even organs like the brain and the thymus have a left and right half. Bilateral symmetry, however, is lost in the center of the body. Humans have one heart and one gastrointestinal system. In the region of the embryo that will develop into the head, neural crest cells help to direct the folding and growth of the tissues to separate the nose from the mouth, and the correct formation of tissues leading from the mouth into the esophagus and the nose into the trachea. When the correct sequence is not followed, cleft lip and/or cleft palate may occur. Further, while it may not be fully identified on visual inspection, the clefting effect can also be present in the esophagus and trachea because when the two halves moved together, they may not have sealed up as they were supposed to. Instead, gaps may persist, resulting in dysfunction of the esophagus and trachea, and incorrect swallowing as a consequence.

This latter developmental issue is a greatly underrecognized issue in patients with DGS, known as gastroesophageal reflux (GER). All patients with DGS are at risk for GER, and may have reflux aspiration, meaning stomach contents may come up the esophagus and go into the lungs, or there may be swallowing difficulties that result in dietary material being swallowed directly into the lungs. All patients with DGS need reflux precautions, should receive GER medications and may require surgery to help ameliorate the problem.

We will continue next issue describing further the poorly recognized features of DGS, which can contribute to unwellness.

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.
IN THE NEWS

Education

Webinar Series Launched for Young Adults Living with Primary Immunodeficiencies

PI Voices is a new young adult webinar series developed by the Immune Deficiency Foundation (IDF) to address the unique needs of young adults living with primary immunodeficiency disease (PI). The series consists of five live webinars covering topics from career management, medical care prioritization, insurance transitions, family planning and health/social life balance. Each offers participants an opportunity to ask questions of panel experts and connect with other audience members who may share their experience of living with PI. While the first three webinars took place in March, April and May, the last two in the series are:

• August 24: Family Planning and PI
• September 21: Balancing Your Health and Your Social Life

The series is sponsored by CSL Behring and requires registration. Recordings of each webinar are available after the live events.

“We recognize that young adulthood is a critical time for people with primary immunodeficiency. For the first time, many are taking responsibility of their own care, impacting all aspects of their life,” says Marcia Boyle, IDF president and founder. “We are so grateful for CSL Behring’s support of our new, interactive program, giving this age group a valuable opportunity to learn how to manage these concerns.”

For more information, visit primaryimmune.org/young-adults/webinars.

Insurance

Many 2016 PPO Plans Remove Out-of-Network Cost Limits

This year, 45 percent of silver-level preferred provider plans (PPOs) offered under the federal health law have no annual cap for policyholders’ out-of-network costs, according to an analysis by the Robert Wood Johnson Foundation. This compares with 14 percent of silver-level PPO plans in 2015. Silver plans, which are the second-lowest cost plans, are the most commonly purchased in the marketplace. With no annual cap, and because out-of-network costs are often harder to find on insurance websites, consumers could be caught unaware and end up with bills that total in the tens of thousands of dollars for hospitalizations or treatment by providers who are not part of the plan’s network.

Out-of-network costs were a concern for consumers prior to the Affordable Care Act, which doesn’t set any limits on what insurers or medical providers can charge policyholders for out-of-network care. And, while some states have considered or put in place consumer protections, they are limited. However, the law does cap what insurers can charge policyholders annually for in-network care. For 2016, plans must limit out-of-pocket costs for in-network services, which includes deductibles and co-payments, to $6,850 for individual coverage or $13,700 for family plans. And, insurers must charge in-network co-payments for emergency department services at non-network hospitals because patients in urgent situations often can’t choose where they go.

The National Home Infusion Association (NHIA) has released a final set of revised Standard Definitions for Patient Outcomes Data that are intended to be used in collecting data related to patient events for assessing the safety, effectiveness and efficiency of home and specialty infusion care. Developed by a volunteer task force, the definitions will allow providers to engage in industrywide benchmarking and research activities, generating the necessary data for demonstrating the quality and value associated with administering infused medications in the home setting.

The revised definitions replace an initial version finalized in 2012. The new definitions include adverse drug reaction, emergency department use, unplanned hospitalization, access device events, medication error and discharge reasons. “In the first version, not every outcome data element had reasons, interventions or outcomes associated with it,” said Connie Sullivan, vice president of research for NHIA. “In this version, every outcome element has those items associated with it. It’s a standardized set of data measures to help us identify the impact of that event on the patient’s home infusion care.”

The definitions are part of NHIA’s ongoing Industry-Wide Data Initiative created to establish industry demographic, operational and clinical quality benchmarks. Next, NHIA will undertake development of national quality measures for home infusion. To see the final Standard Definitions for Patient Outcomes Data, go to www.nhia.org/Data/Data-Definitions.cfm.

Green Cross Corp., a South Korean biopharmaceutical company, has submitted a biologics license application (BLA) for IVIG-SN (human normal immune globulin G for intravenous administration) to the U.S. Food and Drug Administration (FDA) for the treatment of primary immunodeficiency diseases (PIDs). The application is based on positive results of a Phase III study in patients with PI, which were well under the requirement specified by FDA for no more than one acute serious bacterial infection per patient-year. Green Cross is the largest plasma protein products manufacturer in Asia; it has been marketing IVIG-SN in more than 30 countries in Asia, South America and the Middle East.

Bio Products Laboratory has launched a clinical trial evaluating the safety and pharmacokinetics of Subgam-VF 16% liquid immune globulin, made from human plasma specifically for subcutaneous use for the treatment of primary immunodeficiency diseases (PIs). The Phase III multicenter, open-label study, which enrolled its first patient in September, will take place in the U.S. and will enroll 35 patients with PI who will be treated for up to 26 weeks. In the trial, the pharmacokinetics of Subgam-VF will be assessed and compared with previous studies. “Enrolling the first subject is an important step in our ongoing effort to bring high-quality products into the U.S. market,” said Ken McNish, PhD, president of BPL U.S. “Patients with primary immunodeficiency need options for immunoglobulin replacement therapy, and we’re excited to investigate an additional option.”
Research

European Scientists Develop Possible Cure for ADA-SCID

A group of scientists and doctors in Milan have developed a potential cure for children born with adenosine deaminase deficiency-severe combined immune deficiency (ADA-SCID), a rare condition in which a single genetic defect prevents them from developing a robust immune system, leaving them susceptible to infections. Without treatment, children with ADA-SCID rarely live more than two years.

Known as Strimvelis, the procedure involves inserting a new gene into the patient’s stem cells. Over the past 14 years, 22 children have been treated with Strimvelis, and all are still alive, most without needing any further treatment. In 2010, GlaxoSmithKline obtained the rights to market the therapy, and on April 1, it received a positive opinion from the European Medicines Agency’s advisory committee, paving the way for final approval.

Currently, the best available option for treating ADA-SCID is a bone marrow transplant. However, that success depends upon how well-matched the donor is. For the one in four babies who have a well-matched family member, a transplant can be a cure. But, for the rest, success rates can be as low as 50 percent.

If Strimvelis is approved, it will be only the second gene therapy to be sold in Europe. No gene therapies are approved for sale in the U.S.

FDA Issues Recommendations on Zika Virus and Blood Donations

In February, the U.S. Food and Drug Administration issued a new guidance recommending the deferral of individuals from donating blood if they have been to areas with active Zika virus transmission, potentially have been exposed to the virus or have had a confirmed Zika virus infection. Specifically, in areas with active Zika virus transmission, FDA recommends that whole blood and blood components for transfusion be obtained from areas of the U.S. without active transmission. However, blood establishments may continue collecting and preparing platelets and plasma if an FDA-approved pathogen-reduction device is used. In addition, the guidance recommends blood establishments update donor education materials with information about Zika virus signs and symptoms and ask potentially affected donors to refrain from giving blood. In areas without active Zika virus transmission, FDA recommends that donors at risk for Zika virus infection be deferred for four weeks. Individuals considered to be at risk include those who have had symptoms suggestive of Zika virus infection during the past four weeks, those who have had sexual contact with a person who has traveled to or resided in an area with active Zika virus transmission during the prior three months, and those who have traveled to areas with active transmission of Zika virus during the past four weeks.

While there have been no reports to date of Zika virus entering the U.S. blood supply, the risk of blood transmission is considered likely based on the most current scientific evidence of how Zika virus and similar viruses (flaviviruses) are spread and recent reports of transfusion-associated infection outside of the U.S. It is also a concern because four out of five individuals infected with Zika virus do not become symptomatic. “Based on the best available evidence, we believe the new recommendations will help reduce the risk of collecting blood and blood components from donors who may be infected with the Zika virus,” said Peter Marks, MD, PhD, director of the FDA’s Center for Biologics Evaluation and Research.
Autoimmune Corner

Research

Autoimmune Disease May Be Treated Without Compromising Immune System

A new study at The Scripps Research Institute (TSRI) shows how diseases such as lupus, ulcerative colitis and multiple sclerosis (MS) might be treated without causing dangerous autoimmune responses that compromise the immune system’s ability to fight viruses and bacteria. While working on clinical trials of ozanimod, an autoimmune drug candidate under development by Cangene, the researchers found they could administer low doses of the drug and still get a treatment response. This led them to further test the drug’s effects on the immune system. “With this tool, we show a new, important mechanism in disease that can prevent collateral tissue damage while preserving the protective host response,” said TSRI professor Hugh Rosen, who led the study. Ozanimod is not yet approved by the U.S. Food and Drug Administration. The study was published in January in the Early Edition of the Proceedings of the National Academy of Sciences.


Research

FDA Expands Promacta Approval for ITP

The U.S. Food and Drug Administration (FDA) has approved an expanded use for Novartis’ Promacta (eltrombopag) to include children 1 year of age and older with chronic immune thrombocytopenia (ITP) who have had an insufficient response to corticosteroids, immune globulin or splenectomy. The updated label also includes a new oral suspension formulation of Promacta that is designed for younger children who may not be able to swallow tablets. Promacta was approved by FDA as a tablet formulation in June 2015 for children 6 years of age and older and in 2008 for adult patients with the same condition.

The label expansion is based on data from two double-blind, placebo-controlled trials, including the largest Phase III trial in this patient population. The trials showed that treatment with Promacta significantly increased and sustained platelet counts among pediatric patients with chronic ITP with an insufficient response to prior chronic ITP therapies, and some patients taking concomitant ITP medications were able to reduce or discontinue their use of these medications, primarily corticosteroids.

Promacta is intended for use only in those whose degree of ITP and critical condition increase the risk for bleeding. It is a once-daily oral thrombopoietin-receptor agonist that works by inducing stimulation and differentiation of megakaryocytes from bone marrow stem cells to increase platelet production.


Research

Study Shows HSCT Tied to Long-Term MG Remission

A retrospective study has found that treatment with autologous hematopoietic stem cell transplant (HSCT) can result in durable, symptom-free, treatment-free remission of myasthenia gravis (MG). In the study, seven patients with severe cases of MG (five of whom had immune dysregulation-related concurrent autoimmune or lymphoproliferative illnesses) treated with autologous HSCT were followed for a median of 40 months. All patients achieved durable complete stable remission, according to the Myasthenia Gravis Foundation of America clinical classification, with freedom from ongoing therapy and no residual symptoms. After autologous HSTC, three patients experienced transient viral reactivations and one developed a secondary autoimmune disease, but all stabilized or resolved with treatment. The study was reported on in the April 4 online edition of JAMA Neurology.

