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THE TERM “patient-centered care” (PCC) was coined by Planetree nearly 40 years ago, and it is endorsed by the Institute of Medicine and emphasized in the Affordable Care Act. PCC, defined by Planetree, is an organization that pioneered its modern movement as “an approach to care that is 1) organized around the needs of the patient; and 2) promotes relationships between patients, their families and their healthcare teams that nurture trust, transparency, collaboration and individualized care.” According to an article in NEJM Catalyst, with PCC, “patients are partners with their healthcare providers, and providers treat patients not only from a clinical perspective, but also from an emotional, mental, spiritual, social and financial perspective.” But, is PCC reality or fairy tale in today’s medical world? For most, it would seem the latter. Healthcare professionals are so burdened by an overwhelming number of patients and escalating pressures to comply with mandated paperwork and justify medical procedures, it’s difficult for them to take the time necessary to address the physical and psychological aspects of each patient.

Fortunately, for many chronically ill patients, PCC may be closer to reality. Linda K. Barry, MD, MPH, FACS, associate professor of surgery at UConn School of Medicine and author of Barriers to Patient Centered Care: My Perspective as a Physician, Patient, Caregiver and Advocate, says chronically ill patients may stand to benefit more from PCC since “the long-term nature of these diseases affords the opportunity to develop relationships with physicians and medical teams.” Primary immunodeficiency disease patient Whitney Ward sheds light on this reality in her article “What Patients Want Their Doctors to Know” (p.18). Whitney highlights her own experiences with her physicians to explain that if doctors just understood three things patients want, they could make all the difference in improving their patients’ quality of life.

Nursing, of course, is always patient-centered since nurses have to ensure patients’ needs are the primary consideration. But, boundaries can sometimes be crossed, turning healthy relationships between nurses and their patients into unhealthy ones. Rachel Colletta, a certified registered infusion nurse and director of educational resources at the National Immunoglobulin Society, explains how boundaries can become fuzzy in her article “Blurred Lines: Professional Boundaries in the Nurse/Patient Relationship” (p.21). Rachel provides examples of how and why boundary crossings may occur. Her goal is to educate both patients and nurses about how they can benefit from healthy relationships without overstepping the boundaries.

As always, we hope you enjoy these articles, as well as the many more educational and insightful topics presented in this issue of IG Living.

Ronale Tucker Rhodes, MS
Finding the Balance in Pain Management

By Abbie Cornett

IN RECENT YEARS, most states have enacted laws restricting access to opioids in response to epidemic levels of overdoses and deaths resulting from addiction. As a result, because each state’s laws outlining the prescribing of pain medications differ, patient access to medication can vary from state to state.

A major issue that needs consideration when imposing restrictions on opioids is that not all opioids are the same. There are illegal opioids such as heroin and counterfeit fentanyl. And, there are legal opioids such as oxycodone (Oxycontin), hydrocodone (Vicodin) and fentanyl that are prescribed to treat severe pain caused by surgery, acute injuries (e.g., broken bones), cancers or long-term pain conditions related to chronic medical conditions.¹

While everyone can agree the issue of illegal opioids and addiction needs to be addressed, it’s important to ensure people who actually need opioids to relieve their pain still have access to them. Unfortunately, many do not. Patient access to needed medications has been adversely affected due to newly enacted laws.

Because I work with patients who suffer from chronic conditions, I had only considered the impact of restrictions on opioid access for this population. I had not considered the implications of the new laws on people who suffer acute injuries or undergo surgery. This eye-opener became personal when I experienced an unexpected back injury. While at the emergency room, I had to fill out a questionnaire on my use of pain medication. I was a little surprised because I rarely ever take Tylenol. But, the real surprise came when I was questioned about how many times I had been in an emergency room in the last year for pain (zero), and how many times I had taken pain medication in the last year (also zero). After it was determined I had actually injured my back and was not merely seeking drugs, I was given a short supply of medications and referred to a specialist.

This is when access problems to acute pain medication became apparent. The supply of medication from my ER visit was for five days. But, there was no way I could get in to see the specialist within that time frame. Thankfully, I was able to control my pain with over-the-counter medications. However, the experience made me realize reasonable access to pain medication isn’t just an issue for patients with chronic pain conditions but also for those in acute pain.

The new restrictions on opioids were put in place as a result of studies that showed the amount of opioid medications prescribed for acute pain often exceeds the amount actually consumed by patients, and by reducing the amount prescribed for acute pain, the number of unused opioids available for diversion (illegal use) and misuse would likely decrease.²

The problem, though, is opioid prescribing for acute pain is not a one-size-fits-all situation. Many patients require no opioids, whereas others may require more than three days of treatment, particularly if they have contraindications to nonopioid analgesics or lack access to timely follow-up care outside the emergency room.³ Thus, the unintended consequence of the new restrictions is many people in acute pain are not getting their pain needs met.

While no one can argue policymakers need to address the issue, real-world experience tells us that not all people who use pain medications have addiction issues. The majority who report misusing prescription opioids did not get them from a doctor while under medical supervision, and as many as 70 percent reported prior use of substances such as cocaine and methamphetamines.⁴

People who have chronic illnesses, cancer or acute pain need reasonable access to medication. This means finding a balance between setting prescribing limits and meeting patient needs.

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

References

Do diseases have a smell?

Like other respondents, my dog has recently changed his behavior. He’s gone from being aloof to being pasted to my side and wanting to climb in my lap whenever possible (all 55 pounds of him). I thought this change was just him feeling settled after living with us for two years, but now I’m wondering if he, too, has a sensitive nose. Either way, pooch therapy is helping me feel better despite the symptoms of chronic inflammatory demyelinating polyneuropathy.

— Min J

I have stage 3 kidney disease. I have a Jack Russell, and she knows before I do when my levels are high. She sniffs my stomach and will not leave my side. She is not a service dog. They do know.

— Paula B

Do doctors need to talk to one another?

I make sure all my lab work, diagnostics, etc., go to all my doctors, as well as my own files. As patients or a care provider for a primary immunodeficiency disease patient, it is important that all knowledge is shared.

— Birgit C

[Doctor communication is] a great concept, but I have found it doesn’t always work. I have been in the hospital twice in the last few years. Both times, I asked the doctor multiple times on multiple days to please call my rheumatologist. Both times, I ended up [asking my rheumatologist to] call the hospital.

— Liz D

How do you balance life, health and finances?

I don’t really feel I have much of a life because I’m so wiped out and exhausted all the time. Between having polycythemia and doing subcutaneous immune globulin infusions every week, it’s hard to get functional the rest of the week. [It’s hard to do] anything other than hang around on Facebook or watch TV and do surveys for gift cards lately. I have no motivation to really do much exercise.

— Rachel D

[It’s] impossible to balance finances when you have a chronic illness! You do without a lot that most people take for granted!

— Carolyn W

What finances? I’m lucky if I can pay my bills at the end of the month. I haven’t worked since 2007 due to disability and all my illnesses. It’s a good thing I had a lump sum from Social Security in 2013. That’s [the] only thing that [is] saving me from losing my house and everything else I own.

— Rachel D
Abbie » I spoke with Terry O. Harville, MD, PhD, medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, and he stated IVIG and a C1 esterase inhibitor should not result in a rise in ferritin. He feels the most likely cause is your liver is being affected by the autoimmune disorders. The rise may be an indicator the disease is not fully controlled or other medications are affecting the liver. He suggests you ask your treating physician to test more for autoimmunity.

Question » Can IVIG or a C1 esterase inhibitor cause ferritin levels to rise?

My doctor has tentatively diagnosed me with necrotizing autoimmune myopathy, polymyositis with unspecified organ involvement and hereditary angioedema (HAE) type 1. My ferritin (iron) level has been climbing and is now in the low 900s. No cancers have been detected. I have been treated with intravenous immune globulin (IVIG) every four weeks for the past six months. I am also being treated with a C1 esterase inhibitor twice a week for HAE. Could either of these drugs cause ferritin to rise? I don’t have anemia, and I have been tested for hemochromatosis. I have a low total iron-binding capacity, but my iron levels are normal.

Abbie » According to Dr. Harville, pretreatment with Solu-Medrol is not required unless a person has previously had a reaction to IVIG. If you have not, there is no need for routine steroid pretreatment. In addition, non-Hodgkin’s lymphoma should not matter when deciding whether to treat CVID with SCIG. Like any therapy, SCIG treatment will have to be justified to be approved by Medicare, but that should not be a problem.

Question » Does a patient with CVID and non-Hodgkin’s lymphoma need steroid treatment prior to IVIG treatments, and would SCIG be an alternative to IVIG?

I am a 72-year-old male diagnosed with common variable immunodeficiency (CVID) that has been treated with monthly intravenous immune globulin (IVIG) infusions for the past two and a half years. I also have non-Hodgkin’s lymphoma, which is currently in remission for the second time in 13 years.

My oncologist insists on giving me Solu-Medrol (steroids) before the IVIG infusions. Because of many treatments, the skin on my arms and hands is extremely thin, and I bruise very easily, making it appear as though I am prescribed blood thinners, but I am not. I also have trouble falling asleep and staying asleep for two nights after treatment. Are steroids necessary when getting IVIG infusions? If not, are there any side effects?

I have heard about subcutaneous IG (SCIG), but I am unsure if I could do it myself. I asked my oncologist about this option, but he seems to think not enough research has been conducted on IVIG patients with non-Hodgkin’s lymphoma. On the other hand, my allergist, the doctor who discovered I needed IVIG treatment, thinks differently. I would like your professional opinion. I would prefer to have something other than the four- to five-hour monthly infusions. Do you know if Medicare will pay for SCIG treatments at home?

Abbie » According to Dr. Harville, pretreatment with Solu-Medrol is not required unless a person has previously had a reaction to IVIG. If you have not, there is no need for routine steroid pretreatment. In addition, non-Hodgkin’s lymphoma should not matter when deciding whether to treat CVID with SCIG. Like any therapy, SCIG treatment will have to be justified to be approved by Medicare, but that should not be a problem.

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

ABBIE CORNETT is the patient advocate for IG Living magazine.
Type I Hypersensitivity: ‘True’ Allergic Disease, Part 2

By Terry O. Harville, MD, PhD

IN THE LAST issue, we began the discussion of true allergic disease, or type I hypersensitivity, mediated by IgE interaction with mast cells. True allergic disease is what people generally think of as allergies, which are responsible for hay fever (a reaction to pollen grains from trees, grasses and weeds that occurs in seasonal patterns) causing the symptoms of runny nose, itchy eyes, etc., and also occurs due to indoor allergens such as dust mite dander. IgE-mediated allergic disease is also associated with food allergies such as cow’s milk, soy, wheat, fish, shellfish and nuts, particularly peanuts. Life-threatening anaphylaxis may occur after exposure to a food item to which one is allergic due to excessive mast cell degranulation. Consequently, food allergies are a serious issue. (Many other items may cause allergic reactions such as mold spores, but this discussion pertains only to the more common items.)

As previously noted, this form of immunity developed in humans to protect us from parasites. IgE antibodies directed to a parasite and binding to mast cells results in the release of noxious substances to make the parasite want to leave our bodies. Thus, in early times when humans were less sanitary during evolution, this action was an important protective immunity. As humans began to be more sanitary, this part of our immune system began to react to harmless environmental stimuli such as pollen grains. Noteworthy studies of German communities support this theory.

After the fall of the Iron Curtain and Berlin Wall that reunified Germany, studies evaluated the former East Germans compared to the West Germans because allergic disease was uncommon in the former, but common in the latter, even though these people lived in relative close proximity separated only by the Berlin Wall. As the general environment was similar for all Germans, all were exposed to similar pollen allergens in the air. But, within a decade or so of reunification, the extent of allergic disease was similar between the former East Germans and West Germans. However, studies indicated there were some specific differences present before the fall of the Iron Curtain and Berlin Wall, most likely microbial exposures, providing further support of the so-called “hygiene hypothesis.” For example, East German milk may not have been pasteurized before consumption, and there was greater consumption of fermented foods (the type of food preservation and diet before modern refrigeration and processed foods). As a result, East German babies were exposed to the immunologically-expected microbes and parasites from very early in life; hence, their immune systems developed in the normal evolutionary response, which helped to preclude allergies. On the other hand, West German babies were fed processed sterile foods and, thus, did not have the immunologically-expected microbial/parasitic exposures to their immune systems. In their case, the Th2 immunity that looked for parasites to protect us responded instead to other harmless environmental items, thus creating allergies.

It is believed the body or immune system recognizes a parasite is present when it perceives it is being eaten.

To sum up, the common allergens that result in allergic disease appear to have in common with parasites a “digestive enzyme signal” that triggers the Th2 response (protective against parasites) and results in allergic disease manifested by allergens. We will continue with the topic of hypersensitivity and allergic disease in the next issue.

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.
The Dos and Don’ts of Supporting a Friend or Family Member with a Chronic Illness

By Erika Lawrence, PhD, LCP

Loving Someone with a chronic illness can be confusing. One day, chronic illness sufferers seem fine, and the next day, they can’t get out of bed. Symptoms and functioning change day to day, week to week, even hour to hour. They may cancel plans often or not want to make plans with you at all for fear of needing to cancel at the last minute. You want to be supportive, but you don’t know how. Or, you try to be helpful, but your efforts fall flat. It can be frustrating, confusing and challenging. So what do you do? Well, every person is different, and the trajectory of every chronic illness is different, but there are some rules of thumb you can use to get started on the right path. I like to call them the “dos” and “don’ts” of loving someone with a chronic illness.

What Not to Do

Don’t blame them for their symptoms. You may find yourself thinking, “You were fine yesterday” or “You look perfectly fine.” No one can tell what is going on inside someone’s body by looking at the person from the outside. They are not being lazy or stubborn. They can’t help how their body is responding on a given day.

Don’t compare their experience to someone else’s. Chronic illnesses and, in particular, immune disorders are highly unpredictable and variable. The symptoms, pain and impact on functioning can change on a dime. Moreover, the course of these disorders over time is different for every person. Trying to compare your friend or loved one to another person with a different diagnosis, or even to someone with the same diagnosis, is rarely helpful.

Don’t give unsolicited advice. You love this person. You want to help. You want to find answers that make things better. So, you read or consult with others or go online to learn more. Then, you share what you found. Or, perhaps you simply offer general practical advice for healthy living.

Unfortunately, offering unsolicited advice is the most harmful form of help in a close relationship. Do not offer advice unless it is specifically requested. For one thing, people know what they need to do. Additionally, offering unwanted advice can make your loved one feel guilty or resentful, putting a strain on your relationship.

What to Do

Believe. Individuals with a chronic illness spend so much time trying to convince doctors and others that their symptoms are real. Take what your loved one is saying at face value. Their symptoms or pain or inability to do something on a given day is real. It may not make sense to you. It may not be what you expected. Believe them anyway and respond to what they are telling you rather than what you think is going on.

Be empathic. Your loved one’s struggle is hard enough. Listen, sympathize, be compassionate. Let them know you care even though you can’t change the situation. This can be the most powerful thing you do for them.

Ask what kind of support or help they would like. Some people want to vent and simply be listened to. Others want to be hugged. Others need help with daily chores such as household tasks, grocery shopping, making dinner, picking up the kids, etc. Still, others need a pep talk when their spirits are low.

What you would want in this situation is probably different than what your loved one wants in this situation. And, they may want different types of support on different days. Instead of guessing what they want, or doing what you would want in that situation, ask your loved one what you can do for them. Moreover, offer some specific options (like the ones listed here), so the burden is not on them to come up with something you can do.

Follow the Simple Rules

Supporting and helping your loved one can be challenging, but following these simple rules may make all the difference in the world to your loved one and to your relationship with them.

ERIKA LAWRENCE, PhD, LCP, is director of translational science at The Family Institute at Northwestern University, Evanston, Ill.

Reference

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Update on Subcutaneous Immune Globulin

By Michelle Greer, RN

**IMMUNE GLOBULIN** (IG) products have evolved tremendously over the years. Products were once approved by the U.S. Food and Drug Administration (FDA) only for intramuscular or intravenous use. Initially, these products were manufactured in a lyophilized form that required reconstitution for infusions. The next generation of intravenous IG (IVIG) products saw improvements with availability in ready-to-use liquid forms; however, they were low concentration (5%) requiring high-volume infusions. Following the introduction of 5% liquid products, manufacturers continued to refine the manufacturing process and developed 10% ready-to-use liquid products. Most recently, manufacturers developed products for the subcutaneous route of administration. These products are available in several concentrations (10%, 16.5% and 20%), and all are ready-to-use liquid formulations. Subcutaneous IG (SCIG) products give additional options to people who require IG treatment, allowing them to find a product they can best tolerate.

SCIG products were initially manufactured and tested to treat only primary immunodeficiency disease (PI) patients. PI patients typically receive relatively low doses of SCIG that are most commonly infused weekly, which allows for the total monthly volume of an IVIG dose to be divided into smaller-volume weekly doses. These divided doses allow for infusion into the subcutaneous tissue in a fairly short time — between one and two hours. More recently, SCIG products received approval by FDA to treat chronic inflammatory demyelinating polyneuropathy (CIDP), which is an autoimmune disease. There are also reports in the medical literature that show the efficacy of using SCIG products to treat other autoimmune diseases.

**SCIG Products**

SCIG products have been available in other countries for many years, but it wasn’t until 2006 when the first SCIG product, Vivaglobin (CSL Behring), was approved by FDA for use in the U.S. At that time, while most IVIG product concentrations were 5% or 10%, Vivaglobin was 16%. With a higher solution concentration, more drug can be infused in fewer subcutaneous insertion sites and/or shorter infusion times.

Shortly thereafter, Gamunex-C and Gammagard Liquid, both 10% products with prior FDA approval for IV administration, received FDA approval for SC administration in 2010 and 2011, respectively.

Hizentra (CSL Behring) was the first 20% subcutaneous solution to enter the market in 2010, one year prior to the discontinuation of Vivaglobin from the market. Hizentra was also the first and remains the only SCIG product to be approved by FDA for a condition other than PI. Hizentra was approved in 2018 to treat CIDP.

Following Hizentra, HYQVIA (Takeda), a facilitated SCIG (fSCIG) product, was approved by FDA in 2014. HYQVIA is a combination packaged product that includes a 10% IG component with hyaluronidase. Hyaluronidase, which temporarily opens up the subcutaneous space to allow for more volume to be administered in a single site, is administered in the subcutaneous space prior to the infusion of IG. Since the addition of hyaluronidase allows a much higher volume of the product to be infused, most HYQVIA fSCIG infusions require just one or two injection sites and only monthly infusions. In contrast, traditional SCIG products vary in the number of injection sites, length of infusion and frequency of infusion.

**SCIG Products**

<table>
<thead>
<tr>
<th>Product</th>
<th>Manufacturer</th>
<th>Administration Route</th>
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<tbody>
<tr>
<td>Vivaglobin 16% (discontinued)</td>
<td>CSL Behring</td>
<td>SC</td>
</tr>
<tr>
<td>*Gamunex-C 10%</td>
<td>Grifols</td>
<td>IV and SC</td>
</tr>
<tr>
<td>*Gammaked 10%</td>
<td>Kedrion</td>
<td>IV and SC</td>
</tr>
<tr>
<td>Gammagard Liquid 10%</td>
<td>Takeda</td>
<td>IV and SC</td>
</tr>
<tr>
<td>Hizentra 20%</td>
<td>CSL Behring</td>
<td>SC</td>
</tr>
<tr>
<td>Cuvitru 20%</td>
<td>Takeda</td>
<td>SC</td>
</tr>
<tr>
<td>Cutaquig 16.5%</td>
<td>Octapharma</td>
<td>SC</td>
</tr>
<tr>
<td>Xembify 20%</td>
<td>Grifols</td>
<td>SC</td>
</tr>
<tr>
<td>HYQVIA 10% + Hyaluronidase</td>
<td>Takeda</td>
<td>fSC (facilitated subcutaneous)</td>
</tr>
</tbody>
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*same product
depending on the diagnosis and dose, as well as the patient’s subcutaneous tissue.

Additional higher concentration products have been approved by FDA over the last several years, starting with Cuvitru 20% (Takeda) in 2016, Cutaquig 16.5% (Octapharma) in 2018 and Xembify 20% (Grifols) in 2019.

SCIG Benefits

Determining the treatment choice between IVIG and SCIG infusions should be up to patients and their prescribing physicians. While each route of administration has its benefits, the predominant factors patients should consider are lifestyle and tolerability.

Lifestyle. IVIG is typically infused and monitored by a trained registered nurse. SCIG infusions, on the other hand, are self-administered by patients, allowing for more independence and flexibility. For instance, patients can infuse on any day at any time and even when they travel, but they should try to infuse at regular intervals.

Tolerability. While most people tolerate IVIG with minimal to no side effects, some experience reactions that can be severe and debilitating. This is because infusions directly into a vein are more likely to cause systemic reactions due to the immediate administration into the bloodstream. With SCIG, the drug is absorbed more slowly and, therefore, the potential for systemic reactions is greatly reduced. SCIG is also a great option for people who may have difficulty with venous access. For those who have venous access issues and prefer IVIG, a port may be an option, but consideration should be given for the increased potential for clotting and infection.

SCIG Similarities and Differences

Since not all SCIG products are the same, their similarities and differences should be taken into account when selecting a brand.

As stated earlier, all SCIG products are FDA-approved to treat PI, and Hizentra is approved to treat both PI and CIDP. FDA approval is based on clinical trials that showed efficacy within specific dose ranges. Each product has specific dosing protocols, infusion volume per site and maximum rates of infusion per site. Dosing and administration protocols are different for PI than CIDP.

Sugar content, IgA content, sodium content, pH and osmolality of products should also be considered. Unlike IVIG, no SCIG products are stabilized with sugar. However, IgA content varies. IgA is usually only a consideration for people who are severely deficient in IgA and have anti-IgA antibodies, which is very rare. Sodium content also varies and should be considered.

Osmolality of IVIG/SCIG and SCIG products varies and is usually only of concern with IVIG infusions. Most products are within the range of physiologic osmolality. Products that deviate substantially from physiologic osmolality levels may put patients at risk for various infusion-related adverse effects. Similarly, these same patients may be sensitive to the sodium content of SCIG products. If the pH of an injectable product is substantially below physiologic levels, localized reactions at the site of injection may result.

With SCIG, site reactions are the most common side effect. Important factors to consider to reduce local side effects are the supplies used to administer the product and the insertion technique. Determining the correct needle length is important to ensure SCIG is delivered to the subcutaneous space. A needle that is too short or too long may cause the patient to experience pain or welts. Patients should be taught where and how to place the needles to avoid local site reactions as well. Good insertion technique with the right size needle helps reduce local site reactions. While site reactions can range in severity, they typically dissipate over a day or two post-infusion, and diminish as patients remain on therapy.

Lastly, payers may have a product formulary for SCIG that may limit choice in product selection. And, it should be noted that some of the newer SCIG products are awaiting approval for reimbursement by Medicare B in the home setting at the time of this writing.

Product Selection Is Important

Since IVIG and SCIG products can experience shortages, it’s beneficial for patients to have a range of products from which to choose. However, in the same way IVIG products vary, SCIG products also differ from one another. Therefore, all product attributes must be considered when selecting a brand for patient treatment, and product choices should be reassessed on an ongoing basis.

MICHELLE GREER, RN, is senior vice president of sales for Nufactor, a Specialty Infusion Company.

Reference

Because common variable immunodeficiency (CVID) is a diagnosis of exclusion, making it difficult to establish a definite diagnosis, researchers developed a machine learning pipeline that performs automated diagnosis based on flow cytometric immunophenotyping. Using the pipeline, the researchers analyzed the immunophenotypic profile in a pediatric and adult cohort of 28 patients with CVID, 23 patients with idiopathic primary hypogammaglobulinemia, 21 patients with IgG subclass deficiency, six patients with isolated IgA deficiency, one patient with isolated IgM deficiency and 100 unrelated healthy controls. Compared to manual analysis, which is traditionally performed and has severe limitations, the automated pipeline resulted in a more reproducible flow cytometry analysis and improved diagnosis, with the pipeline achieving on average a balanced accuracy score of 0.83 compared to a balanced accuracy score of 0.72 with the manual analysis. The researchers believe this will result in more timely diagnoses that is essential for optimal follow-up and treatment.


Hizentra (Immune Globulin Subcutaneous [Human] 20% Liquid) Receives Orphan Drug Exclusivity as Maintenance Therapy for CIDP

CSL Behring’s Hizentra (subcutaneous immune globulin [SCIG] [human] 20% liquid) has received orphan drug exclusivity from the U.S. Food and Drug Administration (FDA) for the treatment of adult patients with chronic inflammatory demyelinating polyneuropathy (CIDP) as maintenance therapy to prevent relapse of neuromuscular disability and impairment. Hizentra was previously approved by FDA in March 2018 for the treatment of adults with CIDP to prevent relapse of neuromuscular disability and impairment. Orphan drug status provides CSL Behring a seven-year period of U.S. marketing exclusivity for Hizentra in the maintenance and treatment of CIDP with SCIG.

Approval was based on data from the Phase III PATH (Polyneuropathy And Treatment with Hizentra) study, the largest controlled clinical study in CIDP patients to date. In the PATH trial, patients taking Hizentra relapsed or withdrew less often than those taking placebo. Patients in the study also maintained their grip strength, as well as upper- and lower-body strength.

“Orphan drug exclusivity is a significant milestone for the CSL Behring team committed to delivering Hizentra and improving the lives of patients diagnosed with CIDP,” said Bob Lojewski, senior vice president and general manager, North America, at CSL Behring. “We are proud to be the only company to offer an innovative portfolio of subcutaneous and intravenous immunoglobulin therapies for CIDP.”

Research

Stewardship Program Improves IVIG Prescribing

A multidisciplinary team of clinical pharmacists, physicians, nurses, informaticians and IT specialists developed a stewardship program to improve and standardize intravenous immune globulin (IVIG) dosing in their healthcare organization, and found institution-specific ordering patterns may facilitate more efficiency and effective prescribing and optimized drug dosing.

The project involved a four-step approach:

• Automating dose rounding to commercial vial strength to facilitate optimized dispensing practice;

• Generating a complete list of indications for IVIG from internal data capture;

• Developing and implementing an order set with coded indications and dosing, including dose adjustment for indications and patients’ weight; and

• Evaluation and ongoing tool optimization.

The first step was developed after a workflow analysis found prescribing practices and pharmacy preparation processes were contributing to drug waste. The processes included pooling IVIG from individual vials into a large glass bottle to indicate the precise grams prescribed. However, dose rounding to the commercially available increments (such as rounding a 32-gram dose to 30 grams) allowed for dispensation of the entire product vial, which enabled the dispensing practice to change from pooling to commercial bottle dispensing. And, it prevented the waste of unused vials that had expiration dates much longer than 24 hours if stored appropriately.