"STEROID" is often used as a generic term to refer to medicines that treat a variety of inflammatory conditions. Most commonly, steroid is used as a shortened name for corticosteroids, also known as glucocorticoids. These medications play important roles in reducing the inflammation that plagues individuals with primary immunodeficiency (PI), as well as helping with side effects that may accompany some of the treatments for PI. Corticosteroids should not be confused with the banned "anabolic steroids" used illegally by athletes to gain advantage in competition. Instead, corticosteroids are legitimately used steroids that offer no athletic advantage, but rather are very helpful for the treatment of inflammatory disorders. Commonly prescribed corticosteroids include prednisone (sold under many brand names such as Deltasone and Sterapred), methylprednisolone (Medrol), prednisolone (Prelone, Pediapred), dexamethasone (Decadron, Hexadrol) and hydrocortisone (Acticort, Cortef). These products are most typically prescribed to treat diseases that cause inflammation such as multiple sclerosis, lupus, rheumatoid arthritis and other autoimmune diseases, as well as skin conditions and cancers.1

Corticosteroids were once considered miraculous when, in 1948, the first patients with rheumatoid arthritis were treated with daily injections and thought to be cured. Subsequently, between 1954 and 1958, six synthetic steroids were introduced for systemic anti-inflammatory therapy. But, by the 1960s, it was discovered that corticosteroids given in high doses over prolonged periods cause serious side effects, and the term “scare-oids” was coined. As such, physicians began prescribing corticosteroids more conservatively, and some fearful patients even declined treatment. But despite the possible harmful effects, corticosteroids administered within proper guidelines can be very therapeutic. So, understanding how they work and how to reduce harmful effects is essential.2,3
What Are Corticosteroids and How Do They Work?

Corticosteroids are drugs that mimic the effects of endogenous cortisols, hormones that are naturally produced in the adrenal cortex (the outer layer of the adrenal gland). Cortisols help to control salt and water balance in the body and regulate carbohydrate, fat and protein metabolism, and, in particular, they are secreted each day to help reduce the inflammation naturally accumulating in the body. The adrenal glands typically produce about 20 milligrams of cortisol a day, but when the body is stressed by situations such as infection, trauma, surgery or emotional problems, they can produce five times that amount to allow the body to cope with the metabolic needs and need for reduction in inflammation.

Treatment with higher doses of corticosteroids — much higher than what the body normally produces — may also suppress the immune system. As such, treatment with higher doses of corticosteroids may help control an autoimmune disease caused when the immune system mistakenly attacks its own tissues. However, they can also impede the normal function of white blood cells, which could increase susceptibility to infections.

The use of corticosteroids, then, may seem to be counterproductive for treatment of PI patients. Why treat someone with a compromised immune system with medications that can result in immunosuppression? Terry O. Harville, MD, PhD, medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences and a consultant for immunodeficiencies, autoimmunities and transplantation, explains that when corticosteroids are prescribed for PI patients, “the dosages being used are for reducing inflammation, trying to keep the dose low enough to not further add to immune system compromise.” In general, he says, taking low doses of corticosteroids once a day in the morning doesn’t result in much immune system impairment in most people. At other times, though, higher doses may be required, which actually may result in some decrease in immune system activity, especially when trying to control some autoimmune manifestations. According to Dr. Harville, it’s like fighting fire with fire — deliberately raising small controllable fires, which are called “backfires,” to remove any flammable material to deprive it of fuel. Yet, even though low-dose treatment with corticosteroids may not further impair already compromised immune systems, it is recommended to employ a team approach by joining the skills of the immunologist with those of a specialist in treating the organ system involved (i.e., gastroenterology, rheumatology, pulmonology, endocrinology, nephrology, dermatology or hematology).

Corticosteroids come in many forms, including pills, injections (for joints or muscles, or via intravenous infusion), ointments and inhalers. They are prescribed for short-term and long-term use, and to minimize dosages, they are sometimes used in conjunction with other non-steroidal anti-inflammatory drugs (NSAIDs) or other disease modifying anti-rheumatic drugs (DMARDs) to act as “steroid-sparing” medications.

By the 1960s, it was discovered that corticosteroids given in high doses over prolonged periods cause serious side effects, and the term “scare-oids” was coined.

Corticosteroids are sometimes needed in conjunction with PI patients’ primary treatment — immune globulin (IG) therapy — that causes side effects such as hives, rashes, difficulty breathing, headache, diarrhea, chills, body aches and other symptoms. Many of these side effects can be reduced or eliminated by pretreating patients prior to IG infusions with a low-dose oral corticosteroid such as prednisone, or the shorter-acting hydrocortisone. In some cases, higher dosing may be required for amelioration of more serious side effects.

Corticosteroids are also used to treat PI patients who suffer from autoimmune disorders. These include, but are not limited to, autoimmune cytopenias, lung disease (e.g., interstitial lung disease and granulomas in the lung), skin disease (eczema, psoriasis, hair and skin pigmentation changes), gastrointestinal disease (Crohn’s disease [aka granuloma colitis] and inflammatory bowel disease), musculoskeletal disease (e.g., rheumatoid arthritis and dermatomyositis/polymyositis), lupus and Sjögren’s syndrome.

PI patients may also suffer from allergies and asthma. Corticosteroids in the form of over-the-counter or prescriptive nasal sprays are often prescribed for nasal allergies. For asthma or other respiratory conditions.
problems, inhaled corticosteroids are prescribed. For more serious cases, courses of oral or intravenous corticosteroids may be required.

“One problem with PI is that, depending on the type of disease condition, parts of the immune system may begin working overtime and create more inflammation and damage,” says Dr. Harville. “We give corticosteroids to help slow down the unnecessarily overactive parts of the immune system, as well as to reduce inflammation.”

According to Dr. Harville, dosing of corticosteroids depends on the disease condition and severity. In general, the lowest dose that provides benefit is desired. “Typically, the initial dose may be 1 mg/kg to 2 mg/kg, which is tapered to the lowest successful dosing,” he explains. Tapering generally occurs every three days. “A dose between 0.15 mg/kg and 0.25 mg/kg prednisone equivalent is considered to be ‘physiologic,’ or equivalent to the body’s own production of endogenous cortisols. Thus, doses less than this are felt to be somewhat safer since they are below the body’s production level. Most physicians try to reduce the dose even more to every-other-day dosing to further reduce the adverse effect of the corticosteroid on normal physiologic production of cortisols.”

Why sub-physiologic levels of corticosteroids control inflammation is not well understood. “There’s something unique about the synthetic cortisols (i.e., perhaps longer lifespans) versus the natural cortisols that makes them better at controlling disease,” adds Dr. Harville.

Several studies have demonstrated successful treatment of autoimmune disorders with corticosteroids in PI patients. In one study, a 23-year-old man with common variable immunodeficiency and symptoms of chronic diarrhea, malabsorption and weight loss that had been apparent for two years was treated with 30 mg prednisone each day for one month. The prednisone was then tapered weekly by 5 mg until it was discontinued. Three months later, the patient’s clinical symptoms disappeared, and his quality of life improved. During the subsequent nine months follow-up, his body weight increased, and he was able to work without suffering any effects from his illness.6

Harmful Effects of Long-Term Corticosteroid Use

Both short-term and long-term corticosteroid use can result in harmful effects in multiple body systems. Short-term effects could include an increase in blood sugar; upset stomach, with stomach ulcers and bleeding; increased hunger; increased risk of pneumonia, thrush and irritability.4

Common long-term adverse effects of oral corticosteroids include elevated pressure in the eyes (glaucoma); fluid retention, causing swelling in the lower legs; persistent high blood pressure; problems with mood, memory, behavior and other psychological effects; weight gain with excess fat deposits in certain areas; clouding of the lens in one or both eyes (cataracts); high blood sugar that can trigger or worsen diabetes; increased risk of infections; thinning bones and fractures; suppressed adrenal gland hormone production; and thin skin, bruising or slower wound healing.4

Side effects of inhaled corticosteroids include fungal infection in the mouth (oral thrush) and hoarseness. Systemic effects may also occur, and there is some risk for cataracts. Topical corticosteroid side effects include thinning of skin, red skin lesions and acne. And, side effects of injected corticosteroids include skin thinning and loss of skin color at the site of injection, as well as facial flushing, insomnia and high blood sugar.4

According to Dr. Harville, whether due to short- or long-term use of corticosteroids, what can be most troubling in some patients are the psychological effects: “Paranoia is one of the major side effects people will experience, resulting in a potential for outbursts, mood swings and other adverse alterations in thought processes. These can be very disturbing to some patients, because they know that it is not them; it seems to be someone else.”

Reducing Corticosteroids’ Harmful Effects

Both physicians and patients can take measures to help minimize the risks of long-term corticosteroid use.

If possible, physicians will opt to prescribe topical or inhaled therapy rather
than systemic (oral or injected) to reduce the effects. “If a PI patient is having diarrhea or granulomas in their intestines, prescribing a form that doesn’t get absorbed very well into the body will result in fewer systemic side effects, and may bring the symptoms under control,” explains Dr. Harville. “But if someone with common variable immunodeficiency has lung disease, they may need the systemic effects of the corticosteroids to gain control and get improvement of the disease.” In these cases, as mentioned earlier, physicians strive to prescribe the lowest dose that provides the needed benefit.

When stopping corticosteroids, patients should be tapered off treatment. This is so their adrenal glands can once again begin producing cortisols. Stopping corticosteroids without tapering can result in the lack of endogenous cortisols and may cause fatigue, joint pain, muscle stiffness, muscle tenderness, fever and even a flare-up of the condition.7

Unfortunately, there are patients who can never go off low-dose daily corticosteroids. These patients will accumulate damage, which is why self-care is essential. Getting an annual flu shot is important because long-term use may result in some reduction in the normal protective role of the immune system. Taking corticosteroids after a full meal or with an antacid may help reduce stomach irritation that often occurs when they are taken along with NSAIDs such as ibuprofen or aspirin.

Physicians may obtain a bone density test at the start of corticosteroid treatment, which can be repeated to assess the effectiveness of measures to prevent bone loss. They may also prescribe bone-preserving medications such as alendronate (Fosamax), calcitonin (Miacalcin), raloxifene (Evista) and risedronate (Actontel). Measures patients can take include consuming calcium supplements and milk products to increase calcium intake to at least 1,500 mg per day; taking a multivitamin to be sure they get a minimum of 400 IU of vitamin D per day to help absorb the calcium; reducing or eliminating smoking and alcohol use; and conducting weight-bearing exercises such as running, walking and dancing to stabilize bone mass.7

Patients should follow a heart-healthy lifestyle by watching calories and exercising regularly to try to prevent excessive weight gain and the development of atherosclerosis.

Patients should follow a heart-healthy lifestyle by watching calories and exercising regularly to try to prevent excessive weight gain and the development of atherosclerosis. Consuming a low-sodium diet will help reduce fluid accumulation and control blood pressure. And, blood sugar levels should be checked regularly. Those patients who already have diabetes should follow their prescribed medical and dietary regimens.7

For mood changes, physicians can sometimes prescribe another medication. In extreme cases of psychological problems, corticosteroid dosage will have to be decreased.7

Overcoming “Scare-oids”

When used long-term and with high doses, as noted, corticosteroids have many possible adverse consequences. “One major dilemma in some patients fearful of corticosteroids, but become educated about them and their adverse side effects. Then, both patients and their physicians can help to minimize side effects while achieving significant benefits. ■

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

References
There are many traits parents hope to pass on to their children, but primary immunodeficiency disease (PI) is not one of them. Understanding the genetics of PI can help identify risk factors and generate conversations with friends, family and physicians.

By Trudie Mitschang
Protect Yourself From a Lapse in Coverage

CSL Behring Assurance℠

CSL Behring Assurance is designed to help ensure that people who rely on CSL Behring Ig therapies can continue to receive treatment even if they experience a lapse in third-party, private health insurance.

- Earn CSL Behring Assurance points for every month of continuous Ig product use
- Points are redeemable for Ig therapy in the event you lose insurance coverage
- Enroll as soon as possible because points are issued from the date that enrollment eligibility is approved

1-877-355-4447
Monday – Friday
8 AM to 8 PM ET

“No one wants to think they will lose their insurance—even briefly—but when you’re dealing with a chronic disorder you always have to be prepared for the unexpected.”
— Lori K.
Parent

CSL Behring Assurance℠ is a service mark of CSL Behring LLC. Biotherapies for Life® is a registered trademark of CSL Behring LLC.

©2016 CSL Behring LLC
1020 First Avenue, PO Box 61501, King of Prussia, PA 19406-0901 USA
A Gap in Your Insurance Coverage Shouldn’t Mean a Gap in Treatment


How to enroll:
• Call the toll-free IgIQ Hotline at 1-877-355-4447

Earning and redeeming points is easy.
• Earn a point for every consecutive month of CSL Behring product use
• Redeem 3 points for a one-month supply of your CSL Behring product*
• Should you suffer a lapse in your insurance, simply contact a IgIQ Care Coordinator and we will take care of the rest

“At the Immune Deficiency Foundation, we often talk to patients who lose their insurance coverage and we see firsthand how stressful this situation can be. This program not only helps people continue their life-saving treatments but also provides security for the future. It is a valuable option for our patient community.”
— Marcia Boyle
President and Founder, Immune Deficiency Foundation

*Certain limitations apply—see program Terms and Conditions at www.cslbehringassurance.com.
WHEN COUPLES DREAM of starting a family, they delight in imagining the physical traits their child will inherit. Will the baby have Mom’s blue eyes, Dad’s prominent nose, Grandpa’s slim physique or Grandma’s curly hair? Unfortunately, when one or both parents carry the genetic makeup of a primary immunodeficiency (PI), passing along DNA from parent to child can go from joyful anticipation to fear and dread. And, in some cases, if one of the parents is merely the carrier of a genetic disorder, the devastating news of a diagnosis does not come until one or more children are already exhibiting symptoms.

Tragedy Leads to Diagnosis
Jessica Johnson was unknowingly a carrier of X-linked agammaglobulinemia (XLA), a condition that affects the immune system and occurs almost exclusively in males. Children with XLA are usually healthy for the first 1 month or 2 months of life because they are protected by antibodies acquired before birth from their mother. Later, as the maternal antibodies diminish, the affected child begins to develop recurrent infections. “When our first child, Emma, was 2 years old, we had Andy. And although Emma was always very healthy, Andy started to get sick at the age of 4 months,” says Johnson. “He suffered from frequent colds, ear infections and sinus infections. He would get 105-degree [Fahrenheit] fevers, and his white cell count could be through the roof.”

Jessica’s third child, Ethan, wrestled with the same cycle of infections as his 2-year-old brother, and tragically succumbed to complications from his illness just prior to a diagnosis. Armed with medical information and still grieving the loss of Ethan, Jessica and her husband, Bart, made the controversial decision to have a fourth child, Gavin, who also has XLA. “I struggled quite a bit with the decision to have more children after our sons were diagnosed, because the diagnosis came at the same time as my son Ethan’s death,” she recalls. “We lost him and got the diagnosis of XLA at the same time.” While Johnson wasn’t done having kids, she wondered whether she could survive XLA taking away another child.

Understanding the Genetics of PI
A short review from high school biology class helps explain the basics of genetic influence on offspring. In the human body, each cell contains 23 pairs of chromosomes, hence 23 sets of genes. One of each pair of chromosomes is inherited from the mother, while the other is inherited from the father. One gene from each biological parent determines characteristics like eye and hair color. All of the chromosomes except the sex chromosomes are called autosomes and are numbered from 1 to 22 according to size. One additional pair of chromosomes determines the sex of the individual. These are called the sex chromosomes and are of two types, X and Y chromosomes (females have two X chromosomes, and males have an X and a Y chromosome). The sex of the baby is determined by which type of sperm (X or Y) fertilizes the egg.1

Most PIs are inherited in one of two ways: X-linked recessive or autosomal recessive. Recessive genetic disorders occur when an individual inherits two copies of an abnormal gene for the same trait, one from each parent. If an individual receives one normal gene and one gene for the disease, the person will be a carrier for the disease but usually will not show symptoms. The risk for two carrier parents to both pass the defective gene and have an affected child is 25 percent with each pregnancy. The risk of having a child who is a carrier, similar to the parents, is 50 percent with each pregnancy. The chance for a child to receive normal genes from both parents and be genetically normal for that particular trait is 25 percent, and the risk is the same for males and females.1

Rarely, the inheritance of a genetic disease is autosomal dominant. Dominant genetic disorders occur when a single copy of an abnormal gene is necessary to cause a particular disease. The abnormal gene can be inherited from either parent or can be the result of a new mutation (gene change) in the affected individual. The risk of passing the abnormal gene from the affected parent to offspring is 50 percent for each pregnancy, and again, the risk is the same for males and females.1

Of course, statistics can never predict every possible variable when it comes to conception, pregnancy and childbirth. Even when the odds seem to be in favor of delivering a healthy baby, genetic outcomes can be wildly unpredictable.

Terry Harville, MD, PhD, medical director of the Special Immunology Laboratory at the University of Arkansas for
Medical Sciences and a consultant for immunodeficiencies, autoimmunities and transplantation, has decades of experience working with families who have wrestled with family planning issues. In his opinion, denial of risk is often a driving force behind the decision to have children once the possibility of an inherited PI has been established. “In situations where the risk of passing on the disease is 50/50 for each pregnancy, parents who are optimistic by nature will still believe the odds are in their favor with the coin flip,” he explains. “Sometimes, when they already have one or two affected children, they believe they’ve used up all of the potential ‘bad luck.’”

Navigating Guilt

In families stricken with genetic diseases, feelings of guilt can manifest in myriad ways. Parents may blame themselves and one another. Family dynamics are also impacted; healthy siblings may feel intense shame wondering why they were spared the life-altering disorder that affected a brother or sister. Expectant parents who learn their unborn child will be chronically ill also confront feelings of guilt, whether they decide to have the child or abort. Even adult children who have accepted the ramifications of their inherited diseases may feel reluctant to discuss their health challenges with parents for fear of instilling guilt.

Experts believe understanding the many layers of guilt as it applies to families with genetic diseases may help researchers better understand the emotion, and may lead to better counseling and support systems for individuals and families. “Psychoanalytically, we look at guilt as an internal process of punishment. You’ve done something bad, and your superego says you have done something bad and need to be punished. It is an internal way of keeping us moral,” says Dr. Philip R. Muskin, a psychiatry professor at Columbia University. “Guilt can be warranted when you’ve really done something wrong, but you can also feel it without having done anything.”

Advances in medicine have only intensified potential feelings of guilt surrounding inherited diseases. Illnesses that were once chalked up to “God’s will” or a combination of unforeseen factors can now be absolutely attributed to a single bad gene inherited from one parent. Dr. Harville has seen this scenario numerous times in his practice dealing with X-linked diseases. “Many men blame the female disease carrier for ‘giving this disease to my child.’ I tell people, you can pick your friends, you can pick your nose, but you can’t pick your relatives. You are stuck with the genetics you come with,” he says. “In spite of feeling guilt about having a sick child, with the father present, it’s important to address the tendency of the husband to blame the wife and confront that issue head-on.”

Dr. Harville notes that in terms of family dynamics, if fathers tend to place blame, mothers tend to bear the burden of responsibility: “The mother tends to ask, ‘How did this happen; what did I do to cause this?’ These questions are normal, and it’s important to work through the facts so that the family can begin to heal.”

Next-Generation Considerations

Myron Anderson’s* mother has common variable immune deficiency, and he and his brother both inherited the disease.

---

* Name changed for privacy.
Because he was diagnosed at an early age and has lived a relatively normal life thanks to immune globulin (IG) therapy, he determined to never let his diagnosis define or limit him, and that has carried over into his views on starting a family. Recently, Anderson learned that his wife is expecting. “Having PI my whole life made me think more about what tests to have when my kids were born, but I never let my disease limit what I can do, so I didn’t let it limit me from wanting kids,” he says. “I figured if any of my kids have a PI, then I already know what to do. It did make dating harder because some women were concerned about my disease passing on to our future children.”

Treatment options like IG can positively influence family planning decisions, especially when couples have quality-of-life concerns regarding children with genetic diseases. “We believe that Ethan only died because the disease was undiagnosed and he hadn’t been receiving IG infusions,” says Johnson. “We knew our older son and future children would not have that disadvantage. XLA was something we felt we could manage, and from what we knew, the kids, if affected, could still live normal and productive lives.”

Pease agrees: “I have no guilt or regrets about my decision to have my three boys. This challenge has taught them to rely on one another, and as adults, they are extremely close.” However, she does suspect that living with XLA may have made her sons hesitant to marry and have children of their own. At 20, 26 and 33, the men remain single, preferring to share a home with one another.

**Pursuing Genetic Counseling**

Speaking to medical professionals who are well-versed in the risks, treatment options and quality-of-life issues of various PIs can help couples make informed decisions about family planning. Simply allowing spouses and family members to become more familiar with the genetic facts of a particular disease can help spark questions and alleviate fears. “In my experience, families often don’t know what questions to ask,” says Dr. Harville. “In cases where we are dealing with something like severe combined immunodeficiency, couples are so attuned to the fact that we are dealing with the prospect of death that fears regarding the immediate future overshadow concerns about long-term treatment and quality of life.”

Dr. Harville says that while genetic counseling can be a benefit, he advises against in utero testing unless abortion is a viable option: “The risk of damaging the fetus is too high, in my opinion.”

According to a journal article published by the Allergy Society of South Africa, due to the complexity of the genetic mechanisms involved in PIs, genetic counseling for patients and their families is recommended. Genetic counseling can assist affected families by helping them understand the cause of the condition and the expected prognosis, and, importantly, what the implications are for them, as well as for extended family members. Genetic counseling plays an integral part in the management of patients with PI and can facilitate genetic testing and treatment plans.

During a genetic consultation, individuals will be asked to provide a comprehensive family history and obtain a disease diagnosis, if one has not already been established. Further discussions will revolve around disease progression, risk of recurrence for future pregnancies and occurrence in other family members. Testing options may also be discussed, including testing of individuals at risk of being carriers and prenatal testing. Supporting the family may also include referrals to other professionals such as mental health counselors and parent support groups. More information on genetic counseling can be found at the National Society of Genetic Counselors (www.nsgc.org).

Whether or not families choose to seek professional advice from a genetic counselor or other healthcare professional, successfully navigating the emotional minefield of genetically inherited diseases always begins with open, honest communication. Dawn Laney, a genetic counselor, research coordinator and instructor in Emory University’s Department of Human Genetics, has written several children’s books on the topic of genetic disease. After diagnosing patients, Laney frequently has to explain the test results and potential treatment options to patients and their families. “How do you explain to a child that he or she is battling a genetic condition? It’s simple: You explain it simply.”

For Anderson, the joyful anticipation of expecting his first child has pushed any fears or concerns to the background. When and if the time comes, he and his wife have a plan in place to discuss PI with their future son or daughter: “We plan on talking to them about the risk for their kids, while emphasizing that they should not let it hold them back from fulfilling their dreams.”

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

**References**

Managing Multiple Healthcare Specialists

Several strategies and tools can help patients deal with the challenge of being cared for by multiple specialists.

By Cynthia Perry

Patients with multiple diagnoses, multiple specialists and medications prescribed by different providers can find managing their healthcare overwhelming. Ideally, patients’ primary care physicians (PCPs) would play “quarterback” for them, but this rarely happens in the real world. PCPs are very busy with full patient loads, electronic medical records to maintain and insurance paperwork to manage. And, while they don’t have the time to know every aspect of their complex patients’ diagnoses and care, the good ones will let these patients be partners in their care.

Unfortunately, doctors often don’t consult each other about their patients’ care. Most often, this is because they want to respect each other’s expertise and authority. If patients have specialists whose areas of expertise overlap, such as immunology, ENT or pulmonology, problems could present if these specialists’ views differ about how patients should be treated.

Understanding these issues, patients can use some strategies and tools to better help manage their medical team.