The second step included ascertaining IVIG usage patterns. Using the indication and dose entered by providers in combination with patient demographics, the researchers conducted an evaluation on the usage and dosing ranges of IVIG. After an 18-month period, a literature search was conducted for supportive scientific evidence and recommended dosing information for the most frequently prescribed clinical indications.

In the third step, clinical experts in each therapeutic area convened over six months to discuss appropriateness of IVIG use and dosing for each indication. Specific recommendations were aggregated into an IVIG order set using contextualization, in which the order set was adapted according to the medical condition of the patient as the basis of the next step in the approach. The new order set consisted of four main sections. The first section included two drop-down menu options for the therapeutic category and specific indications and a free-text field for indications not found among the provided options. In the second section, the prescriber was prompted to answer whether a patient had a known immunoglobulin A deficiency, which would select the correct IVIG product for dispensing. Prescribers were also prompted to indicate whether the patient would undergo plasmapheresis treatment, and if so, a general nursing order was generated so the nurse knew to coordinate with the pharmacy for drug dispensation to occur after plasmapheresis. And, finally, a medical logic module (MLM) was programmed with the specific details regarding the weight-based dosing for each indication, as well as defaulted frequencies and durations, so the recommended dosage was based on the provider’s answers in the first two parts of the order set. The MLM also indicated the latest recorded weight and height for the patient to determine whether actual weight or adjusted body weight should be used in the dosing calculation, and then performed the dosing corrections as necessary.

In the last step, the IVIG dose was recorded. The recommended dosage was calculated and entered, but providers could override the dosage based on their clinical judgment. And, premedications to prevent or mitigate infusion-related reactions and preset infusion rates were generated in the order and comment fields to optimize the safe administration of IVIG.

During 36 months before the implementation of the IVIG order set, out of 1,965 IVIG orders reviewed, the prescribed IVIG dose varied considerably from the expected dose (mean = −1.8, range = −4.9–1.5). However, in the 27 months after order set implementation, out of 848 IVIG orders reviewed, the prescribed IVIG dose was closer to the expected dose (mean = −1.2, range = −3.9–2.6).

Conference

13th Annual Neuropathy Action Awareness Day Is June 19

The 13th Annual Neuropathy Action Awareness Day will be held Friday, June 19, at the InterContinental Hotel in Los Angeles. This largest neuropathy awareness and education event in the U.S. provides an opportunity for patients to interact with other patients, providers and exhibitors, as well as to learn about neuropathy and how to cope with the disease, policy issues and patient advocacy. The day begins with an exhibit area and educational sessions in the morning, followed by a luncheon and additional educational sessions and exhibit area with refreshments in the afternoon. Additional features include a celebrity speaker, elected officials and a silent auction that includes trips, activities and other fun items.

To ensure as many patients can participate as possible, the Neuropathy Action Foundation (NAF) has booked a block of rooms at the hotel for a special, reduced rate. For those with a financial hardship, NAF will pay for up to 10 flights and hotel rooms for the night of the event or the night before the event for patients from outside the Los Angeles area. In addition, those unable to attend in person can listen free of charge via Livestream technology using a computer with an Internet connection. Those individuals will also be able to ask speakers questions and receive answers in real time. The event will be recorded, so individuals can watch the event even after it has ended.

To register, go to the NAF website (neuropathyaction.org) and click on “register today” on the right side of the homepage. Once registered, those watching via Livestream will receive a link to view the event. Registration is $25 for patients and caretakers for the event and luncheon. For nonpatients and noncaretakers, the event is $125 per person. The event is free for Livestream participants.

Medicines

FDA Approves Benlysta to Treat Children 5 Years and Older with Lupus

GlaxoSmithKline’s Benlysta (belimumab), a B lymphocyte stimulator (BlyS)-specific inhibitor, has been approved by the U.S. Food and Drug Administration (FDA) to treat children 5 years and older with active, autoantibody-positive systemic lupus erythematosus (SLE) who are receiving standard therapy. This approval extends the current indication in the U.S. approved by FDA in March 2011 for the intravenous formulation of Benlysta in adults with SLE.

Approval was based on data from a post-approval commitment study (the “PLUTO” study) that assessed the efficacy, safety and pharmacokinetics of 10 mg/kg intravenous belimumab plus standard therapy compared with placebo plus standard therapy for one year in children ages 5 years to 11 years and 12 years to 17 years with SLE. Since pediatric lupus is an uncommon disease, a fully powered study was not feasible. The proportion of children achieving a clinically meaningful improvement in disease activity, as assessed by the SLE responder index response rate, was numerically higher in patients receiving belimumab plus standard therapy (52.8 percent) compared with placebo plus standard therapy (43.6 percent) at week 52. The proportion of patients experiencing more than one adverse event (AE) and a serious AE was 79.2 percent and 17.0 percent for the belimumab group compared with 82.5 percent and 35 percent for the placebo group, respectively. AEs that led to discontinuation were lupus nephritis, hepatitis A, hypertransaminasemia, acute pancreatitis, post herpetic neuralgia, retinal vasculitis and pancreatitis.

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A recent study has found children with Kawasaki disease (KD) make antibodies to hepacivirus peptides, and KD ICI contain protein with a hepacivirus-like epitope, which strongly suggests a new human virus, closely related to the hepaciviruses and with a respiratory portal of entry, is etiologically related to KD.

In the study, researchers sequenced 1,156 peripheral blood plasmablasts (PB) from 11 KD patients, and identified 44 sets of oligoclonal PB in these patients. They then prepared 61 monoclonal antibodies (Mab) from oligoclonal PB and from IgA PB that showed high levels of somatic mutation. Ten of these antibodies strongly bind to KD ICI, and 23 weakly bind. Animal virus peptide array revealed that Mab KD4-2H4 (from patient KD4), which strongly binds ICI, recognized multiple similar peptides from a nonstructural protein of hepacivirus C with an identified motif that was highly significant. Patient KD4 had negative hepatitis C serology. Peptide substitution analysis was performed to identify optimal amino acids for binding of KD4-2H4 at each position. ELISA using an optimized peptide revealed that four other KD Mab from two additional KD patients also recognized this peptide; all three patients had coronary aneurysms. The strong ICI binding of KD Mabs KD4-2H4 and KD6-2B2 was completely blocked by pre-incubation with the optimized peptide.

The researchers believe identification of the specific etiology of KD could revolutionize diagnosis and treatment of the disease in the future.
What Patients Want Their Doctors to Know

Patients’ quality of life can be positively impacted when doctors are mindful of three things about patient-doctor interaction.

By Whitney L. Ward

ONE OF THE most basic human needs is the desire for human relationships. But, relationships can be difficult. To make them work, there has to be the perfect blended recipe of a dash of giving and a pinch of taking. Yet, despite the blood, sweat and tears, without special people in our lives, we would feel a void.

For those of us with a chronic illness such as a primary immunodeficiency disease (PI), doctors are included in the list of special people. We PI patients may not have asked for these men and women with white coats to play a significant role in our lives, but for better or worse, we need them. Our immune-suppressed bodies depend on specialists, which can be frustrating, especially when it seems we are just a number or a medical case being shuffled through a revolving door of other medical cases. However, mutual admiration and respect between doctors and patients is possible when doctors care about their patients’ concerns, show compassion about any fears they may have and understand the three things patients want their doctors to know about patient-doctor interaction.
1) Patients want a relationship with their doctors that has depth.

There are two types of doctors. There are those who have conversations with their patients about life and what exciting things they are involved in. And, there are those who just get down to brass tacks: “This is how you’re doing, and this is what needs to be changed in your patient care; see you in six months.” It’s clear to me which doctor/patient relationship has a stronger connection. The National Institutes of Health estimates 25 million to 30 million Americans are living with a rare disease, and the National Health Council estimates 133 million Americans have a chronic illness. These Americans are a blended community of men, women and children who need compassion and assurance during scary unknowns and uncertain futures. While there are doctor/patient lines that can’t be crossed, when doctors want to invest in their patients and get to know who they are aside from their medical cases, it eases the patients’ fears and allows them to have another level of trust in the care they are receiving.

I was 6 years old when my PI began to significantly affect my health. It was a new and frightening experience, but the relationship I had with my rheumatologist helped with my anxiety. At every visit, he would sit me on the examining table, look at my mom and say, “OK, mom, you just sit over there and stay quiet. Whitney and I need to chat for a few minutes.” And, that’s what we did. We talked about school, shoes, clothes, how my sister was doing — how life was outside of my disease — and because of this, I was comfortable and ready when it came to discuss my health.

Doctors who take the time to interact on a personal level with their patients make a difference in their patients’ lives. When a milestone happens in life, these doctors are on the list of people patients want to share it with, because they celebrate with them. The doctors who make the biggest impact are the ones who not only care about their patients’ medical lives, but also their quality of life.

2) Patients don’t want their dreams belittled or diminished.

When patients tell their doctors they are going to attempt to do something that may risk their health, the doctors’ initial response may be to diminish or belittle their patients’ dreams. Patients need to understand their doctors care about them and their well-being, so their response stems from fear. Doctors are human, too, and not only do they remember when their patients were at their worst physically, they also have the medical knowledge of what could happen to their patients’ health if they take risks. That being said, doctors have to remember their patients know when their bodies are weak or strong. They know when they can push their bodies and when their bodies have said, “That’s enough!” The millions of people living in this country with a disease can make a difference and impact this world if they are encouraged to not allow their disease define their abilities and ambitions. If patients aren’t allowed to try, they will never know what limitations they can surpass. For those with a chronic illness, that type of fulfillment and purpose is important.

My immunology nurse practitioner once told me with a chuckle: “Whitney, your doctor and I don’t want you to live in a bubble, but some of the things you attempt to do scare us to death!” I can’t tell you how happy her comment made me, because it showed me I’m truly defining my disease. And, even though it made my doctor nervous, he was beginning to understand how important it was to allow me to dream.

3) Patients don’t want their doctors, in their excitement about a teaching moment, to treat them like a human petri dish.

At one time or another, most PI patients have heard one of these phrases from their specialist: “You aren’t textbook,” “We talk a lot about your disease with our colleagues because it’s so rare” or “You’re very unique.” Fact of life: Most PI patients will be “teaching moments” for doctors because, in
the medical world, we aren’t “horses,” we’re “zebras.” All PI patients understand this.

However, when specialists bring in another colleague, resident or fellow so they can show them their patients’ medical abnormalities, it sometimes takes a toll on these patients’ emotions and self-esteem. All patients know the routine: The doctor comes in the room, tells the patient he or she looks good, and then asks if the resident or fellow can look at the patient’s deformities. When this happens, patients often wonder, “Are they trying to tell me I look ‘good’ for someone with a rare disease that produces rare abnormalities?” According to a National Health Interview Survey conducted by the Centers for Disease Control and Prevention in 2018, 68.2 percent of Americans see a doctor every six months or less. Who wants to be reminded frequently that the embarrassing blemishes or noticeable deformities aren’t “normal”?

I have a specialist who has been treating me for almost 17 years. We have a great relationship, and I know he truly cares about my well-being. But, when I first began seeing him, tactfulness wasn’t his strong point. Nearly every appointment, he would ask to see the warts on my feet that my immune-compromised body produced. When I agreed to his request, he would back away from me, standing as far from me as he possibly could with his hands behind his back, craning his neck to get a good look at my “abnormal feet.” One day when I was in my late teens, he asked, “Whitney, can I see your feet?” Of course, I obliged, but on this day, I was quite fed up with his insistence of observing my “scientifically abnormal feet.” I turned my back to him to take my shoes and socks off, and for the first time in our doctor/patient relationship, he was standing right beside me, but I didn’t know. I took the first shoe and sock off without a hitch. I then took the second shoe off, but then had trouble with my sock. It was stuck. I pulled and pulled, and then finally yanked it off as hard as I could. The sock flew behind me and hit my doctor right in the face. I giggled, my doctor was shocked and offended, and my mother looked like she was about ready to clock me. Today, my immunologist and I look back on the incident and laugh. But, that awkward incident was a breakthrough that made my specialist realize he needed to treat me with more compassion and not as a science experiment.

Doctors aren’t intentionally malicious, but they are scientists. And, in their excitement to potentially learn something from their patients’ condition, they sometimes forget their patients are insecure and self-conscious about their medical deformities, so their doctors’ teaching moments make them feel like a medical oddity on display. Patients understand teaching moments are essential for scientists to have medical breakthroughs so more people can be helped. However, doctors need to remember that during those teaching moments, they should go the extra mile to make patients feel comfortable, understanding the vulnerabilities patients might experience when being exposed in such a way.

Seeing the Whole Person, Not Just the Case

PI patients and others with chronic illnesses rely on their doctors to help improve their quality of life. This relationship is not a preference; it’s a necessity. When doctors look past the medical case and truly see the patients sitting across from them, they understand that the care and treatment they provide their patients can give them the ability to live life to the fullest, filled with dignity and purpose despite their patients’ diseases.

WHITNEY L. WARD was not only the first person in the world diagnosed with MAGIS syndrome, she had the honor of naming the new primary immune deficiency. MAGIS means “more” in Latin, and Whitney hopes to instill in her readers the message they are more than their disease. Find out more about Whitney’s journey at www.whitneylaneward.com.
Blurred Lines: Professional Boundaries in the Nurse/Patient Relationship

Patients and nurses both benefit by respecting the boundaries of professional care.