Managing the PCP Relationship

Many PCPs are limiting visits to discussing one or two issues for insurance billing reasons. As such, complex patients might benefit from making appointments every three months to six
months (in addition to their regular wellness exam) to bring their PCP up to date on medical issues, specialists they have seen, what specialists have recommended, etc. Those appointments are also a good time to review medications and side effects, how well the medical team is working and whether any new specialists would be beneficial. Good PCPs will also check on how patients are holding up under the stress of dealing with complex and chronic health issues and offer suggestions for stress management.

**Medical Summary**

Patients can create a medical summary, which should be updated often, to carry with them to all appointments. Helpful things to include in the summary are current doctors and their specialties and phone numbers, pharmacy contact info, major diagnoses and dates of diagnosis, allergies, daily medications with dosages, as-needed medications, and past surgeries and procedures. The first sheet should summarize as much information as possible; subsequent sheets can offer more detail, if needed.

An easy way to create a medical summary is to sign up for an annual subscription service such as MedicAlert. These types of services provide step-by-step templates for in-case-of-emergency (ICE) situations and full medical histories. Patients can order jewelry with an ID unique to them that will provide healthcare workers the toll-free number to call to get the information needed to care for them during a medical emergency.

Once a medical summary is created, patients can simply write “see attached” on new patient forms and attach a copy of it. This saves patients from having to repeat their story over and over again. It also ensures they don’t leave anything out when meeting new specialists.

**Building the Team**

There are many ways for patients to find specialists for their medical team. Patients can look for university-based providers, HMOs with integrated care models such as Kaiser, or community-based providers in private practice or affiliated with non-university hospitals. Referrals can come from patients’ PCPs, local patient support groups, other trusted specialists, family and friends, or local doctor ratings guides. Sometimes it is a good idea to check with multiple sources and find specialists who are repeatedly recommended.

University-based healthcare providers are often involved in research studies and may know the very latest information about treating patients’ conditions. They are also apt to have electronic medical records that are integrated across departments, facilitating communication between specialists. However, university hospitals can have long waits for services and be difficult to get into for urgent care.

Integrated HMO models can also simplify care coordination for patients without long waits that can be found in university-based health systems. The downside is that, normally, patients can be treated only by in-network specialists. The exception to this is if there is no in-network physician of a particular specialty; in that case, special authorizations need to be obtained to go out-of-network.

Community-based providers are often easier to get in to be seen than university-based caregivers. They may also keep more “sick appointments” open for their patients’ urgent healthcare needs. The care patients receive may feel more intimate, but the doctors may not know as much about the latest treatments available for patients’ care. Also, patients will likely have to do much more of their own care coordination.

**Patients can create a medical summary, which should be updated often, to carry with them to all appointments.**

Regardless of the site of care, it is a good idea for patients to ask new specialists if they are willing to take on their care given their complex medical histories. Not all specialists want to work with patients with complex medical conditions, so it’s better to find this out right away and look for a better match than to waste time on a specialist who isn’t going to want to work with a patient. It is also a good idea for patients to ask a specialist exactly what role they want to play in their care.

**Negotiate Specialists’ Boundaries**

Sometimes, when specialists’ areas of expertise overlap, it may become necessary for patients to decide which specialist they want to take care of each aspect of their health. For instance, an ENT may treat sinus infections, while an immunologist treats all other infections. Patients may actually need to negotiate this...
with their specialists so everyone knows where they fit into the patients’ care team. If the specialists agree, great! If they don’t, patients may need their PCPs’ help in creating a different set of boundaries that will work for the specialists. Or, they may have to find new specialists.

**Running List for Specialists**

A paper notebook or electronic journal can be used for making a separate running list for each specialist. Patients can use this to jot down questions and information they want to communicate whenever something pops into their heads. Sometimes specialists convey information to each other through patients rather than calling each other. For instance, if one specialist wants another to run a test or consider a new medication or dose, they may ask patients to discuss this with the other specialist the next time they see them. The running list is a great way to keep track of these types of requests.

**Symptom Tracker**

Patients can create a symptom tracker to track basic vitals, any major symptoms doctors want reported, days bedridden, sleep habits, antibiotic usage, etc. An easy way to build this is in Excel with dates across the top and symptoms down the side. A simple “x” when symptoms are experienced may be sufficient, or patients can use a pain scale or record any other information in the grid that will be helpful to them and their doctors.

Tracking symptoms over time in a consolidated tool can show important trends in patients’ health, and it can help doctors optimize their patients’ treatment plans. This information can also be used for ongoing disability paperwork.

**Managing All Those Meds**

When complex patients are treated by multiple specialists, there is an increased risk of adverse medication interactions. For patients with a large number of daily and as-needed medications, it can be very helpful to fill all of their prescriptions at a single pharmacy, avoiding mail order if at all possible. Using a single pharmacy improves the odds that potentially dangerous drug interactions are found before any harm is done.

Pill sorters can also be helpful — one by the sink for morning and night pills, and one to carry for daytime pills. Patients can pick a day of the week to fill the pill sorters, which allows them to see if they need to order any prescription refills for the following week. This will ensure they never run out of necessary medications. It is also advisable for patients to travel with at least a few extra days of pills in case there are delays.

**Managing Medical Records**

The length of time for which doctors and medical facilities are required to retain patients’ medical records varies by state. Typically, it ranges from seven to 10 years. However, for certain diagnoses, it can be critical to have records for hospitalizations, surgeries, complete diagnostic testing, etc., for patients’ entire lives. This is especially true for conditions such as primary immunodeficiencies or neurological conditions for which treatments alter future test results, and only cessation of treatment would “prove” patients are still ill.

Patients who have a need to keep decades of old records can create paper or electronic files, one for each diagnosis, and update them with new records as needed. These packets of information can then easily be copied when patients are meeting a new specialist, need to provide proof of disability, or need the specific records related to a diagnosis for any other purpose.

**Say Thank You Often**

Treating patients with complex medical conditions is very rewarding for some specialists because they know they are making a difference in their patients’ lives. A verbal heartfelt thanks in an email or a card on “doctor appreciation day” are all appreciated by providers brave enough to take on the challenges of helping patients with complex and chronic medical conditions.

**CYNTHIA PERRY** is a wife, mother and advocate. She started her career as a technical writer, later transitioning into marketing analytics and strategic planning. Her life and work have taken her to many unexpected places at the forefront of medical and genetic research, and she now shares those experiences with others through her writings.
You can lament what is lost to you, whether it’s opportunity, a person or your health, but clinging to anger is no way to experience life.” — Rebecca Zook in “Life Lessons,” excerpted from Chronic Inspiration. Download a daily dose of inspiration with this heartfelt compilation of writings on life with chronic illness. From coping strategies and parenting tips to “from the trenches” advice on dealing with family and friends who simply don’t get it, these personal stories are sure to uplift, challenge and inspire. Honest and candid, Chronic Inspiration: Heartfelt Perspectives on Life with Chronic Illness gives voice to those who refuse to let their diagnosis define who they are or what they can accomplish.

“For the patient community, this was invaluable. When I downloaded it, I knew this would be something I would refer to over and over again.” — Jenny Gardner

Chronic Inspiration can be purchased on iTunes, Amazon and Barnes and Noble.com
Negotiating Healthcare Costs

Many options are available to reduce the cost of care — before and after treatment.

By Amy Scanlin, MS
HEALTHCARE IS EXPENSIVE, even for those in good health. Add a chronic condition, and costs can be pushed past the breaking point. To keep premiums lower, patients can opt for higher deductibles and co-pays. But this leads to a greater risk of financial hardship when payments for big-ticket services come due.

There has been tremendous growth in the numbers of people, whether uninsured or underinsured, who suffer financially from the cost of healthcare. And while it may seem that these costs are insurmountable, that is not always the case. Hospitals, doctor offices and insurance companies are the primary negotiators of the cost of care, but patients can also impact their own costs. Smart oversight, asking the right questions, knowing where to turn for assistance and keeping a close eye on medical bills can have a positive effect on the bottom line.

Smart Oversight

More and more, consumers expect to understand what things cost, says Mark Rukavina, MBA, founder of Community Health Advisors LLC. “However, with healthcare, that cost can be a mystery.” The multi-party system, he says, involving payees, providers and patients, leads to a lack of knowledge and, in some cases, an inability to get good information.

That’s why patients must keep track of the who and what when it comes to tests and procedures. Going out-of-network and having a lack of clear justification for care are two culprits of unexpected and crippling medical costs. In fact, unexpected out-of-network bills were a top consumer complaint, according to a New York State Department of Financial Services report.1 A simple referral to a specialist or for a specific test by a trusted physician can unwittingly lead to financial disaster. Whether it’s a broken arm, a second opinion or a specialist, any number of unexpected circumstances can arise that can cause costs to skyrocket. It is up to patients to ultimately understand what is covered by their insurance plan. As such, it is imperative they ask the right questions and manage where everything and everyone falls within the system.

Ask the Right People the Right Questions

Calling the insurance company to verify provider participation and covered tests allows patients to go back to their doctor to confer and determine if a more cost-effective option is available. This also enables patients to request better justification from the insurance company in its determination of coverage pre-approval.

“Your health plan can tell you what services or procedures require pre-approval,” says Karen Thomas, director of Healthcare Finance Policy, Consumer Engagement, at Healthcare Financial Management Association. “It’s ultimately the patient’s responsibility to make sure that pre-approval has been obtained when it’s necessary. Ask your doctor if he or she will request the pre-approval, or if you should. Either way, call your health plan before receiving the care to ensure the pre-approval is on record.” Of course, in emergency situations, pre-approval requirements generally do not apply.

Gathering information upfront will help to reduce the element of surprise later. Unfortunately, too many people realize after the fact the questions they should have asked beforehand. “Your health plan can help you compare price and quality for different physicians and hospitals,” says Thomas. “Many health plans have a price estimator, often called a price transparency tool, on their websites for use by their members. Many hospitals can also provide you with price information upon request.”

In addition, online cost aggregators such as FAIRHealth are options that allow patients to see the anticipated costs for services in their area. Costs passed on to patients vary widely depending on negotiations between hospitals and providers and which insurance plan a patient has. So shopping around for a better price, without compromising care, can be a good strategy. But, patients should be sure to compare apples to apples by asking for the exact name of the procedure and the specific billing codes.

Thomas suggests patients and physicians ask questions such as the following to help bring costs down: Are lab tests pre-authorized, including those conducted off-site? Are certain tests even necessary, or can a previous result provide needed detail? What about prescribed medications? Is there a less-expensive alternative?

Gathering information upfront will help to reduce the element of surprise later.

Patients should bring written lists of questions that are prioritized to make the best use of time. “If you are concerned that you won’t remember or be able to process everything,” says Thomas, “bring a friend or a family member with you to your doctor’s appointment. Don’t move forward with treatment until you feel comfortable that you know what to expect and that all your questions have been answered.”
More than 10,000 patients and providers have put their confidence in Hizentra¹

Hizentra offers a range of dosing options:

- DAILY
- ONCE EVERY 2 WEEKS

Hizentra—Ig therapy that fits your lifestyle.

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

WARNING: Thrombosis (blood clotting) can occur with immune globulin products, including Hizentra. Risk factors can include: advanced age, prolonged immobilization, a history of blood clotting or hyperviscosity (blood thickness), use of estrogens, installed vascular catheters, and cardiovascular risk factors.

If you are at high risk of thrombosis, your doctor will prescribe Hizentra at the minimum dose and infusion rate practicable and will monitor you for signs of thrombosis and hyperviscosity.

Always drink sufficient fluids before administration.

Complete Support, All in One Place

IgIQ is a resource center for people who depend on the Ig therapies offered by CSL Behring, including Hizenta.

What can our friendly and knowledgeable IgIQ staff do for you?

- Answer general (non-medical) questions*
- Provide helpful resources
- Enroll you in peer support and financial assistance programs
- Answer questions related to insurance coverage

*IgIQ personnel are not medical experts; medical questions will be forwarded to the appropriate CSL Behring department.

For more information, call 1-877-355-IGIQ (4447) Monday–Friday, 8 AM to 8 PM ET.

Important Safety Information (continued)

Tell your doctor if you have had a serious reaction to other immune globulin medicines or have been told you also have a deficiency of the immunoglobulin called IgA, as you might not be able to take Hizenta. You should not take Hizenta if you know you have hyperprolactinemia (too much prolactin in your blood).

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

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

Tell your doctor about any side effects that concern you. Immediately report symptoms that could indicate a blood clot, including pain and/or swelling of an arm or leg, with warmth over affected area; discoloration in arm or leg; unexplained shortness of breath; chest pain or discomfort that worsens with deep breathing; unexplained rapid pulse; and numbness or weakness on one side of the body. Your doctor will also monitor symptoms that could indicate hemolysis (destruction of red blood cells), and other potentially serious reactions that have been seen with Ig treatment, including aseptic meningitis syndrome (brain swelling); kidney problems; and transfusion-related acute lung injury.

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

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

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

Please see brief summary of full prescribing information for Hizenta on adjacent page. For full prescribing information, including box warning and patient product information, please visit Hizenta.com.

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

Hizenta is a registered trademark of CSL Behring AG. CSL Behring is a registered trademark of CSL Behring LLC.

©2016 CSL Behring LLC
1020 First Avenue, PO Box 61501, King of Prussia, PA 19406-0901 USA
Hizentra®, Immune Globulin Subcutaneous (Human), 20% Liquid
Initial U.S. Approval: 2010

**BRIEF SUMMARY OF PRESCRIBING INFORMATION**

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

**WARNING: THROMBOSIS**

* See full prescribing information for complete boxed warning.

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

**INDICATIONS AND USAGE**

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

**DOSE AND ADMINISTRATION**

For subcutaneous infusion only. Do not inject into a blood vessel. Administer at regular intervals from daily up to every two weeks (biweekly).

**DOSAGE (2.2)**

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

- Weekly: Start Hizentra 1 week after last IGIV infusion
  - Initial weekly dose = Previous IgG dose (in grams) x 1.37
  - No. of weeks between IGIV doses.
- Biweekly: Start Hizentra 1 or 2 weeks after the last IGIV infusion or 1 week after the last weekly Hizentra/IGS infusion. Administer twice the calculated weekly dose.
- Frequent dosing (2 to 7 times per week): Start Hizentra 1 week after the last IGIV or Hizentra/IGS infusion. Divide the calculated weekly dose by the desired number of times per week.
- Adjust the dose based on clinical response and serum IgG trough levels.

**Administration**

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

<table>
<thead>
<tr>
<th>Infusion Parameters</th>
<th>1st</th>
<th>2nd to 4th</th>
<th>5th</th>
<th>6th and above</th>
</tr>
</thead>
<tbody>
<tr>
<td>Volume (mL/site)</td>
<td>≤ 15</td>
<td>≤ 20</td>
<td>≤ 25</td>
<td></td>
</tr>
<tr>
<td>Rate (mL/hr/site)</td>
<td>15</td>
<td>≤ 25</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* As tolerated

**ADVERSE REACTIONS**

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

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

**DRUG INTERACTIONS**

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

**USE IN SPECIFIC POPULATIONS**

- Pregnancy: No human or animal data. Use only if clearly needed.
- Pediatric: No specific dose requirements are necessary to achieve the desired serum IgG levels.

Can IgIQ help you? If you answer YES to any of these questions, call 1-877-355-IGIQ (4447) Monday–Friday, 8 AM to 8 PM ET.
Whether upfront or after a service has been rendered, patients have another option for trying to reduce the cost of care, and it is often the most successful. Simply ask. “The squeaky wheel gets the grease,” says Rukavina. “That is true here with negotiating. Some wonder if negotiating the cost of care is appropriate, and the answer is ‘yes.’ There are all kinds of negotiations going on all the way up the chain.”

If the quoted costs seem higher than area norms, or if they aren’t affordable, patients should feel free to talk with the provider and explain, using the gathered data, why they feel that way. They should also ask about interest-free payment plans or even a discount if the entire bill is paid upfront.

Even asking for a reduced rate after the bill is received can be a viable option. And, self-pay discounts may be provided to those who opt out of insurance or who do not qualify for Medicaid. With 1.7 million Americans declaring bankruptcy due to medical debts, according to Nerd Wallet, finding an acceptable payment option is in the best interest of the patient, provider and insurance company. After all, a fee going to collections is bad for all parties.

Even Asking for a Reduced Rate After the Bill is Received Can Be a Viable Option.

Keep a Close Eye on Bills

Patients should keep a close eye on medical bills as they are received so that concerns can be addressed and payment solutions can be found immediately. While, historically, medical bills have been confusing, this is changing. “Hospitals and doctors are working on developing price estimates for a patient’s share of the bill that give patients the right amount of detail without overwhelming them,” explains Thomas. “It’s a big job, considering that the average hospital price list may include tens of thousands of items. But we know people don’t want the whole phone book, they just want the particular ‘entry’ that corresponds to the care they will receive, taking into consideration the specifics of their health plan coverage. And that’s what we’re trying to give them.”

In the case of extraordinarily high healthcare bills, Rukavina adds that assistance, termed “medical hardship” may also be available. This is true even for those who are not considered low income as long as a large portion of the person’s bills are going to healthcare. Those with an ongoing health need that limits their ability to work may also be eligible for programs that can cover the cost of their care. Hospitals, insurance companies, foundations and associations all may offer assistance, as well as provide additional resources for places to turn to when in need of assistance.

When it comes to assistance, nonprofit hospitals are required to provide financial help, and many for-profit hospitals offer help as well. Assistance may be available even for those who have opted out of purchasing insurance or who do not qualify for other insurance discounts. By the end of 2016, nonprofits will be required by law to have their financial information posted in plain language, including what financial assistance programs are available, as well as an application to apply for assistance.

Rukavina suggests that patients could also reach out to their state ombudsman and other patient protection options in their public sector offices such as the state health insurance commissioner.

Help Is Available

Understanding what assistance is out there and how to pursue it can be a hurdle when in the midst of treatment or caring for a loved one. Help is available, however. Whether upfront or after a bill is received, there are countless resources — including patients themselves — that can ease the financial burden of care.

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

References

Patient-Centered Resources
- USA.gov Help with Bills: www.usa.gov/help-with-bills
- Healthcare Financial Management Association: www.hfma.org/transparency
- FairHealth: fairhealthconsumer.org
- Nerd Wallet Health: www.nerdwallet.com/blog/category/health

Amy Scanlin, MS, is a freelance writer specializing in medical and fitness issues.
MG

Understanding

An incurable but generally treatable autoimmune neuromuscular disease that leads to muscle weakness and fatigue, MG is often misdiagnosed due to symptoms similar to other conditions.

By Jim Trageser

**WITH NO TYPICAL** case — and with the most common symptoms mimicking those of amyotrophic lateral sclerosis (Lou Gehrig’s disease), velopharyngeal incompetence or even a stroke — myasthenia gravis (MG) can be difficult to diagnose. And yet, because it leads to the weakness and extreme fatigue of the voluntary muscles, MG can be life-threatening in many patients, making an early diagnosis and beginning treatment all the more critical.

**What Is MG?**

MG is an autoimmune neuromuscular disease that occurs when the nervous system has difficulty communicating with the voluntary muscles, resulting in weakness and rapid fatigue. The condition generally affects only a select group of muscles in any particular patient at onset, with the most frequent initial symptoms occurring in the facial and ocular muscles. However, MG typically grows more severe, with up to 90 percent of patients experiencing a generalized spread throughout their body within a year of onset.

When the muscles used for breathing are affected, it can prove fatal. Weakness in the mouth or throat can lead to aspiration pneumonia if drooling or difficulty swallowing allows food into the lungs. Patients with MG will experience heightened symptoms following physical activity, and will show marked improvement after resting.

MG was first described in 1672 by English doctor Thomas Willis. In 1877, Samuel Wilks, a London physician, provided the first modern definition of the disease. However, it wasn’t until the 20th century and the discovery of the chemical acetylcholine and its role in neurotransmission that knowledge of MG progressed further. In 1973, a study by J. Patrick and J. Lindstrom established a link between blocked acetylcholine...
receptors and antibodies, giving us our first full understanding of the disease. Subsequent discoveries in molecular biology have further revealed how the disease works at the cellular level.

What Causes MG?

MG is caused by antibodies produced by the patient’s own body interfering with the normal communication between nerves and muscles. When a healthy person wants to move a muscle (for instance, bring a spoonful of breakfast cereal to his or her mouth), the brain sends a signal through the nervous system. When the nerves along the muscles of the arms and hands get the signal, they release acetylcholine, which then travels the very short distance to special neuroreceptors on the muscle (a special protein that reacts to acetylcholine), causing the muscle to contract. When everything goes smoothly, the muscles contract in proper order, and the person delivers a spoonful of raisin bran to his or her mouth.

In patients with MG, however, the body produces rogue antibodies that attack and/or block the neuroreceptors on the muscle tissue so that the muscle never notices the acetylcholine sent by the nerves. No reception, no contraction.
As with many autoimmune diseases, the root cause of the production of these antibodies is not fully understood. MG does not appear to be hereditary. Some cases of MG appear to be tied to abnormalities in the thymus gland — oftentimes a tumor. Scientists suspect that many cases of MG — perhaps most — are triggered by an earlier infection that confuses the body into attacking the neuromuscular proteins on the muscles. MG does not involve the acetylcholine receptors. This is a variant of the disease known as antibody-negative MG. Researchers believe this type of MG involves antibodies attacking another protein known as lipoprotein-related protein 4.

Symptoms of Progression of MG

Initial symptoms of MG vary widely, ranging from double vision or droopy eyelids (more than half of all cases) to difficulty swallowing. Less-common initial symptoms include unstable walking, slurred speech, shallow breathing or general weakness in the limbs. Onset of symptoms is generally rapid, and most patients will see a continuing, significant worsening of their condition — with almost 90 percent experiencing a widespread, general condition within a year of the first symptom. A small number of patients will regress to the point that they suffer a myasthenic crisis, in which the muscles used for breathing are too compromised to provide adequate ventilation. Respiratory assistance is used to stabilize them while treatment is begun to restore muscle strength.

Women under 40 (typically in their 30s) and men over 60 are most likely to develop MG, although it affects all ages and nationalities. Juvenile MG is relatively rare, however.

Spontaneous remission is also rare, except in neonatal cases where the baby contracts it from the mother during pregnancy; these cases usually clear up within a few months of birth.

Diagnosing MG

Because the symptoms of MG are also associated with other conditions, it can be difficult to make a diagnosis.

When patients exhibit any of these symptoms, physicians may examine the eyes for weakness and conduct other standard neurological tests: reflexes, muscle tone, coordination, etc. If MG is suspected, a blood test for the antibodies can confirm most cases (except for antibody-negative MG). About a third of patients with antibody-negative MG will test positive for the anti-MuSK antibody on a blood test, further enhancing doctors’ ability to provide a definitive diagnosis.

For patients who exhibit symptoms but do not display the antibodies on either blood test, doctors may administer the edrophonium test, which involves an intravenous dosage of edrophonium chloride. If symptoms are relieved, this is further evidence of MG. Another test used in determining a diagnosis is single fiber electromyography, which uses mild electrical pulses to test muscle reaction. Further, if the thymus is suspected of being the cause of MG, a chest MRI or CT scan may be ordered.

Treating MG

There is currently no cure for MG, nor a method to prevent it. However, most patients will respond to one of the treatment methods currently available — and be able to lead normal lives.