By Rachel Colletta, BSN, CRNI, IgCN, VA-BC

FOR MANY YEARS, nursing has been ranked as the most widely respected and trusted profession. The term “nursing” is synonymous with words like “compassion” and “empathy.” Nurses see patients as more than a group of symptoms and disease states. They also serve as bridges between families, physicians and other healthcare providers. Nurses’ to-do lists are never-ending and ever-expanding based on the needs of patients in their care. Aside from physical care, patients often also need emotional and spiritual support. Therefore, it’s not surprising close friendships form between nurses, patients and families. But, it can be difficult to define where the role of nurse ends and friendship begins. This delineation is known as the professional boundary. Like lines in a parking lot, professional boundaries have clear lines in place to protect both patients and nurses. So, it’s essential to understand these boundaries to maintain a healthy relationship that best serves patients.

Professional Boundaries

Florence Nightingale first spoke about nursing boundaries when she said nurses will “hold in confidence matters committed to their keeping and devote themselves to the welfare of those committed to their care.” This statement referred to the responsibility of nurses to maintain the boundaries relating to patients under their care.

Fast forward to the present day and the evolution of the term professional boundaries as defined by the American Nurses Association states: “Nurses must recognize and maintain the boundaries that establish appropriate limits to relationships.” But, what does that mean? And, how do nurses and patients manage their healthcare partnerships and maintain healthy boundaries?

To successfully navigate boundaries in the nurse/patient relationship, it is essential to understand the terms used to define professional boundaries, and what they mean to...
patients and their healthcare providers.

“Professional boundaries” are defined as the spaces between nurses’ power and patients’ vulnerability. In any nurse/patient relationship, nurses are always in a position of power, which comes from the amount of detailed, sensitive information nurses know about patients compared to the information patients know about their nurses. Because this creates a very unbalanced relationship, it is the responsibility of nurses to respect this power and use it for the greater good of patients.

“Boundary crossings” are thought of as brief trips across the line. Imagine, for example, a person accidentally takes up two spaces in a parking lot. That person can move his or her car immediately, and there is no harm done. The next time that person parks, he or she will likely check the car’s position. Boundary crossings are similar; they are inadvertent and are often done as a thoughtful gesture with no harm intended. Examples of boundary crossings by nurses include:

- Divulging personal information about themselves to provide comfort or reassurance to patients;
- Accepting small gifts like cookies or cake;
- Performing tasks outside of the job description such as doing household chores or cooking;
- Bringing food or other groceries to patients; and
- Transporting patients to the mall or a physician appointment, even when they are off duty.

Boundary crossings are usually considered acceptable when they are done in the best interest of patients; however, if performed long-term, they can lead to more serious violations.

“Boundary violations” occur when there is confusion between the needs of nurses and the needs of patients. Imagine that while parking the car, the person determines it is his or her right to take up two spots. That person continues to park in two spots regularly, and it becomes a habit. A few examples of boundary violations by nurses include but are not limited to:

- Divulging detailed personal information to patients, which can include financial problems, marital issues or other personal information not related to the care of patients;
- Accepting money or a loan from patients, even in small amounts;
- Refusing to allow another healthcare professional to provide care for patients;
- Keeping secrets from other healthcare professionals involved in patients’ care;
- Developing an intimate relationship before, during or after caring for patients;
- Friending patients on social media; and
- Attending weddings, graduations or other family events.

These violations are more severe and may lead to a reversal of roles, leaving patients in a vulnerable position. Oftentimes, patients don’t realize this is happening until a serious incident occurs.

“Therapeutic touch” is a practice derived from an ancient technique called “laying on of hands.” The practice of nursing requires physical contact between nurses and patients. For hundreds of years, nurses have provided comfort by holding a hand or giving a hug. In addition to physical contact needed to provide care, many patients find the act of touch comforting. It helps them to feel they are not alone. But, while most patients appreciate the act of therapeutic touch, some do not. Family members may also be uncomfortable with this form of nursing intervention. Honesty and an open discussion early in the relationship will help to establish these boundaries.

The “zone of helpfulness” occurs when the nurse/patient relationship is in perfect balance (Figure). When thinking of a professional relationship as a straight line, the zone of helpfulness is right in the middle. It is in this area where most interactions between patients and nurses should occur. To the left or right of center are areas of underinvolvement or overinvolvement. Relationships with healthcare providers often drift in either direction, and this tool can be used to help both parties evaluate the status of the partnership and make adjustments as needed. It is not necessary to evaluate every interaction that occurs between nurses and patients. More importantly, interactions should be looked at as a whole to determine where they fit on the continuum of care.
Social Media: Friend or Foe?

The use of social media is a staple of communication in today’s society. Since the 1980s, cell phone subscriptions have grown to almost 6.8 billion. Facebook, Twitter, Snapchat and Instagram are platforms that enable people to stay connected every minute of every day to any person in the world. Yet, while these platforms provide an excellent method of communicating with friends and family members, they can also make it difficult to protect the privacy of patients. Using social media can further blur the lines of what is considered a healthy nurse/patient relationship. Boundary violations occur when nurses and patients friend each other and post comments and pictures on social media, even if it occurs during the nurses’ downtime. As innocent as it sounds, these actions can lead to violations of patient privacy and confidentiality. Healthcare professionals should abide by the policies provided by their organization regarding social media activity. By having an open discussion with their healthcare professionals, patients can prevent these sorts of violations from occurring.

**Warning, Warning**

Red-flag behaviors can help both nurses and patients evaluate and adjust the nature of the professional relationship. These behaviors include but are not limited to:

- Secretive behavior
- A feeling of possessiveness experienced by patients or nurses
- Discussions of intimate or personal issues from nurses to patients
- Speaking poorly about other nurses or an employer in the presence of patients
- Meeting with patients in a nonwork setting or after work hours
- Friending on social media

**Facing the Consequences**

Reports of boundary violations to a state’s board of nursing can lead to serious consequences for nurses. Reprimands may be administered requiring nurses to undergo re-education about professional boundaries. And, suspension or permanent revocation of professional licensure, if ordered, can impact nurses’ ability to practice for many years. More importantly, patients may be left feeling a lack of trust in the nursing profession and can experience emotional trauma, anxiety or depression. These experiences may lead to a setback in patients’ health.

Ensuring Patient-Centered Care

Nurses have duties to both care for and protect patients. While accomplishing this, it is entirely reasonable for friendships to form. It could be said this is a hazard of the job that is virtually unavoidable. Nurses use the skills they have obtained and their innate compassion for people to lead patients on a journey to wellness. It feels quite natural to do favors for patients to make this journey easier.

Nurses caring for patients in the home setting are more likely to participate in these types of activities as they form long-lasting relationships with patients with chronic illnesses. It is when emotional attachments form that boundary violations become more prevalent. Therefore, it is vital for patients to have an open, honest relationship with their healthcare providers and to discuss these issues early and often.

Nursing care is always patient-centered, and nurses have to ensure patients’ needs are the primary consideration. Professional boundaries were established to protect both patients and nurses. With education and self-awareness, both can benefit from maintaining a healthy relationship within the boundaries of the profession while providing the highest level of care all patients deserve.

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**Resources**

WHEN MEETING WITH new clients to set goals, I always ask what their top three priorities are in life. And, almost always, regardless of the medical diagnosis, my clients express the desire to stay in their own home. Yet, many have admitted they aren’t completely honest with their family and medical providers about safety and independence at home. Usually, this is because they have concerns, but they are afraid disclosing them will force a conversation about relocation.

Comments such as “Maybe home isn’t the safest place for you any longer, Mom,” “We could start looking at places this weekend where you could get some extra help,” “You could move into that extra bedroom we have and stay with us” or “I’m sure we could find another good home for the dog” are not uncommon. And, even though these comments may be practical solutions given the circumstances, they are also very difficult to hear, because they can signal a loss of independence.

I don’t advocate for staying at home at all costs. However, if clients would like to remain home or don’t have other options but to remain home, there are a number of solutions to safely do so. Having said that, using a lawn chair in the shower, a piece of quarter-inch plywood for a ramp, or stacking boxes in the living room so there is something to hold onto every couple of steps are not safe solutions. I’ve seen just about everything.

Medication Management

One of the greatest dangers to a person’s safety at home is medication errors, which are frequently related to confusion or memory concerns. However, they may also be related to weakness, fatigue or pain if a person is not able to independently open a pill container or if he or she has medications spread throughout the home and doesn’t have the stamina to gather them when it’s time.

Medications should be kept in an organizer and in one place so there are no missed or inaccurate doses. Many people use inexpensive plastic pill boxes. This is a better strategy than keeping medications in a shoebox or in several places throughout the home. However, if reminders are needed to take medications or if a person has difficulty remembering if he or she has taken medications, a tinted medication lockbox or other automated dispenser should be considered. The dispenser still needs to be organized by someone so it will automatically open the correct door on the pillbox or dispense the medication after alerting the individual of the proper time to take it.

Another alternative offered by some pharmaceutical companies is prepackaged and delivered medications in a color-coded blister pack for morning, afternoon and/or evening administrations, making organization relatively simple.

Functional Adaptations of the Home for Those with Weakness, Fatigue and/or Pain

Accommodations to manage medications and reduce fall risk can help patients remain in their own homes.

By Matthew D. Hansen, DPT, MPT, BSPTS
Falls

Risk of falls is another common safety issue at home. Falls are often people’s greatest fear, particularly if they live alone and are prone to balance issues associated with neuropathy, dizziness or weakness. Standard practice for most home health agencies is to conduct a home safety evaluation as part of their initial assessment. It’s a great place to start for anyone. If a person doesn’t contract with a home health agency, an evaluation by a skilled private-duty agency can be requested. Or, there are several safety checklists on the Internet to use to review the home with a friend or family member (see “Improving Safety, Mobility and Activities of Daily Living at Home” in the February-March 2017 issue of IG Living at www.igliving.com/magazine/articles/IGL_2017-02_AR_Improving-Safety-Mobility-and-Activities-of-Daily-Living-at-Home.pdf).

Following are modifications that can be considered for each area of the home:

Yard and front porch. Common hazards outside of the home include uneven ground and sidewalk/driveway surfaces, ice, wet leaves, sprinkler heads and tools or garden hoses that are not properly stored. Fallen leaves and grass clippings should be picked up promptly, and icy surfaces should be treated with ice melt or sand where ice melt products are not allowed or recommended.

Consider using an assistive device for support or a yard wagon to transport garden tools, supplies, etc., if two hands would otherwise be required. And, try to keep items as close as possible to the area they are needed (this is a great principle for inside and outside the home).

Look for ways to work in the garden without bending over. If sitting on the ground is not an option, consider sitting in a chair or using long-handled tools instead of bending over from a standing position.

For many, the greatest need for permanent modifications outside the home are on the front and back porches. There should be at least one handrail in place for any set of stairs, even when there aren’t more than one or two steps to navigate. Anti-slip tape can be added to the edge of the stair or tread for additional security, and special door delivery of mail can be requested from the post office.

For those who use an adaptive walker or a wheelchair, a ramp may be needed. If the home has only a couple steps, a prefabricated aluminum ramp may be sufficient. Otherwise, a customized metal or wood ramp may be required. Again, a piece of plywood might be functional, but it’s not safe and could lead to a life-altering or life-threatening fall.

Living spaces. In addition to keeping living spaces clutter-free, proper footwear inside and outside of the home is important. Heels, flip-flops and any footwear with an open back or slippery sole can be bad news. Additionally, if an assistive device has been prescribed, it should be used properly, even if it’s inconvenient. I’ve seen too many walkers and canes that were supposed to be aids become hazards because they were left in the way. Be sure to check rubber cane tips for wear, and replace them when needed.

Loose cords/cables and oxygen tubing are other common tripping hazards. Cords should be kept together and close to the wall rather than strewn across walking paths. A number of retail products can help with organization. Special colored tubing can be purchased for supplemental oxygen users, or bright-colored tape can be placed at 12-inch intervals along the tubing to alert the user and others of its presence.

Wherever possible, household and food items should be moved down from top shelves to eliminate the need for reaching overhead or using a step stool. Similarly, items on bottom shelves near the floor should be moved up to eliminate the need to bend below the waist. If moving low items is not an option, sitting down to access them may be a better one.

There are several considerations for improving transfers in the living room or bedroom. Oftentimes, having a properly height-adjusted bed is enough to allow someone to continue transferring independently. Motorized and manual height-adjustable beds and hospital beds are available. Other transfer-assist options include bed canes, transfer poles, power-lift recliners and hydraulic or ceiling track patient lifts to transfer someone to and from bed and a wheelchair or shower chair, or in the case of a ceiling track lift, to another location in the home (a second person is required to operate the lift).

Though they can be expensive modifications, some people may need to have inside doorways widened to accommodate a walker or wheelchair, and/or they may need a stair lift or elevator put in place if they are unable to negotiate stairs in the home and can’t live solely on the ground level. Where possible, laundry washers/dryers and other essentials should also be placed on the same level as the person’s living space to reduce the number of required trips up and down the stairs.

Finally, take advantage of developments in technology. Most appliances or other devices plugged into outlets (thermostat, television, washer/dryer, doorbell and other security
Important Safety Information

WARNING: Thrombosis (blood clots) 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 blood clots, your doctor will prescribe Hizentra at the minimum dose and infusion rate practicable and will monitor for signs of clotting events and hyperviscosity. Always drink sufficient fluids before infusing Hizentra.

See your doctor for a full explanation, and the full prescribing information for complete boxed warning.