Depending on the symptoms, the patient’s history and the specific diagnosis, a doctor may prescribe anticholinesterase drugs, such as neostigmine and pyridostigmine. These help block the rogue antibodies, allowing the nerve to once again communicate effectively with the muscles. Other immunosuppressive drugs may be used to limit production of antibodies, including azathioprine, mycophenolate mofetil, cyclosporine or tacrolimus. However, extended use of immunosuppressants can increase the chance of infection, and cause liver or kidney damage.

Another treatment option is plasmapheresis, which is a blood-filtering process similar to dialysis. For MG, it is used to remove the antibodies causing the disease.

If a tumor on the thymus is suspected of being the cause, surgery to remove the thymus may be scheduled. About half of patients who receive this treatment recover completely. In some cases, even those patients without a tumor or thymus abnormality may benefit from removal of the thymus and elect to undergo the procedure.

One of the more recent treatment options is the use of intravenous immune globulin (IVIG), which provides the body with...
normal antibodies that don’t block the muscle receptors, thus providing relief to many patients. IVIG also is less likely to provoke side effects than immunosuppressants or plasmapheresis, although it can take as much as a week before patients begin to show improvement.

IVIG provides the greatest relief to patients with the most severe symptoms, but its effectiveness wanes in some patients after a period of three weeks to six weeks. Thus, it is often used to bring improvement to seriously ill patients until other treatment options can be explored (although some patients will continue on a long-term regimen of IVIG if they continue to show improvement).

**MG Research**

Important advances are already being made in treating the symptoms of MG. There are more than four dozen ongoing studies involving MG listed on the National Institutes of Health’s clinicaltrials.gov website. Among them is a study looking at whether the lymphoma-leukemia drug Rituximab can help control MG. There is also a study investigating whether 3,4-diaminopyridine — used in other countries to treat another autoimmune neuromuscular disease, Lambert-Eaton myasthenic syndrome — might also help the symptoms of MG.

One study from St. Louis University planned to explore whether a subcutaneous (SC) administration of IG would be more effective than IVIG in treating MG. While it was recently terminated due to a lack of test subjects, it is possible it will be restarted. Another study by University Health Network in Toronto, Canada, looked at whether plasma exchange rather than IV was a more effective way to administer IG in MG patients. This study has been completed, but the results have not yet been published.

Additional studies are researching whether self-administered SCIG can be effective in treating MG, whether IVIG is effective in treating advanced (generalized) MG, whether IVIG can be used to ease patients off of long-term corticosteroid treatment, and whether IVIG might help treat MG patients with certain exacerbations.

As researchers gain additional insight into how the neurological system interacts with the muscular system, studies based on that new discovery are likely to yield improved treatment options in the decades to come.

**MG Outlook**

Since there is no cure, nor any preventive treatment, the prognosis for those diagnosed with MG varies according to the severity of each case. Fortunately, most MG patients respond positively to current treatment options and are able to lead full, productive lives.

---

**JIM TRAGESER** is a freelance journalist in the San Diego area.

---

**References**

LET’S TALK

PROFILE:
Ashlie Huss

By Trudie Mitschang

Trudie: You were diagnosed with CVID in December 2014. What led up to that diagnosis?
Ashlie: I’ve had a history of recurring sinus infections and bronchitis. I have also had two cases of shingles and a few cases of pneumonia. My doctors didn’t really think much of it until I saw a rheumatologist for a second opinion. She ordered many rounds of blood work, including my immunoglobulin levels. My IgG, IgM and IgA all came back very low. They thought it was just my lupus medications causing this, and didn’t seem too concerned. I was eventually referred to a clinical immunologist at the University of Pennsylvania. She’s the one that ultimately made the CVID diagnosis.

Trudie: How often do you have infusions?
Ashlie: I administer Gammagard subcutaneously once a week. What I love about subcutaneous administration is that you can take it with you and literally infuse all over the world. Besides infusing at home, I’ve infused at Disney World and in Disneyland hotel rooms, in New Orleans at the Immune Deficiency Foundation conference and even in Hawaii.

Trudie: You live with several chronic illnesses. How do you stay optimistic?
Ashlie: I live for the moments when I get to cross things off my bucket list and go to places I’ve never been before — the moments when I do things I used to believe were impossible; the moments of sheer goofiness with friends and family when we end up laughing so hard that we’re crying. Despite all my pain and challenges, life is still so beautiful and wonderful, and I just have to keep reminding myself to keep finding and appreciating those moments and to enjoy the little things.

Trudie: What inspired you to start running half marathons?
Ashlie: In the spring of 2014, I was sick of being tired and tired of being sick. I knew I needed to make some changes in my life, and I was inspired by a former colleague who completed the Disney Princess Half Marathon to give running a try. I officially caught the running fever one morning when I was counting my pills. I counted 13 of them and something hit me. I knew I needed to run the half marathon — one mile for every one of those stupid pills I needed to take before I became “functional” every morning. My first-ever half marathon was in February 2015. It was such a struggle, but after I crossed that finish line, I was hooked and couldn’t wait to tackle my next one.

Trudie: Many people with primary immunodeficiency diseases (PIDs) find exercise a challenge. What advice do you offer them?
Ashlie: I’d recommend you just do what you can and celebrate the good days when it’s less of a struggle to get out of bed. Even if you just complete a 15-minute walk, go for it; you can slowly build up your stamina and endurance.

AT 25, ASHLIE Huss is an outgoing young woman who loves running half marathons, volunteering in her community, traveling and almost anything “Disney.” As a patient living with multiple chronic illnesses, including lupus and common variable immune deficiency (CVID), she’s also living proof that with optimism, tenacity and courage, you can overcome even the most insurmountable odds.

Ashlie Huss was diagnosed in her early 20s with common variable immune deficiency after recurring infections and two cases of pneumonia.
I’ve learned that every step, every mile is something worthy of celebration. I’ve also had plenty of bad days when I’m too sick to leave my couch and running seems impossible.

**Trudie:** On your blog, you shared an interesting experience about traveling on a plane while wearing a face mask. Tell us about that.

**Ashlie:** It was early December 2014 when I knew my immunoglobulin levels were low, but I didn’t have the official CVID diagnosis. I was in the Orlando airport struggling about whether I should put on my face mask or not. I knew it was cold and flu season, and I was at a much higher risk for infections, yet I was really self-conscious. Wearing a mask was a physical sign to the outside world that I was quite sick. I ultimately chose to wear it, and I’m glad I did.

Once on the plane, I had an encounter with a little girl and her family who were coming home from a Make a Wish trip to Disney World. I’m guessing the little girl was also struggling with not wanting to wear a face mask because her mom said: “Look at this big girl wearing her face mask like a good girl.” At that point, I unintentionally became a role model. For the rest of the flight, I proudly rocked that face mask.

**Trudie:** Tell us about your goals regarding advocacy work.

**Ashlie:** I’ve found ways to incorporate spreading awareness into my outfits for my half marathons. I still have a voice and such a unique story, and I can’t be afraid to share it. I want to impart change, help raise much-needed funds for research and help get the word out that, yes, these illnesses do exist and they can impact people at any age. If I had known more about PI, I probably would have gotten my diagnosis much sooner.

**Trudie:** What has living with chronic illness taught you about yourself?

**Ashlie:** Chronic illness has taught me all about bravery. The sheer amount of bravery it takes to get through another day during a flare or to face another specialist can be mind-boggling at times. Bravery is found every day when fighting your hardest not to become another disease statistic despite the overwhelming odds stacked against you. Bravery is found in owning your story and not being afraid to open yourself up and share that story. Living with these illnesses has taught me that I’m braver and more courageous than I ever thought I could or would be.

**Trudie:** Many PI patients “don’t look sick.” How do you deal with people who don’t understand your invisible illness?

**Ashlie:** I tell them that’s the thing about invisible illnesses, you can’t see what’s going on inside, nor would you want to see all that chaos. For the most part, people have been quite understanding (or do their best to understand), and I’ve only had to deal with a few who have said I’m not truly sick and I’m making all of this up for attention.

**Trudie:** What’s the biggest emotional challenge you face on a regular basis, and how do you overcome it?

**Ashlie:** The biggest emotional challenge is knowing that these illnesses aren’t going away anytime in the near future, and they’ll probably be with me the rest of my life. All of that can get kind of overwhelming and depressing. Instead of getting stressed out about the big picture and what the future may hold, I try to just focus on the present and enjoy each day for what it is.

**Trudie:** What inspires you?

**Ashlie:** I spent a few summers volunteering at a summer camp for children with chronic or life-threatening illnesses. My campers inspired me to keep living life to the fullest and to embrace the struggles. I also belong to a group called I Run 4 that matches people with disabilities to runners. The runners dedicate their runs or workouts to their buddies and end up forming relationships with their families. I’ve been matched as a runner since September, and I’ve been so inspired by my little buddy and her family. Thinking of her and her family inspires me through my challenging days.

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

"Tired of being sick," Ashlie was inspired by a former colleague to begin running half marathons.
Coping with the Loss of a Support System Star

By Stacy Oliver

HAVING A CHRONIC illness is hard on so many levels: physically, mentally, socially and the list goes on. Over the years, I have developed a core group of people who are my lifeline to feeling loved, keeping a positive attitude and helping me cope with medical setbacks. Since being diagnosed with a rare autoimmune disease, a little part of me has always thought: “I’m going to die before all the people I love.” Even though immune globulin (IG) therapy helps me live a full life and I’ve not been told that I will pass away any time soon, death just seems imminent because my body is so medically complicated. So to have one of my greatest supporters die before me is a void I can’t comprehend.

Last autumn, my mother passed away. She had a quick and painful battle with lymphoma that ended with her dying in a month’s time. For so many reasons, I am devastated she is gone. But along with my husband, she was one of the biggest parts of my support system. When my hands were too weak to work, she would cut my food. When I couldn’t drive to the neurologist one more time because I was gripped with fear over what might be discovered, she drove me. When I was depressed and in the depths of despair, she gave me encouragement. How am I supposed to carry on now without her guidance and support?

I’ve always known that death is a part of life. As my therapist once jocularity stated, “No one gets out of here alive.” My own relationship with death is very simple: I don’t fear dying; I never have. I can’t control that fate, so when it happens, it happens. But it’s those left behind, dealing with my absence, that I will feel sadness for. Now, I am that person left behind, trying to fill the hole in my heart. It has been a soul-searching period that has profoundly affected me, and I am humbled by this life-altering change. To anyone who has experienced this kind of loss, I hope what has worked for me is useful to you. I offer these thoughts as a fellow human being trying to connect with my kindred spirits treated with IG:

• Take all the good memories and loving moments and wrap yourself in them — like lotion you put on your skin to feel hydrated and healed. All those positive memories are yours to keep, making you feel strong and peaceful.

• Feel it all: the deepest sadness, the angriest hurt, the numbness of not being able to feel anything, and then the bits of joy that life gives you, because life does go on. Listen to your heart and head as they both come to terms with the reality of loss.

• Share your feelings, your stories and your life. You don’t have to carry your grief alone. Those around you want to help, and you’ll find you’re more connected to people than you thought. Take comfort in others’ help. As John Donne writes in his poem “No Man Is an Island”: “No man is an island, entire of itself; every man is a piece of the continent, a part of the main.” An honest moment of truth about your pain or sharing a story about your loved one might change not only your life, but help another person as well.

As fellow IG patients, we share a special bond. We gather strength from our community. I am honored to connect with you through this helpful magazine. ■

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

By Ilana Jacqueline

MY MOTHER AND I both suffer from primary immunodeficiency disease. My mother also has fibromyalgia that she chooses to treat without pain medication. For a woman who works in a high-stress job as a television producer, it’s hard to see her hurting when I know she has so much weight on her shoulders.