Hizentra is a prescription medicine used to treat:

- Primary immune deficiency (PI) in patients 2 years and older
- Chronic inflammatory demyelinating polyneuropathy (CIDP) in adults

Treatment with Hizentra might not be possible if your doctor determines you have hyperprolinemia (too much proline in the blood), or are IgA-deficient with antibodies to IgA and a history of hypersensitivity. Tell your doctor if you have previously had a severe allergic reaction (including anaphylaxis) to the administration of human immune globulin. Tell your doctor right away or go to the emergency room if you have hives, trouble breathing, wheezing, dizziness, or fainting. These could be signs of a bad allergic reaction.

Inform your doctor of any medications you are taking, as well as any medical conditions you may have had, especially if you have a history of diseases related to the heart or blood vessels, or have been immobile for some time. Inform your physician if you are pregnant or nursing, or plan to become pregnant.

Infuse Hizentra under your skin only; do not inject into a blood vessel. Self-administer Hizentra only after having been taught to do so by your doctor or other healthcare professional, and having received dosing instructions for treating your condition.

Immediately report to your physician any of the following symptoms, which could be signs of serious adverse reactions to Hizentra:

- *Ig=immunoglobulin
• Reduced urination, sudden weight gain, or swelling in your legs (possible signs of a kidney problem).
• Pain and/or swelling or discoloration of an arm or leg, unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, or numbness/weakness on one side of the body (possible signs of a blood clot).
• Bad headache with nausea; vomiting; stiff neck; fever; and sensitivity to light (possible signs of meningitis).
• Brown or red urine; rapid heart rate; yellowing of the skin or eyes; chest pains or breathing trouble; fever over 100°F (possible symptoms of other conditions that require prompt treatment).

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

The most common side effects in the clinical trials for Hizentra include redness, swelling, itching, and/or bruising at the infusion site; headache; chest, joint or back pain; diarrhea; tiredness; cough; rash; itching; fever, nausea, and vomiting. These are not the only side effects possible. Tell your doctor about any side effect that bothers you or does not go away.

Before receiving any vaccine, tell immunizing physician if you have had recent therapy with Hizentra, as effectiveness of the vaccine could be compromised.

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

You can also report side effects to CSL Behring’s Pharmacovigilance Department at 1-866-915-6958.
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 indicated for:
* Treatment of primary immunodeficiency (PI) in adults and pediatric patients 2 years and older.
* Maintenance therapy in adults with chronic inflammatory demyelinating polyneuropathy (CIDP) to prevent relapse of neuromuscular disability and impairment.

LIMITATION OF USE: Maintenance therapy in CIDP has been systematically studied for 6 months and for a further 12 months in a follow-up study. Continued maintenance beyond these periods should be individualized based on patient response and need for continued therapy.

For subcutaneous infusion only.

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

CONTRAINDICATIONS
- Anaphylactic or severe systemic reaction to human immune globulin or components of HIZENTRA, such as polysorbate 80
- Hyperprolinemia (type I or II) (HIZENTRA contains the stabilizer L-proline)
- IgA-deficient patients with antibodies against IgA and a history of hypersensitivity

WARNINGS AND PRECAUTIONS
- IgA-deficient patients with anti-IgA antibodies are at greater risk of severe hypersensitivity and anaphylactic reactions.
- Thrombosis may occur following treatment with immune globulin products, including HIZENTRA.
- Aseptic meningitis syndrome has been reported with IGIV or IGSC, including HIZENTRA treatment.
- Monitor renal function, including blood urea nitrogen, serum creatinine, and urine output in patients at risk of acute renal failure.
- Monitor for clinical signs and symptoms of hemolysis.
- Monitor for pulmonary adverse reactions (transfusion-related acute lung injury [TRALI]).
- HIZENTRA is made from human plasma and may contain infectious agents, e.g., viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.

ADVERSE REACTIONS
The most common adverse reactions observed in >5% of study subjects were local infusion site reactions, headache, diarrhea, fatigue, back pain, nausea, pain in extremity, cough, upper respiratory tract infection, rash, pruritus, vomiting, abdominal pain (upper), migraine, arthralgia, pain, fall and nasopharyngitis.

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.

Based on March 2018 revision
cameras, lighting, etc.) now have an option (or model) that can be controlled from a remote control or smartphone. Amazon Echo, Google Home and other smart devices have many features to make life easier. The point isn’t to eliminate physical movement, but to increase “smart” movement so energy can be conserved, safety improved and pain reduced.

Bathroom. Walk-in bathtubs or showers with a barrier-free threshold are ideal; however, they aren’t an alternative for everyone. To improve transfers and safety in a more traditional tub shower, a transfer bench can be used. If transfers aren’t the problem but maintaining a standing position in the shower is, utilize a shower chair (not a lawn chair). Handheld showerheads help to wash and rinse difficult-to-reach areas, as can a long-handled sponge. Don’t forget nonslip bath mats (inside and outside of the shower/tub).

Grab bars are an important installation in any bathroom, and often in other areas of the house, too. Be sure to have them mounted properly per the manufacturer’s instructions, and don’t use suction cup grab bars, even though the convenience of a movable product might be tempting. They simply aren’t as safe. An occupational or physical therapist can help to indicate proper positioning of the bars.

To make toileting easier, a raised toilet seat and/or toilet frame can be acquired. Availability of a commode chair might be a good idea for those who aren’t able to safely make it into the bathroom or if they don’t have enough energy to do so as often as their body demands.

Personal Care

When more help is required than what family, friends or someone can provide, employing the services of a personal care agency may be considered before looking at other options. Caregiver aides can assist with just about anything a person might need help with or can no longer do on their own, including meal preparation, errands, shopping, transportation, bathing, dressing, toileting, companionship and housekeeping. An agency designated as “skilled” can also provide nursing services at home, including medication management, infusions, triage/wellness checks, wound care, etc., even when insurance will not pay for services through a home-health episode.

Unfortunately, personal care still falls in the realm of private duty and, unless someone has a long-term care insurance policy or Veteran’s Administration (VA) benefits, these services will have to be paid out of pocket. And, while the Centers for Medicare and Medicaid Services now allows Medicare Advantage plans to cover personal care services, there aren’t many companies paying for them yet.

Smart Spending

Much home adaptation equipment typically not covered by insurance (e.g., grab bars, shower chairs, raised toilet seats, transfer benches) can be found in local home-improvement stores or drugstores.

Insurance companies don’t yet pay for bathroom equipment. Therefore, before purchasing something new, used equipment can sometimes be found on the Internet by searching for “adaptive equipment foundations” or “adaptive equipment loaning closets.” Or, equipment can sometimes be found by searching for “adaptive home modifications,” “funding adaptive home modifications” and “adaptive housing.” In addition, local thrift shops often have used equipment. Be sure, though, that any used equipment is thoroughly inspected for safety. Veterans should also check with the local VA clinic to ask for help.

For more expensive equipment and modifications (e.g., stair lifts, patient lifts, assistive devices), a doctor’s order and a good letter of medical necessity from a therapist or physician may be sufficient to get insurance approval. The letter should focus on how the equipment will improve the intended user’s independence and protect their safety, with an explanation of what anticipated consequences would ensue without the equipment. Also, some patient advocacy groups have equipment grants for individuals with a specific medication condition.

Staying in Place

Don’t get discouraged if money is tight, and don’t be afraid to spend the money if support is needed. While some modifications can be expensive, there are resources to help. The alternatives, whether they be voluntary relocation or a required rehab stay in a hospital or skilled nursing facility due to injury or self-neglect, can be much more costly to the pocketbook, the medical system and a person’s independence.

MATTHEW DAVID HANSEN, DPT, MPT, BSPTS, is a practicing physical therapist in Utah and president of an allied healthcare staffing and consulting agency named SOMA Health, LLC. He completed his formal education at the University of Utah, Salt Lake City, and has additional training in exercise and sports science, motor development and neurological and pediatric physical therapy.
Four Most Common Antibody Deficiencies in the First Year of Life

Infants are often diagnosed with these four antibody deficiencies, two of which can cause serious complications, however they resolve with time for most.

By E. Richard Stiehm, MD

**Physiologic Hypogammaglobulinemia**, specific antibody deficiency (SAD), hypogammaglobulinemia of the newborn and transient hypogammaglobulinemia of infancy are the four most common antibody deficiencies infants experience in their first year of life. The infant in the clinical vignette (opposite page) had all four antibody deficiencies, two of which are present in every young child (physiologic hypogammaglobulinemia and SAD), while the other two (hypogammaglobulinemia of the newborn and transient hypogammaglobulinemia of infancy) are less common and more serious after 6 months of age.

All of these disorders predispose infants to infection, contributing to the high risk of death during their first year of life. The first month of life is the most critical with deaths due to profound prematurity; birth defects; maternal complications; placental, cord and membrane complications; and bacterial sepsis. The next five months of life are also hazardous because of congenital malformations, sudden infant death syndrome, unintentional injuries, circulatory disorders and homicide.¹ These disorders will be detailed below, preceded by a brief summary of B cell and immunoglobulin development.

**Fetal B Cells**

B cells (CD19+), the basic cells of the immunoglobulin system, first appear in the liver, bone marrow and circulation at approximately the eighth week of gestation. Fetal B cells increase steadily all through gestation so, by term birth, the B cells represent 15 percent of peripheral blood lymphocytes and number approximately 400 cells/ul.² These intrauterine B cells do not develop into plasma cells for immunoglobulin production for the circulation or secretions, in part because of immaturity and in part because of lack of antigen stimulation within the sterile womb. Thus, these B cells are antigenically naive with a CD19+CD27-phenotype, unlike memory B cells that have a CD19+CD27+phenotype.

**Transplacental IgG Immunoglobulin**

The lack of fetal immunoglobulin production is compensated...
by the placental transfer of maternal IgG immunoglobulin beginning about the 12th week of gestation. This transfer accelerates during late pregnancy so, by the time of term birth, the infant’s IgG level is slightly higher than the maternal IgG level. This transfer is accomplished by the FcRn, the neonatal FcReceptor. The Fc stands for the tail of the Y-shaped IgG immunoglobulin molecule. This receptor is present on placental syncytotrophic cells at the maternal-fetal interface where it ingests maternal IgG and transfers it to the infant’s circulation. This transfer is restricted to IgG; no other maternal immunoglobulin (IgA or IgM) is transferred (Figure).

Maternal IgG provides infants with protective antibodies to the microbes they have encountered either from infection or from immunization. This emphasizes the importance of immunization for prospective mothers before or during pregnancy (excluding live virus vaccines during pregnancy). Premature infants will receive less of these protective antibodies and thus are more susceptible to infection than are term infants.

Immunoglobulin Levels in the First Year of Life

The maternal IgG transferred to her newborn has a half-life of 30 days so it disappears gradually from the infant’s circulation over a six-month period. By that time, infants have started making their own IgG and IgM and are able to respond to vaccines and microbes. Newborns first make IgM, then IgG and finally IgA, so by 1 year of age, newborns have an IgM 75 percent of an adult’s, an IgG 60 percent of an adult’s and an IgA 35 percent of an adult’s as shown in the Figure.

I. Hypogammaglobulinemia of the Newborn

Prematurity is the most common cause of neonatal hypogammaglobulinemia. At 8 weeks of age, he started his routine childhood vaccines, except for the live oral rotavirus vaccine that is not given in the newborn nursery because of possible spread to other newborns. He was discharged at 10 weeks of age with a weight of 2,200 grams. His IgG level was 240 mg/dl, thus he was diagnosed with hypogammaglobulinemia of the newborn.

Asa did well at home with weekly visits by a visiting nurse and monthly checks by his pediatrician. His vaccines were continued, including oral rotavirus vaccine, and he was started on monthly Synagis for respiratory syncytial virus prophylaxis since it was the winter months. At 4 months of age, his IgG was 280 mg/dl, his IgM was 28 mg/dl and his IgA was less than 5 mg/dl. He was then diagnosed with physiologic hypogammaglobulinemia of infancy, aggravated by his low IgG level at birth.

He did well at home and by 8 months of age, he weighed 7 kg and had only one febrile illness with rhinorrhea (thin nasal mucus discharge) and a cough that lasted a week. His IgG level was 320 mg/dl, his IgM level was 40 mg/dl and his IgA level was 8 mg/dl. He had low but protective antibodies to tetanus and diphtheria and several pneumococcal serotypes. He was then diagnosed with transient hypogammaglobulinemia of infancy.

At 11 months of age, Asa developed bilateral otitis media (earache affecting both ears) resistant to amoxicillin. A myringotomy (a tiny surgical incision in the eardrum to relieve pressure caused by excessive buildup of fluid from the middle ear) with culture revealed a pneumococcus serotype 14N not present in the conjugated pneumococcal vaccine (Prevnar 13) but present in the 23-valent pneumococcal polysaccharide vaccine (Pneumovax 23). For this reason, Asa was given the latter vaccine and his titers were checked before being vaccinated and one month after vaccination. This disclosed protective titers to eight of the Prevnar serotypes but only two of the unique serotypes in Pneumovax (not present in Prevnar). His IgG had normalized at 450 mg/dl, and he was then diagnosed with specific antibody deficiency (also known as impaired polysaccharide responsiveness), which is common at this age, that persisted until age 3 years.

A Clinical Vignette

Baby boy Asa was born prematurely at 31 weeks because of maternal bleeding. It was the 21-year-old mother’s first pregnancy, and she was in good health with a normal IgG level of 1,090 mg/dl. Asa’s immunoglobulins at birth were IgG 280 mg/dl, IgM 5 mg/dl and IgA less than 5 mg/dl. A heel stick TREC test excluded severe combined immunodeficiency disease.

Asa did well in the nursery on two weeks of tube feedings and supplemental oxygen. At 8 weeks, he started his routine childhood vaccines, except for the live oral rotavirus vaccine that is not given in the newborn nursery because of possible spread to other newborns. He was discharged at 10 weeks of age with a weight of 2,200 grams. His IgG level was 240 mg/dl, thus he was diagnosed with hypogammaglobulinemia of the newborn.

He did well at home with weekly visits by a visiting nurse and monthly checks by his pediatrician. His vaccines were continued, including oral rotavirus vaccine, and he was started on monthly Synagis for respiratory syncytial virus prophylaxis since it was the winter months. At 4 months of age, his IgG was 280 mg/dl, his IgM was 28 mg/dl and his IgA was less than 5 mg/dl. He was then diagnosed with physiologic hypogammaglobulinemia of infancy, aggravated by his low IgG level at birth.

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hypogammaglobulinemia (an IgG level less than 400 mg/dl during the first two months of life). Hypogammaglobulinemia is generally present in infants born before 32 weeks of gestation and/or weighing less than 1,500 grams. Ballow et al. reported the mean IgG level of prematures born at 25 weeks to 28 weeks of gestational age was 251 mg/dl (range 114-552 mg/dl), and the mean IgG level of infants born from 29 weeks to 32 weeks gestation was 368 mg/dl (range 186-298 mg/dl). Despite their immaturity and in the absence of other illnesses, these newborns are able to respond to routine childhood vaccines by 8 weeks of age. But, they are kept in isolation until ready for discharge at 38 weeks and weighing 2,100 grams if they are otherwise well.

Hydrops fetalis due to maternal-fetal Rh incompatibility was a common cause of neonatal hypogammaglobulinemia before its prevention with Rh immunoglobulin. These critically ill infants have generalized edema (swelling caused by excess fluid trapped in the body’s tissues) associated with severe anemia (a lack of healthy red blood cells) due to lysis (destruction) of their Rh positive red cells by transplacentical maternal Rh antibodies. The anemia results in heart failure with loss of plasma proteins, including IgG, into the soft tissues and serous cavities. Placental edema may also decrease maternal-fetal IgG transfer.

Less-common causes of hydrops include severe anemia due to congenital hemoglobinopathies, maternal infections, particularly parvovirus, congenital heart disease with heart failure and several other genetic disorders. All require circulatory support, correction of the anemia and replacement of serum albumin and immunoglobulin.

Maternal hypogammaglobulinemia will result in newborn hypogammaglobulinemia, dependent on the maternal IgG level and the newborn gestational age. The most common cause is maternal rituximab therapy, a monoclonal antibody to CD20 on B cells, which causes a marked decrease in B cells, plasma cells and IgG levels. Rituximab is used in several diseases associated with harmful autoimmune antibodies such as immune thrombocytopenic purpura or autoimmune hemolytic anemia. It also renders mothers hypogammaglobulinemic with less IgG transferred to their infants. If rituximab is given during late pregnancy, it may depress B cells in infants. Other immune-suppressive drugs may also decrease maternal IgG levels, but not usually to the degree as rituximab therapy.

Additional causes of maternal hypogammaglobulinemia include untreated immunodeficiencies such as common variable immunodeficiency, parvovirus infection, hemodialysis (procedure to remove fluid and waste products from the blood and to correct electrolyte imbalances) or immunoglobulin loss into the GI tract, urine or serosal spaces such as the pleura (chylothorax) or peritoneum (chylous ascites).

Immunoglobulin loss in infants’ circulation may also result in neonatal hypogammaglobulinemia. This loss can occur into the urine in congenital nephrosis (kidney disease), into the bowel in early onset protein-losing enteropathy (abnormality of the intestinal tract) or into serosal spaces such as the pleura or peritoneum. Immunoglobulin loss can also result with extensive blood loss due to hemorrhage, surgery (particularly cardiopulmonary bypass surgery), frequent blood sampling or the twin-twin transfusion syndrome (one twin bleeds into the other, causing anemia and hypogammaglobulinemia in the donor twin).

Management of these disorders is first directed at the underlying cause of the disease. Immune globulin (IG) therapy may be considered if there is refractory infection not responding to antibiotics. Sometimes IG is used to reduce hemolysis.

II. Physiologic Hypogammaglobulinemia of Infancy

All infants have a significant decrease in their IgG levels between 2 months and 6 months of age as maternal IgG levels disappear and the infants’ own IgG synthesis is not yet well-established. This is markedly severe and prolonged if babies are born prematurely as noted above. The presence of systemic infection or inflammatory diseases may accelerate IgG catabolism (the part of the metabolism responsible for breaking complex molecules down into smaller molecules) with resulting aggravation of the already low IgG levels. Blood or protein loss due to any reason will also aggravate the hypogammaglobulinemia. Despite these low levels and in the absence of systemic disease, routine immunizations should be continued. IG therapy is not indicated simply based on low IgG levels.

The first six months of life is the period of highest infant mortality, as noted above. Hypogammaglobulinemia may contribute to infants’ demise, particularly in infections, sudden infant death syndrome and the pneumonia that accompanies many of these disorders.

III. Transient Hypogammaglobulinemia of Infancy (THI)

THI occurs in infants older than 6 months of age who have an IgG less than two standard deviations below the mean for age with or without low levels of IgM and IgA. I prefer the simple definition of an IgG less than 400 mg/dl with measurable levels of IgA and IgM immunoglobulins, thus excluding selective IgA or IgM deficiency. Antibody titers are usually present but at reduced titers.

THI is among the most common antibody deficiencies in
several immunodeficiency registries that include children, with its incidence similar to selective IgA deficiency and SAD. Two-thirds of infants with THI are boys. THI usually resolves by 5 years of age but may persist up to 10 years of age and beyond. Thus, one authority suggests it should be termed transient hypogammaglobulinemia of childhood. A definitive diagnosis cannot be made until patients recover. The cause is not known, and it is not associated with known genotypes.

In infants with THI, immunoglobulin levels should be measured every 6 months to 12 months to follow progression of the disease. Antibody titers to protein vaccine antigens are usually protective but often slightly depressed. Most children have a concomitant SAD that may be prolonged (see below). A complete absence of an antibody response suggests a primary antibody deficiency. And, while B cell and T cell numbers are normal, B cells show a naive phenotype.

Many of these children remain well, but others have frequent respiratory infections, asthma, food intolerance or eczema. These children should be given their childhood vaccines. Continuous antibiotics and intravenous IG use is reserved for refractory and/or recurrent infections, with most infants experiencing only mild infections.

IV. Specific Antibody Deficiency (SAD)

SAD is defined as an inadequate response to polysaccharide antigens but normal responses to protein antigens, normal levels of immunoglobulin and no other immunodeficiency syndrome. SAD is present in most infants after their IgG normalizes at about 6 months of age, and it persists in most infants until 24 months of age and often longer.

SAD is usually diagnosed by an inadequate response to the 23-valent pneumococcal polysaccharide vaccine. Adults should develop protective titers to 70 percent of the polysaccharide serotypes, while children under 6 years should respond to 50 percent of them.

In children, SAD is associated with frequent respiratory infections, including sinusitis and chronic otitis (ear infection). It is also the most common antibody disease in adults, usually manifested by chronic rhinosinusitis. Up to 40 percent of patients with these symptoms have SAD, as do 10 percent of adult controls. Patients over age 60 years are more likely to have SAD. And, although SAD in children often disappears after several months, the diagnosis in adults usually persists for a lifetime.

The cause of SAD is not known and probably multifactorial. It is a common secondary immunodeficiency in many chronic conditions such as hematologic malignancies, immunosuppressive therapy and older adults. Treatment includes antibiotics and occasionally IG therapy in refractory or persistent infection.

The Need for Long-Term Follow-Up and Vaccines

Two of the four common antibody deficiencies, THI and SAD, are experienced by all infants but are usually asymptomatic with a good prognosis. Neonatal hypogammaglobulinemia is common in premature infants but also associated with serious conditions. THI is often associated with frequent respiratory infections and allergies. Long-term follow-up of these illnesses is imperative, since most infants recover with time. Rarely are these antibody deficiencies precursors for adult disease. Vaccines are used both as therapy and diagnosis. Vaccines for pregnant mothers, infants and their families are important parts of optimal management, and they may prevent the need for antibiotics and illness in infants and their family members.

E. RICHARD STIEHM, MD, is professor of pediatrics at the David Geffen School of Medicine at the University of California, Los Angeles.

References
New diagnostic tools and treatment strategies have emerged during the past several years for this rare neurological disorder.

By Ronale Tucker Rhodes, MS

THE FIRST probable cases of chronic inflammatory demyelinating polyneuropathy (CIDP) were described by J.H. Austin, MD, in 1958 as a “fluctuating motor-predominant neuropathy that produced severe weakness that would either improve spontaneously or in response to corticosteroids” with “focal areas of segmental demyelination rather than axonal degeneration” likely the pathological cause. It wasn’t until 1975 that the name chronic inflammatory polyradiculoneuropathy was coined by Peter James Dyck, MD, and colleagues after conducting a historical study of 53 personally evaluated patients with these symptoms. Later, the term demyelinating was subsequently added, which defined CIDP as a separate disease entity.1 (The terms polyradiculoneuropathy and polyneuropathy are used interchangeably.)

CIDP is rare, affecting approximately 40,000 people in the U.S. Its estimated incidence is 0.7 cases to 1.6 cases per 100,000 persons per year, and its overall prevalence is estimated to be 5 cases to 9 cases per 100,000 individuals, with an estimated prevalence in children to be 0.5 cases per 100,000. The disease can affect any age group, and onset can begin during any decade in life. However, CIDP affects twice as many males as females, and the average age of onset is 50 years.2,3
Since its distinction as a separate disease from similar neuropathies, advances have continued to be made in understanding this debilitating disorder. In fact, the past several years have produced some significant new diagnostic tools and treatment strategies.

**What Is CIDP?**

CIDP is the most common chronic immune-mediated polyneuropathy, and is sometimes called chronic relapsing polyneuropathy or chronic inflammatory demyelinating polyradiculoneuropathy because it involves the nerve roots. It is a progressive autoimmune disease that destroys the myelin sheath (the fatty covering that wraps around and protects nerve fibers and assists in nerve signal transmission) of peripheral nerves.¹ The result is a slowing of the nerve signals and subsequent weakness in the muscles they control. CIDP has a variable course that can be relapsing-remitting (relapses and periods of stability in between relapses), stepwise progressive or gradually progressive.²

There are two categories of CIDP: typical and atypical. Typical CIDP is the most common subtype and accounts for at least 50 percent to 60 percent of all cases. It is a fairly symmetric sensorimotor polyneuropathy in which proximal and distal motor involvement exceeds sensory involvement. It presents with gradually progressive symptoms over the course of several months or longer. Some patients present with more rapidly progressive symptoms, which have been termed “acute-onset CIDP.” However, the diagnosis of CIDP is dependent on progression or relapse of the disease over greater than eight weeks.

Atypical variants of CIDP are distinguished by their clinical presentation and/or pathogenic (organism causing disease) mechanism. These include:³

- Lewis-Sumner syndrome, also known as multifocal acquired demyelinating sensory and motor neuropathy, a well-described atypical variant that accounts for 5 percent to 10 percent of CIDP cases;
- The sensory-predominant form of CIDP characterized by symptoms and signs consistent with large fiber sensory dysfunction, including balance problems, pain, paresthesias (pins and needles) and dysesthesias (abnormal sensation);
- Distal acquired demyelinating symmetric neuropathy, a distal and sensory-predominant variant of CIDP, which is usually more slowly progressive than typical CIDP;
- A proximal variant of CIDP in which inflammatory demyelination is confined to dorsal (sensory) nerve roots;
- A pure motor variant of CIDP reported in a small number of cases;
- Neurofascin (NF) antibody-mediated CIDP with autoantibodies to NF155 that appear to be younger and more likely to have sensory ataxia (lack of muscle control or coordination) and prominent tremor, which can cause severe dysfunction; and
- Contactin 1 (CNTN1) antibody-mediated CIDP caused by autoantibodies of the IgG4 class to CNTN1 or CNTN-associated protein 1, which is found in a small subset of patients.

**Symptoms of CIDP**

CIDP symptoms usually begin in the feet and move slowly over time up the legs and arms, typically affecting both sides of the body (Table 1). Both proximal and distal muscles can be involved. Symptoms reported include:⁴

- Initial limb weakness, both proximal and distal
- Sensory symptoms (e.g., tingling and numbness of hands and feet)
- Motor symptoms (usually predominant)
- Symptoms of autonomic system dysfunction (e.g., orthostatic dizziness)
- Preceding infection (infrequent)
- A relatively acute or subacute onset of symptoms in about 16 percent of patients
- Usually a more precipitous onset of symptoms in children

**Table 1. Common Manifestations of CIDP**

- Gradually worsening paresthesia (pins and needles feeling) and numbness
- Muscle weakness in the legs and arms
- Areflexia (absent tendon reflexes) without wasting
- Preferential loss of vibration of joint position sense
- Foot drop and difficulty getting out of a chair
- Difficulty with fine finger control
- Sensory ataxia
- Fatigue

When the condition is associated with other diseases, symptoms may include:

- Signs of cranial nerve involvement (e.g., facial muscle paralysis or diplopia)
- Gait abnormalities
- Motor deficits (e.g., symmetric weakness of both proximal and distal muscles in upper and lower extremities)
- Diminished or absent deep tendon reflexes
- Sensory deficits (typically in stocking-glove distribution)
- Impaired coordination

The rate and severity of progression of weakness varies from person to person; however, CIDP usually presents slowly over several months and has ongoing symptoms for more than eight weeks and usually does not improve unless ongoing treatment is given. This is in contrast to the acute form of demyelinating neuropathy known as Guillain-Barré syndrome (GBS). GBS presents with a rapid progression of symptoms occurring over days or weeks that usually warrants hospitalization due to involvement of the breathing muscles. Respiratory involvement does not occur in CIDP.