She has always been a caretaker for me, and, now, as her parents age, she’s become the main caretaker for them, too. Between my migraine spiral, my grandfather’s lung cancer and my grandmother’s colitis, my mother has spent almost the entirety of this month at someone’s bedside in the emergency room. She’s there to take the lead, tell the story and share our preferences when we are no longer able to voice them ourselves.

And her disease doesn’t care. It’s not like she gets to take a step back and say: “Whoa there, aching joints! Somebody else needs my attention!” She’s at the mercy of her symptoms just as much as we all are at any inopportune moment.

I’ve often pondered the strength of caregivers. I know my new husband, RJ, loves me, but the fact that he can keep his cool when I roll over in the middle of the night and say it’s time to go to the ER is something that’s still hard to process. There are lots of events and activities I care about. There are parties I want to go to and friends I want to see, but if I’m knee-deep in a bad flare, even these fun times won’t happen.

So, how do patient-caregivers make moments of incredible support happen when someone else is in need?

On Saturday night, RJ and I went to a housewarming party for one of our old friends. We hadn’t seen most of our social circle since we got back from our honeymoon. We spent three hours (about an hour longer than I usually last at parties) catching up. When we finally got in the car to leave, my body felt so weighted and exhausted, I felt like I might just sink through the leather seat. It had been a stressful touch-and-go day all around. My mom and grandfather had been monitoring my grandmother, who was in critical condition at the hospital. We had wanted to stop by before the party, but were told to just come in the morning.

When we got home around midnight, I was lying on my couch thinking all I wanted to do was jump in the bath, cozy up and go to bed. Instead, my phone rang. I knew immediately that it was going to be bad news and, without hesitation, we were out the door and in the car, begging to catch every green light on our way to the hospital.

My husband parked while my stepdad met me out in front of the emergency room. The two of us rushed through the hallways, my heart pounding. There was no exhaustion anymore, no pain. Just someone I loved waiting for me to come say goodbye.

When we got to my grandmother’s room, I wasn’t surprised to see my mother already there and holding her hand. I went to her other side and took her hand. You could tell just by her eyes that this was the end. She wasn’t going to make it through the night.

And I felt so grateful to be in this moment. There was incredible grief, but my tears were for the realization that if I had even stopped to think about how bad my body hurt after that day, I might not have come in the middle of the night. I might have waited until the morning and lost my chance to say goodbye.

I leaned on my husband as my own fatigue hit while we were waiting, watching over her. I knew that we could stay as

We all rely on someone to take care of us, till the very end.

When we got home around midnight, I was lying on my couch thinking all I wanted to do was jump in the bath, cozy up and go to bed. Instead, my phone rang. I knew immediately that it was going to be bad news and, without hesitation, we were out the door and in the car, begging to catch every green light on our way to the hospital.

My stepdad took care of my mother that night. And all of us (including my uncle who had flown down in the nick of time) were there to support my grandfather — and each other. There is no secret to the willpower of withstanding our own pain as patient-caregivers; it’s a simple explanation: We all rely on someone to take care of us, till the very end.

ILANA JACQUELINE is a 26-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.letsfeellbetter.com is both a personal collection of anecdotes about life with chronic illness, as well as a resource for patients of all ages.
SENDING A CHILD off to college is a milestone of success in the parenting journey. It’s a sign that parents have raised their children to adulthood and prepared them to face the world. And while they’re proud of their children and the things they’ve accomplished in 18 short years, parents may feel a great deal of anxiety when their children leave the nest and they’re no longer under the same roof or within range of their parents’ watchful eyes.

For the parent of a child with primary immunodeficiency disease (PI), this anxiety is compounded. Beyond the normal parental concerns, like whether the child is studying enough or partying too much, parents of PI college students have to worry about whether their children are staying healthy. Will they take their health condition seriously? Without Mom or Dad’s gentle reminders, will they remember to take their medicine or do their treatments regularly?

While it may present some obstacles, PI does not have to stand between any young adult and his or her college degree. It is estimated that roughly 7 percent of college-aged people (18 years to 23 years) in the United States today are living with some type of chronic illness. And even though college students are adults and should be able to manage their own healthcare and academics, they don’t need to do it alone. Parents can help their children succeed during the college years — even before the first day of class.

Find a new doctor before leaving home. Unless the college-bound child is attending a local community college in your hometown, going to college will require moving across the state, or even across the country. When that first illness strikes, it is important that the student has already established a relationship with a nearby doctor who understands his condition. Before the child leaves home, talk to his primary doctor and ask for recommendations of doctors who specialize in, or are at least educated about, PI or whatever his specific condition may be.

John Boyle, vice president of development for the Immune Deficiency Foundation, who completed his undergraduate and graduate degrees while living with X-linked agammaglobulinemia, is no stranger to the challenge of balancing higher education and chronic illness. He recommends finding a doctor who takes the condition seriously and is willing to communicate with specialists who may have been part of the child’s care team in the past. Boyle says, “If [the student’s] condition is reasonably well-managed and they have a good primary care physician, they can hopefully take care of the specialist appointments while on break.”

It is also a good idea for parents to go over their child’s health insurance policy before he or she leaves for college, to ensure that it will cover out-of-state costs.

Locate special services on campus. With so many young people living with a chronic disease today, colleges have become more proficient at accommodating students with special medical needs. When parents and their child arrive on campus for orientation, the first stop should be the office of disability services. These helpful people can arrange certain accommodations, such as a single room or a special diet, for those living with a chronic medical condition. If the student’s illness is one that may affect class attendance, the office of disability services can work with the instructors if an extension is needed for tests or papers. If there is no disability office on campus, the student services department should be able to assist in finding help.

It’s also a good idea to stop by the campus health center and get to know the staff. The medical personnel on campus should be familiar with the student’s condition and know what steps to take should an emergency arise.
The college admissions department can assist students with purchasing a student insurance policy, which is often quite affordable, if necessary. By learning to utilize the school’s health programs and medical services, the first few months of college will likely seem much less challenging.

**Decide on living arrangements.** If the college-bound student is attending school far from home, he or she will have to decide where to live while school is in session. Many college freshmen choose to live in dorms. Living in close quarters with others makes college a breeding ground for infections; and certain diseases, like meningitis, tend to be more prevalent on college campuses. As such, having a chronic illness makes staying healthy in a college setting all the more challenging. Should a student with PI avoid living in a dorm altogether or opt for a single room?

According to Boyle, dorm life was one of the best things about college: “For me, dorm life was great. If your condition is reasonably well-managed, there’s nothing like the experience of living in a dorm. I understand that many parents and some young people living with PI can be skittish about the prospect of a dorm, and it may not be the right choice for everyone. That being said, if it’s even a possibility, I would strongly advise that young people with PI consider going for it if it’s of interest. While you have to keep yourself healthy, you can’t let fear of infection cause you to miss out on amazing experiences.”

Of course, there are ways to make dorm living more “healthy” for the PI college student. If the student is able to receive vaccinations, parents should make sure all immunizations required by the school are up-to-date, plus those recommended by the doctor. To avoid picking up a bug, chronically ill students should avoid sharing with their roommates too liberally. While it may seem like common sense, it doesn’t hurt to remind them not to share personal items like toothbrushes, razors, eating utensils and towels.

**Create a support circle.** College is an exciting time when young adults can reinvent themselves, make new friends and begin to live their own lives. No college student wants to be labeled as “chronically ill.” While there’s no need for any student to walk around with the letters “PI” tattooed across his or her forehead, it’s still a good idea to tell a few trusted individuals, such as a roommate, resident advisor or professor, about the condition. Someone other than the student needs to know what to do in case of an emergency. Those individuals should know where the student keeps his or her medication, and should have the name and number of the student’s doctor, as well as numbers of family members back home.

**Let them fly.** As a parent, Boyle understands the protective impulse to make the situation as “safe” as possible. But he’s also an adult living successfully with PI. “If your children are going to thrive on their own in life,” he says, “you have to let them get used to them taking the reins. Give them the tools that they need, and then let them go.”

And for the future college student, Boyle’s advice is this: “The one thing that you can’t do is to ignore the absolute importance of getting your immune globulin (IG) therapy when you’re supposed to get it. If you’re out on your own at college, worry less about all of the hypotheticals (mono, communal bathrooms, etc.) and more about the one thing that is guaranteed to affect your health if you’re not careful about it. Whether it’s an issue of the hassle of scheduling an intravenous IG infusion, or slacking off when it comes to infusing your subcutaneous IG, you have to be mature enough to take care of that one area of your life. There’s no ‘taking a break’ from your IG. If you understand that, then you’re ready for college.”

Yes, it can be downright scary for parents to let go, to lengthen the parental reins and eventually hand them over to their children completely. Nevertheless, children with PI must become adults with PI. Taking the time to prepare them for college by educating them about their choices and the services available to them before they leave home can make the transition to adulthood less traumatic, and instill parents with the confidence that, yes, their children will be all right.

**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**

**While it may present some obstacles, PI does not have to stand between any young adult and his or her college degree.**
THE GASTROINTESTINAL tract is the largest lymphoid organ in the body, so it is not surprising that food allergies and intestinal diseases are common among primary immunodeficiency disease (PI) patients. One such disease affecting the PI community is celiac disease, in which affected people experience gastrointestinal symptoms after ingesting anything containing gluten.

Understanding Celiac Disease

Celiac disease is an autoimmune disorder in which the body mistakenly reacts to gluten, a protein found in wheat, barley and rye, as if it were a poison. It’s important to note that celiac disease is not a food allergy, although it is sometimes treated like one. The disease affects approximately one in 133 people, although it is often difficult to diagnose.

When individuals with celiac disease eat anything containing gluten, their immune system reacts by destroying the part of the small intestine that absorbs vital nutrients. This malabsorption can lead to serious illness.

Symptoms of celiac disease are numerous and can include:
• bloating, gas, diarrhea
• weight loss or gain
• constant fatigue or weakness
• headaches
• infertility
• depression that does not respond to medication
• abdominal pain and bone pain
• anemia

Diagnosis of celiac disease is made through a series of blood tests, followed by a biopsy of the small bowel to determine if the absorbing lining is damaged. The only treatment for celiac disease is to follow a strict, gluten-free diet.

Gluten-Free Guidelines

The challenge for many people going gluten-free is the emphasis on what not to eat. The list of foods to avoid is lengthy, and often includes many dietary favorites such as bread, cookies, cakes, cereal and pasta. The good news is the food industry and even many restaurants are now catering to individuals who must avoid gluten for health reasons. Focusing on what one can eat rather than what one must avoid can help individuals focus on getting and staying symptom-free. For example, instead of wheat flour, individuals with celiac disease can use potato, rice, soy or bean flour. There are also many gluten-free types of bread, pastas and other products in stores, and many websites offer them. The foundation of a gluten-free meal plan will consist of meat, fish, rice, fruits and vegetables.

One of the reasons going gluten-free is an initially daunting task is that wheat and wheat products are often used as thickeners, stabilizers and texture enhancers in foods that might otherwise seem wheat-free, such as popular salad dressings. Since the term “gluten” is rarely used on product labels, it is vital that a person on a gluten-free diet learn the typical places that gluten can hide. Thanks to the Food Allergen Labeling and Consumer Protection Act of 2004, if a food or an ingredient contains wheat or protein derived from wheat, the word “wheat” must appear clearly on the food label.

Other common foods that may be made with gluten-containing grains include bouillon cubes, rice mixes, potato chips, hard candy, licorice, jelly beans, cold cuts, hot dogs, salami, sausage, French fries, gravy, soy sauce and many low- or nonfat products. Surprisingly, even some medicines and mouthwash contain gluten as an inactive ingredient. When reading medication labels, individuals should look for the words “starch” or “stabilizer” in the inactive ingredients list, and consult with a pharmacist if they’re not sure.