Causes of CIDP

While it is unknown what causes CIDP, it is believed to be an autoimmune disorder. Autoimmune disorders occur when the body’s natural defenses (antibodies and lymphocytes) against invading organisms suddenly begin to attack healthy tissue. With CIDP, the autoimmune disorder causes the immune system to attack the myelin cover of the nerves causing inflammation of nerves and nerve roots.

Table 2. CIDP-Related Disorders

- Lewis-Sumner syndrome: A rare neurological disorder characterized by asymmetric or multifocal weakness and sensory dysfunction affecting the arms and legs.
- Multifocal motor neuropathy: A rare disorder characterized by asymmetric or multifocal weakness of the arms and legs without sensory signs or symptoms.
- Charcot-Marie-Tooth disease: A group of inherited neurological disorders that progressively affect movement.
- Multiple sclerosis: A chronic disease of the brain and spinal cord that may be progressive, relapsing, remitting or stable.

In addition, CIDP may occur in association with other conditions such as diabetes (more than half of people with diabetes develop some type of neuropathy), infections, medications (especially those used to treat cancer), bone marrow disorders (including an abnormal protein in the blood, a form of bone cancer, lymphoma and amyloidosis), other diseases (including kidney disease, liver disease, connective tissue disorders and hypothyroidism) and alcoholism. However, it should be noted that the vast majority of patients with CIDP cannot identify a specific cause.

While previously there did not seem to be a genetic link to CIDP, one study did suggest a possible genetic cause. In that study, patients with CIDP had a high frequency of perforin gene variations that impair the function of cytotoxic T and natural killer cells, which may influence the development and course of CIDP.

Diagnosing CIDP

Not only is CIDP difficult to diagnose, it can go undiagnosed for months or years either because symptoms aren’t prominent enough or because nondebilitating symptoms in the early stages make it difficult to make a definitive diagnosis. In addition, according to recent studies, CIDP is misdiagnosed in up to 50 percent of cases, with alternative diagnoses given such as motor neuron disease, diabetic and inherited polyneuropathy, and even conditions clinically distinguishable from CIDP such as fibromyalgia or multiple sclerosis. Notably, there are some related disorders that can be useful in a differential diagnosis of CIDP (Table 2).

While many sets of diagnostic criteria have been developed for CIDP, the one used most often in current clinical practice was developed by the European Federation of Neurological Societies and the Peripheral Nerve Society (EFNS/PNS). This is mainly due to the high specificity and lack of sensitivity of other diagnostic criteria that can lead to underdiagnosis. For instance, a study of 151 CIDP patients and 162 control patients found the EFNS/PNS criteria had a sensitivity of 81.3 percent and specificity of 96.2 percent for definite or probable CIDP, whereas sensitivity of other diagnostic criteria ranged from 45.7 percent to 79.5 percent.

According to the EFNS/PNS criteria, “CIDP should be considered in any patient with a progressive symmetrical or asymmetrical polyradiculoneuropathy in whom the clinical course is relapsing and remitting or progresses for more than two months, especially if there are positive sensory symptoms, proximal weakness, areflexia (no muscle response to stimuli).
without wasting, or preferential loss of vibration or joint position sense.” EFNS/PNS criteria also state electrophysiologic tests are mandatory. These tests can help determine whether an individual has CIDP by looking at the demyelination process. Other features that support a diagnosis include elevated cerebrospinal fluid protein without an increased leukocyte (white blood cell) count; MRI evidence of gadolinium enhancement or nerve root plexus hypertrophy; nerve biopsy finding of primary demyelination; and improvement following immunotherapy.

Most recently, it has been found a small subset of patients harbor one of two autoantibodies: isoforms of NF155 and NF186, as well as anti-CNTN1 antibodies, which target a specific portion of peripheral nerves and may have important treatment implications. Anti-CNTN1 antibodies are found in between 2.2 percent and 8.7 percent of patients, and NF155 and NF186 have been detected in between 4 percent and 18 percent of CIDP patients. Anti-CNTN1 antibody-positive patients are clinically distinct with predominant involvement of motor fibers and axonal (the appendage of the neuron that transmits impulses away from the cell body) damage. Clinical features associated with NF155 seropositivity are younger onset, tremor and sensory ataxia. Patients with NF186 (and NF140 found in less than 2 percent of patients) have associated autoimmune disorders, are severely affected and present with sensory ataxia but not tremor.

**Treating and Managing CIDP**

Treatment of CIDP is complex and has to be individualized for each patient. The goal of treatment is to block immune processes to stop inflammation and demyelination, as well as to prevent secondary axonal degeneration. Secondary goals of treatment are to reduce symptoms such as weakness and pain and to improve overall functional status, as well as to reduce the frequency of relapses and slow disease progression.

First-line treatments include corticosteroids to suppress the immune system, intravenous immune globulin (IVIG) to infuse antibodies into the blood, and plasmapheresis to remove harmful antibodies.

Corticosteroids have been used to treat patients for more than 40 years, yet there remains no consensus about the optimal regimen. There was a retrospective study that compared regimens (daily oral prednisolone, pulsed oral dexamethasone or pulsed intravenous methylprednisolone) in 125 CIDP patients, 60 percent of whom responded to them with no significant difference in safety and efficacy between the three regimens. The main benefit of corticosteroids is their low acquisition cost. The downside, however, is adverse effects from long-term use, which can include osteoporosis and fractures, adrenal suppression and Cushing syndrome, hyperglycemia, hypertension, psychiatric disturbances, cataracts, weight gain and immunosuppression. Studies have shown dexamethasone and pulsed intravenous methylprednisolone have a lower risk of adverse effects than prednisolone.

The benefit of IG therapy is attributed to anti-inflammatory activity. Currently, there are three U.S. Food and Drug Administration (FDA)-approved IVIG products to treat CIDP: Gamunex-C (Grifols), Gammaked (Kedrion) and
Privigen (CSL Behring). The standard maintenance treatment regimen is 1.0 g/kg every three weeks, with studies showing high response rates. One study showed a response rate of almost 70 percent after 52 weeks at this maintenance regimen.

It should be noted that both anti-CNTN1-positive and NF155-positive patients show a poor response to IVIG. However, some case studies suggest these patients may benefit from treatment with rituximab (a monoclonal antibody discussed below). NF140 and NF186-positive patients do respond well to IVIG or corticosteroids.

In 2018, FDA approved Hizentra (CSL Behring) as the first subcutaneous IG (SCIG) therapy to prevent relapse of neuromuscular disability and impairment. It is the only SCIG approved for this indication based on data from the Phase III PATH (Polyneuropathy And Treatment with Hizentra) study, the largest controlled clinical study in CIDP patients to date. In the PATH trial, patients taking Hizentra relapsed or withdrew less often than those taking placebo. Patients in the study also maintained their grip strength, as well as upper- and lower-body strength. Another study found both SCIG and IVIG are equally effective. That study compared CIDP patients who received SCIG for five weeks to patients who received IVIG for five days after 10 weeks of opposite treatment, and found both treatments had similar effects on muscle strength. However, there are three differences between IVIG and SCIG. SCIG offers improved quality of life since patients can self-administer at home and a lower relative risk of systemic adverse effects (fever, headache, nausea). SCIG is also a therapeutic alternative for patients who suffer wear-off phenomena (loss of benefit) before the start of their next cyclical IVIG infusion.

PE, although used less frequently, is a method for removing unwanted substances (toxins, metabolic substances and plasma parts) from the blood. With PE, blood is removed from the individual, blood cells are separated from the plasma, the plasma is replaced with other human plasma and the patient’s blood cells are transfused back into the individual, thus removing only the plasma and its constituents. PE is similar to IVIG since it is only effective for a few weeks, so it requires chronic intermittent treatments.

In cases where first-line treatments fail, other immunosuppressants can be tried. These include azathioprine, mycophenolate mofetil and methotrexate. Monoclonal therapies that have been found to be effective in multiple sclerosis are also being studied to treat CIDP. Rituximab, a monoclonal antibody that targets a certain portion of B cells, including those that play a role in the immune response thought to occur in autoimmune conditions, has shown to be beneficial particularly in those positive for anti-CNTN1 and anti-F155 antibodies. And, alemtuzumab, which targets B cells and T cells, could provide a broader attack on the immune system. However, one that has not shown to be effective is fingolimod, a drug that affects the immune response thought to occur in autoimmune conditions.

After treatment is started, it must be continued in those who respond to it until the condition is stabilized or greatly improved. Response to treatment is measured by improvements in sensation, strength and the performance of activities of daily living. Assessment tools to monitor response to therapy include the Rasch-Built Overall Disability Scale to measure disability and the Martin Vigorimeter to measure grip strength, among others (Table 3).

Table 3. CIDP Outcome Measures

- Inflammatory neuropathy cause and treatment (INCAT) disability scale and sensory subscore: Measures upper and lower limb dysfunction
- Overall disability sum score (ODSS): Measures the function of the upper and lower limbs
- Overall neuropathy limitations scale (ONLS): Modified ODSS; ODSS item “Does the patient have difficulty walking?” was changed to “Does the patient have difficulty walking, running or climbing stairs?” The remaining scoring criteria are not different from ODSS
- Rasch-built overall disability scale (RODS): Measures upper and lower limb disability
- GAITrite: Measures gait parameters (velocity, cadence, swing phase, double support time, stance phase)
- Timed up and go test (TUG): Time taken to stand up from a chair, walk a short distance, turn around, return and sit down again
- 10-meter walk test (10MWT): Measures walking speed
- Dynamometer or vigorimeter: Measures grip strength
- Manual muscle strength testing and isokinetic strength testing: Muscle strength testing
- Fatigue severity scale (FSS): Measures fatigue
- SF-36: Quality-of-life measure (physical functioning, role functioning, social functioning, body pain, mental health, vitality, general health perception and change in health)
- Chronic acquired polyneuropathy patient reported index (CAP-PRI): Quality-of-life measure (physical function, social function, pain and emotional well-being)

Lastly, physiotherapy plays an important role in treatment. Physiotherapists prescribe gait aids to assist with balance and ambulation, manual therapy to prevent joint contractures and maintain available range of motion, and exercises to promote muscle strengthening and aerobic conditioning.10

A Costly Disease

While rare, CIDP poses a substantial clinical and economic burden among the thousands of patients affected by this neurological disorder of the peripheral nervous system. In a recent retrospective case-control analysis using data from the IQVIA Real-World Data Adjudicated Claims, adults newly diagnosed with CIDP between July 1, 2010, and June 30, 2014, were identified and direct matched to controls without CIDP. The researchers assessed and compared baseline characteristics over a six-month pre-index period; healthcare resource use, costs and clinical characteristics over a two-year follow-up; and total cost differences over the two-year follow-up between matched cohorts. They found that compared to controls, more CIDP patients had greater than or equal to one hospitalization (26.2 percent versus 9.0 percent), and a higher mean number of outpatient prescription fills (62.8 versus 32.0) and physician office visits (34.7 versus 13.0). In addition, CIDP patients had 7.5-times higher mean total costs ($116,330 vs. $15,986). Important cost drivers were costs for outpatient ancillary, radiology and HCPCS drugs (mean $76,366 versus $4,292) and costs for inpatient care (mean $16,357 versus $2,862). CIDP therapy (inclusive of both outpatient pharmacy and medical claims) accounted for 51.2 percent of mean total costs.11 In an earlier study conducted in 2011 that analyzed insurance claims data for 73 CIDP patients, the annual health plan cost per patient was almost $57,000. Pharmacy claims were the primary cost driver (57 percent), with IVIG therapy accounting for 90 percent of those costs.2

Fortunately, a lot has been learned over the last few years. According to Jeffrey A. Allen, MD, a member of the GBS/CIDP Foundation Global Medical Advisory Board, there are currently a multitude of studies around the world exploring ways to get more out of current CIDP treatments. And, he says, equal to this is identification of new treatment options.13

Yet, while much progress is being made, there remains an unmet need to improve diagnostic criteria and find clinical or biological variables that can predict treatment response. But, it is believed these issues can be uncovered through collecting and analyzing data from patients with CIDP in large registries and biobanks. According to the GBS/CIDP International website, a multidisciplinary group at the European Neuromuscular Center workshop recently compared eight currently ongoing international CIDP registries that included a total of more than 1,300 patients to assess infrastructure and collect clinical data, diagnostic data and biomaterials. The outcome was a decision to set up a central database, known as INCBase, to upload data from current registries and databases while these registries continue to exist. The global database is expected to be operational in mid-2020 and will collect data from thousands of CIDP patients to enable solving some of the important challenges in diagnosing and treating CIDP.17

RONALE TUCKER RHODES is the editor of IG Living magazine.