Lastly, it’s not just what one eats but also what one drinks that may trigger symptoms of celiac disease. Distilled alcoholic beverages such as vodka, gin and whiskey are gluten-free, even though they may be made from gluten-containing grains like barley. Wines are also gluten-free, but beer, ale, lager and malt vinegar are not; all contain small amounts of gluten and must be avoided on a gluten-free diet.

TRUDIE MITSCANG is a contributing writer for IG Living magazine.
Dining-Out App
Find Me Gluten Free helps individuals search for gluten-free dining options by entering a specific location or browsing the “popular chains” option, which includes links to well-known restaurant websites and menus.
Free; findmeGlutenFree.com

Gluten-Free Restaurant Cards
This handy tool from CeliacTravel.com ensures an individual’s message regarding the need for a gluten-free meal does not get lost in translation on its way to the kitchen. The restaurant cards come in 54 languages to use when dining out at a restaurant either in the U.S. or abroad.
Free; CeliacTravel.com

Gluten-Free Medication Guide
This helpful guide from the Celiac Disease Foundation allows individuals to see which medications to avoid and helps educate about common over-the-counter products containing gluten.
Free; celiac.org/live-gluten-free/glutenfreediet/gluten-medication

Cook Smart
Gluten Free Cookbook for Busy People on a Budget: 50 Delicious 30-Minutes-or-Less Recipes for Weight Loss, Energy & Optimum Health features tasty recipes for those who need to live a delicious, nutritious gluten-free lifestyle. The book touts that each dish can be made in 30 minutes or less.
Paperback $7.19; Amazon.com

Dining-Out App

Gluten-Free Restaurant Cards

Gluten-Free Medication Guide

Gluten-Free On-the-Go
$2.99; Amazon.com

Gluten-Free On-the-Go

Gluten-Free Resources

Gluten-Free On-the-Go

Gluten-Free Medication Guide

Gluten-Free on-the-Go

Gluten-Free Resources

Gluten-Free Medication Guide

Gluten-Free On-the-Go

Gluten-Free Resources

Gluten-Free Medication Guide

Gluten-Free On-the-Go

Gluten-Free Resources
BOOK CORNER

**How Many Marbles Do YOU Have?: Helping Children Understand the Limitations of Those with Chronic Fatigue Syndrome and Fibromyalgia**

Author: Melina Malott
Publisher: Amazon Digital Services

A mom uses a jar-and-marble analogy to teach her son about her limitations related to chronic fatigue syndrome and fibromyalgia. The book uses marbles as a measure of the mother's limited energy. Using a jar and some marbles, the author conveys difficult concepts such as taking preemptive rests to have more energy later, finding alternate ways to perform tasks that use less energy, and postexertional malaise in terms that children can understand. The concepts in the book are relevant to someone with one or both illnesses, and can be applied to other physically limiting conditions as well. The book reminds the reader that although illness may limit a mother's activities, it never diminishes a mother's love for her children.

**How to Heal Yourself When No One Else Can: A Total Self-Healing Approach for Mind, Body, and Spirit**

Author: Amy B. Scher
Publisher: Amazon Digital Services

Using energy therapy and emotional healing techniques, this book shows patients how to achieve complete and permanent healing by loving, accepting and being themselves no matter what. After overcoming a life-threatening illness, energy therapist Scher had an epiphany that healing is more than just physical. Her story serves as a powerful example of how beneficial it is to address emotional energies, particularly when nothing else works. Scher presents an easy-to-understand, three-part approach to using energy healing for removing blockages, changing a person's relationship with stress, and coming into alignment with who a person truly is.

**The Spectrum of Family Caregiving for Adults and Elders with Chronic Illness**

Editors: Louis D. Burgio, Joseph E. Gaugler and Michelle M. Hilgeman
Publisher: Oxford University Press

This book is written for individuals in the helping professions who are in roles that interface with or serve family caregivers who are supporting an adult or elder with a chronic condition. The volume includes eight disease-specific chapters written by experts from various disciplines discussing the caregiving role, including related care needs, issues and challenges unique to that chronic illness. An Evidence Table is included in each chapter so readers can easily judge the quality of evidence supporting the intervention studies. This book is written for clinicians and those in the helping professions, as well as academics and researchers with an interest in the study of family caregiving and caregiver interventions, and to health administrators, public officials and policy makers.

**Living Incurably Despite Chronic Illness**

Author: Ashley Jane Kneeland
Publisher: Amazon Digital Services

This book is a little bit of tough love and a little bit of irreverent humor. The author describes it as “self-help, but of the direct and concise variety, because when you have an incurable disease, while you do need someone who can relate to what you’re going through, what you don’t need is collective misery.” This book takes what is common in all chronic illness conditions, and focuses on making patients feel less alone, all the while encouraging them to avoid being a victim.
### Ataxia Telangiectasia (A-T)
- **WEBSITES**
  - A-T Children's Project: [www.atcp.org](http://www.atcp.org)

### Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
- **WEBSITES**
  - GBS/CIDP Foundation International: [www.gbs-cidp.org](http://www.gbs-cidp.org)
  - The Foundation for Peripheral Neuropathy: [www.foundationforpn.com](http://www.foundationforpn.com)

### Evans Syndrome
- **ONLINE PEER SUPPORT**
  - Evans Syndrome Research and Support Group: [www.evanssyndrome.org](http://www.evanssyndrome.org)

### Guillain-Barré Syndrome (GBS)
- **WEBSITES**
  - GBS/CIDP Foundation International: [www.gbs-cidp.org](http://www.gbs-cidp.org)
  - The Foundation for Peripheral Neuropathy: [www.foundationforpn.com](http://www.foundationforpn.com)

### Idiopathic Thrombocytopenic Purpura (ITP)
- **WEBSITES**
  - ITP Support Association – UK: [www.itpsupport.org.uk](http://www.itpsupport.org.uk)
  - Platelet Disorder Support Association: [www.pdsa.org](http://www.pdsa.org)

### Kawasaki Disease
- **WEBSITES**
  - American Heart Association: [www.heart.org/HEARTORG/Conditions/More/CardiovascularConditions/Childhood/Kawasaki-Disease_UCM_308777_Article.jsp#T1T2boePWE0](http://www.heart.org/HEARTORG/Conditions/More/CardiovascularConditions/Childhood/Kawasaki-Disease_UCM_308777_Article.jsp#T1T2boePWE0)
  - Kawasaki Disease Foundation: [www.kdfoundation.org](http://www.kdfoundation.org)
  - KidsHealth: [kidshealth.org/parent/medical/heart/kawasaki.html](http://kidshealth.org/parent/medical/heart/kawasaki.html)

### Mitochondrial Disease
- **WEBSITES**
  - United Mitochondrial Disease Foundation: [www.umdf.org](http://www.umdf.org)
  - MitoAction: [www.mitoaction.org](http://www.mitoaction.org)

### Multifocal Motor Neuropathy (MMN)
- **WEBSITES**
  - The Foundation for Peripheral Neuropathy: [www.foundationforpn.com](http://www.foundationforpn.com)

### Multiple Sclerosis (MS)
- **WEBSITES**
  - All About Multiple Sclerosis: [www.mult-sclerosis.org/index.html](http://www.mult-sclerosis.org/index.html)
  - Multiple Sclerosis Association of America: [www.msaa.org](http://www.msaa.org)
  - Multiple Sclerosis Foundation: [www.msfocus.org](http://www.msfocus.org)
  - National Multiple Sclerosis Society: [www.nationalmsociety.org](http://www.nationalmsociety.org)

### Myasthenia Gravis (MG)
- **WEBSITES ANd CHAT ROOMS**
  - Myasthenia Gravis Foundation of America (MGFA): [www.myasthenia.org](http://www.myasthenia.org)
  - Genetic Alliance: [www.geneticalliance.org](http://www.geneticalliance.org)

### Myositis
- **WEBSITES**
  - The Myositis Association: [www.myositis.org](http://www.myositis.org)
  - Juvenile Myositis Family Support Network: [www.myositis.org.uk](http://www.myositis.org.uk)

### Myositis Support Group – UK: [www.myositis.org.uk](http://www.myositis.org.uk)

### Peripheral Neuropathy (PN)
- **WEBSITES**
  - Neuropathy Action Foundation: [www.neuropathyaction.org](http://www.neuropathyaction.org)
  - Western Neuropathy Association: [www.pnhelp.org](http://www.pnhelp.org)
  - Texas Chapter of the Neuropathy Association: [www.handsfeetheart.org](http://www.handsfeetheart.org)
  - The Foundation for Peripheral Neuropathy: [www.foundationforpn.com](http://www.foundationforpn.com)

### Primary Immune Deficiency Disease (PI)
- **WEBSITES**
  - Immune Deficiency Foundation: [www.primaryimmune.org](http://www.primaryimmune.org)
  - Jeffrey Modell Foundation: [www.info4pi.org](http://www.info4pi.org)

### Pemphigus and Pemphigoid
- **WEBSITES**
  - The International Pemphigus and Pemphigoid Foundation: [www.pemphigus.org](http://www.pemphigus.org)
  - Multiple Sclerosis Association of America: [www.msaa.org](http://www.msaa.org)
  - National Multiple Sclerosis Society: [www.nationalmsociety.org](http://www.nationalmsociety.org)

### Pemphigus and Pemphigoid
- **WEBSITES**
  - The International Pemphigus and Pemphigoid Foundation: [www.pemphigus.org](http://www.pemphigus.org)

### Scleroderma
- **WEBSITES**
  - Scleroderma Foundation: [www.scleroderma.org](http://www.scleroderma.org)
  - Johns Hopkins Scleroderma Center: [www.hopkinsscleroderma.org](http://www.hopkinsscleroderma.org)

### Scleroderma Support Forum:
  - Scleroderma Network: [www.sclero.org/support/forums/a-to-z.html](http://www.sclero.org/support/forums/a-to-z.html)

### Stiff Person Syndrome (SPS)
- **WEBSITES**
  - Stiff Person Syndrome: [www.stiffpersons.net](http://www.stiffpersons.net)
  - Living with Stiff Person Syndrome (personal account): [www.livingwithspss.com](http://www.livingwithspss.com)
  - Stiff Person Syndrome: [www.stiffpersons.net](http://www.stiffpersons.net)
Help IG Living Magazine Go Green
Join our campaign to reduce unnecessary paper consumption!

Here’s how you can help: If you can forgo receiving a hard copy of the magazine and utilize the digital version instead, go to www.IGLiving.com to select the Go Green tab to sign up for the electronic version and opt out of the print version.

The Benefits of Going Digital for You!
- Get notified and enjoy earlier online access to every new issue
- Print individual articles to keep or hand out to friends, family and care providers
- Easily share articles instantly on Social Media
- Read the issues anywhere at any time on all of your digital devices (smartphone, computer, iPad, tablet)
- Quickly access all published articles
Making a difference in Our Patients’ Lives.

Specialty Solutions in Chronic Care

- Immune Globulin Intravenous
- Immune Globulin Subcutaneous
- Antihemophilic Factors

NuFACTOR has the distinction of carrying all U.S.-approved immune globulin products. Committed to exceptional customer service, product and patient safety, and secure product availability and affordability, we’ve earned the most respected name in homecare because our customers know we care about them. And that makes all the difference.

NuFACTOR Specialty Pharmacy  
(800) 323-6832  •  www.NuFACTOR.com

*2015 NuFACTOR is the specialty pharmacy subsidiary of FFF Enterprises, the nation’s most trusted distributor of plasma products, vaccines and other biopharmaceuticals.
Take Control of your flu vaccine supply

with MyFluVaccine.com easy online ordering

YOU PICK THE DELIVERY DATE » Conveniently secure YOUR best delivery date(s)
YOU PICK THE QUANTITY » Choose from a broad portfolio of products
WE SAFELY DELIVER » Count on FFF’s secure supply channel with Guaranteed Channel Integrity™

TO ORDER | MyFluVaccine.com | 800.843.7477

©2016 FFF Enterprises, Inc. FL337 Rev 0615