References
Profile: Erica Erdman

By Trudie Mitschang

WHEN ERICA Erdman’s son Braxton was born, his Down syndrome diagnosis took the family by surprise. But, any concerns about that diagnosis were soon eclipsed by Braxton’s frequent illnesses and hospital stays. Suspecting something was wrong, Erica fought to get an accurate diagnosis, and at 3 years old, her son was diagnosed with a primary immunodeficiency. Since then, the family has faced numerous hurdles, including the recent nationwide immune globulin (IG) shortage that has threatened Braxton’s access to care. With a true mother’s tenacity, Erica fiercely advocates for her son daily, and she is actively raising awareness about the increased need for plasma donors.

Erica Erdman’s son Braxton was diagnosed with Down syndrome and hypogammaglobulinemia, and while Braxton has not been affected by the immune globulin shortage to date, Erica is making every effort to encourage people to donate plasma.

Trudie: You describe Braxton as your miracle baby. Tell us about that.

Erica: This is a bit of a loaded question. Despite having a Down syndrome diagnosis, Braxton was born relatively healthy and only spent seven days in the neonatal intensive care unit for a couple of issues. It was at about 2 months of age that he became very ill, and from then on, we have spent countless nights away from home in and out of a hospital. The doctors kept saying it was a “Down syndrome thing,” but I knew there was something more going on, and I had to advocate for almost three years to figure it out. I knew other Down syndrome families that only had to deal with ear tubes, and we were being med-lifted to our Children’s Hospital because our local hospital could not support Braxton’s numerous complications. Braxton had several bouts of pneumonia, sinus and ear infections, upper-respiratory infections, etc., which would always land us in the hospital on significant life-support measures.

Trudie: How long did it take to get an accurate diagnosis?

Erica: It wasn’t until Braxton was almost 4 years old that we received the hypogammaglobulinemia diagnosis. I had to advocate a lot to get an immunologist referral. Even now, we are coordinating a second opinion and working with the Immune Deficiency Foundation (IDF), since it is suspected he has additional diagnoses yet to be made. I have learned so far in this journey with Braxton’s immune condition that, as a patient, you often must advocate to be heard, understood and given support to have a fighting chance at a “normal” life, which is really unfortunate. I have to be Braxton’s voice because he is nonverbal, and that adds a whole other level of difficulty to everything.

Trudie: Braxton has had numerous hospital visits. How have you coped with the stress?

Erica: The time away from home has been very difficult for our family, and I ultimately had to quit my job to stay home to care for Braxton. I guess if there was anything at all positive about the numerous hospital stays, it’s that nurses and doctors get to know you pretty fast. We have amazing relationships with those who care for Braxton when he’s hospitalized, and because he is pretty cute, it doesn’t take much for them to be wrapped around his thumb. Our hospital room often looks like home as well. For simple appointments away from home, we are very lucky to have an amazing Ronald McDonald House, and yes, they all know us too! We recently got a puppy that will be trained as a service dog to help with some medical alert items, along with anxiety during treatments, appointments and stays away from home.

Trudie: Tell us about his intravenous
IG (IVIG) treatment plan.

**Erica:** Braxton started IVIG every 28 days in January 2019. He started off with a lot of the typical side effects and ultimately struggled with IV placement since his tiny veins would blow multiple times during treatment. It was very traumatic and subcutaneous IG (SCIG) wasn’t an option due to his medical complexity. Right now, we are in a standstill with treatment because Braxton’s IgG and IgM levels are both out of reference range, along with some other white cell count issues, and he has been pretty sick but they want him to get sicker before starting back on IG therapy.

**Trudie:** When did the IVIG shortage begin to impact your son’s care?

**Erica:** We have not been told Braxton can’t receive treatment, and the Children’s Hospital pharmacy has been pretty good at ensuring it’s in stock. But, his last treatment was different and it took a bit longer since they did not prepare things until we stepped foot in the treatment center the day of his appointment. We were told they would no longer prepare things the day prior due to the shortage. Considering the likelihood of Braxton having to go back on regular IVIG treatments, my fear is, at some point, we will be one of those patients receiving a cancellation call.

**Trudie:** How are you hoping to raise awareness and encourage people to donate plasma?

**Erica:** I have an amazing friend, Ally Peters, who works at a local television station (WAOW Channel 9), in Wausau, Wisc., and she filmed a segment about Braxton’s immune condition and encouraged our community to donate plasma because of the shortage. I share a lot via social media about Braxton’s immune issues and educate people at his school. I would like to work with our local plasma center and IDF at some point, and possibly host a local awareness event like a walk. Even with this, we still have family and friends who don’t understand why we cannot go to a party held at a water park or why we haven’t had a birthday party for Braxton in three years.

**Trudie:** Do you belong to any support groups for parents of kids with immune diseases?

**Erica:** Unfortunately, there is nothing local for those with immune conditions I am aware of. I am part of several immune groups via Facebook, but none are specifically for kids.

**Trudie:** What advice would you offer other parents who are new to this challenge?

**Erica:** Do what you can to learn about the diagnosis, what lab test results mean, common symptoms, side effects, IG therapy options, everything you can. The immunology world is complex and overwhelming, and things are not the same among patients with the same diagnosis. Don’t be afraid to ask questions, and don’t be afraid to question medical professionals about their recommendations (or lack thereof). You know your child best!

**Trudie:** How do you stay positive and hopeful?

**Erica:** I stay positive and hopeful through lots of coffee! Really, the one and only thing that helps is simply Braxton himself. When I see him have good days, which are few and far between, I capitalize on them. We run around the house, we bake something, we try to learn something new, we laugh and dance like nobody is watching!

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.
PATIENT PERSPECTIVE

Breathe Deep

By Stacey Philpot

NOT LONG AGO, my husband and I moved our family across the country into a beautiful home with Brazilian hardwood floors and a master bedroom I’ve been dreaming of my entire life. It was positioned within walking distance from a large pool and playground inside the gates of a highly sought-after community. I remember describing it as “my dream home” when my husband sent me the listing, complete with a disability-friendly shower. But, in time, I realized it was not my dream home. From each of the lovely windows in my house, I gazed upon other homes, cars hurriedly rushing by, garages, enclosures and, finally, only a tiny patch of grass behind my two-story beauty and my neighbors. There was no place in my home where I could escape the sounds of cars passing by or the conversations of those tightly packed in around me. It felt oddly suffocating, and I couldn’t figure out why.

Having moved from Florida, I missed the beach, where I found solace sitting by the water on days when the pain seemed unending and questions bubbled to the surface of my soul like oxygen escaping from the lips of the frolickers ahead. I knew the sunlight through the trees at sunset grounded me in a way I hadn’t understood, bringing me back to gratitude when bitterness tried to take root. What I hadn’t known was I needed these things to breathe.

I was accustomed to many days of not being well enough to leave my home or only being well enough to sit by the water for a few moments. But here, I could not find a place of beauty accessible to me. While others could easily drive for hours for a weekend getaway or hike to a particular destination, I could not. I felt like I was withering from within. And, while this was troubling, it was also a gift. I discovered what my soul needed to breathe.

What does your soul need to breathe? Where does your pain lose its power so you can find your footing again? Where or what makes you feel most alive? Is it sitting beside a mountain? Is it taking stunning photos? Is it playing an instrument or supporting a charity? To what extent do we allow our souls to breathe deep?

Perhaps, like me, illness has had its way with some of those loves that breathe life into your soul. I can no longer go for a 3-mile run each morning and chant, “You can have the body of a hottie,” to the sound of my feet hitting the pavement. But, sometimes, I can push off the shore in a kayak and sit on the water for just a few moments, breathing ever so deeply, before I have my husband pack up the kayak for another, hopefully, stronger day. I breathe deep whenever, wherever I can.

So, we sold the dream house to some truly lovely people. We live in a gift now. It is not fancy. But every day, I step out onto my front porch, sit in a rocking chair, and my soul finds life. The view from every window is absolutely stunning. My favorite view is from my master bedroom. This view is critical, because as you know, I will spend many days here, and yet, it is healing and life-giving. What less drastic changes will we have to make in our lives to allow our souls to breathe? What is it that will bring us back to gratitude, laughter, hope, purpose, connection and our best selves? Let’s sit here for a while and breathe deep.

STACEY PHILPOT is an author, goofball and avid reader. You can find her blog at chronicallywhole.com, where she shares her journey of making the most of a life touched by common variable immunodeficiency, Lyme disease and rheumatoid arthritis.
Taking a Break from My Disease: An Experiment of Extreme Measures

By Ilana Jacqueline

AFTER YEARS of really struggling with managing my immune deficiency, I’d finally had enough. At the start of 2019, I was exhausted. I couldn’t remember what life was like without taking antibiotics every day and dealing with their side effects, yet still getting one new infection after another. It was weighing more on my mental health than my physical health. I had forgotten what it felt like to be comfortable in my own body. I just wanted to take a step back from all of it.

So, I did. I went on a drastic lockdown. I’m talking a very intentional quarantine from everyone and everything nonessential that added risk to my daily life. I was already working from home, so having a daily commute or office where I had to interact with people was not an issue. I went off the grid with friends and family, seeing only my mom and husband. I used apps to order food and groceries to avoid crowded stores. I shopped for all my clothes on Amazon. If I had to go to a store for anything, I did my best to go during “quiet” hours at 24-hour locations. I switched to a pharmacy with a drive-through window. I cancelled all nonessential doctor appointments. If a waiting room was busy, I’d hand the front desk clerk my phone number and ask for a call when they were ready for me. I walked empty hallways to avoid sitting next to people. All in all, avoiding the entire world wasn’t that hard. It was a lot easier than living with a constant head cold. At the end of 2019, I’d gone my first full year without a hospitalization.

What did it cost?

Literally all of my friendships. I wish I could say I stayed in contact with even one friend during my entire quarantine, but I didn’t. I didn’t go to parties. I didn’t celebrate girlfriends’ birthdays. I didn’t spend nights out. It was lonely, and there were some days I wondered whether it was worth it. But, I just kept reminding myself this season of my life wasn’t forever — and my body needed the break.

A lot of money. Think staying home is cheap? Not in today’s world. I paid through the nose to tip delivery drivers, subscribe to delivery services and cover shipping fees for things that wouldn’t have cost me half as much to pick up on my own.

My social skills. Now that I’m rejoining society, I feel the most awkward I’ve ever felt having conversations with people. I spent a lot of alone time. Even though I had plenty of virtual meetings for work, I lost all the ease I used to have when talking to strangers.

I’m having to relearn how to be myself around other people.

And what did I gain?

I caught up on my medical debt. That’s right! The hospital even wrote me a check for accidentally overcharging my tab after I paid off the last of my hospital stays. Without incurring any new visits, I even had its billing office perplexed!

I reduced my medication. I was able to stop taking antibiotics, which meant much less damage to my stomach. In peacetime, I also decided it was safe to stop taking some of my pain medications.

Appreciation for so many things. My body’s ability to heal. My ingenuity when it came to finding creative ways to avoid risk. My husband and mother who helped me do the things I couldn’t. A brand-new awareness for how much we actually need people in our lives, no matter how much of a risk they might present.

While a quarantine may not be a great experiment for everyone, it is possible with planning and perseverance, and it can do wonders to give your immune system a rest!
Summer Camps for Chronically Ill Children

By Jessica Leigh Johnson

With summer just around the corner, many kids and their families are eagerly making plans. It’s amazing how quickly the calendar fills up with family vacations, summer baseball leagues and day trips to beaches or amusement parks. Another favorite summer activity for kids is summer camp. But, is it a safe choice for children with a chronic illness such as primary immunodeficiency disease (PI)?

As the mother of three children with PI, I know full well the anxiety that can accompany sending children with chronic illness to a summer camp for a week. But, I also know it can be one of the most fun and rewarding experiences for them. My three boys have been attending the same summer camp every year for the past nine years, and it is always a highlight of their summer. With a few extra precautions and some detailed planning, there is no reason kids with chronic conditions should have to miss an amazing experience like summer camp.

Choosing the Right Camp

After parents have made the decision to send their children to summer camp, the next step is to choose which one they’ll attend. Some camps focus on activities such as wakeboarding, skiing, art or science. Others are faith-based camps. Since there are so many choices, the best place to start is the American Camp Association’s (ACA) website. From there, parents can narrow down their choices by selecting which state they live in, which type of camp they’re interested in (day or overnight) and if their child has any special needs or disabilities such as chronic illness. For a camp to be ACA-accredited, it must meet stringent health requirements. ACA lists camps that specialize in serving campers with certain conditions, as well as those that serve campers exclusively with specific conditions.

Keeping Kids Healthy During Camp

The biggest concern for parents who send their chronically ill kids to overnight camp is: “What happens if they get sick?” Hopefully, with camps occurring during the summer months, there are fewer germs circulating. But, certain illnesses such as strep and measles occur year-round. So, for peace of mind, there are some steps parents can take to ensure their children’s health is monitored while camp is in session.

• Make sure the camp has a first-aid plan. Camp staff should know how far they are from medical facilities and whether the hospital is a major trauma center. Medical release forms should be filled out before the children’s first
day at camp, allowing camp staff to transport children to the hospital in case of acute illness or injury.

- Choose camps with medical staff on-site. Some camps staff a full-time physician or nurse, while others have volunteer nurses who stay only for the week. The doctor or nurse should be made aware of the children’s condition, as well as which medications they take and when they should be taken. If children need to take medication before bed, will the nurse still be on duty? If not, other arrangements must be made. If the children’s medications need to be refrigerated, there should be a designated, safe storage area for them. Be sure several of the camp staff — not just the nurse — are trained to respond to medical emergencies such as seizures or severe allergic reactions to food or bee stings. And, make sure the selected camp is able to accommodate IV medications, if necessary.

- The children’s cabin counselor should also be made aware of any health issues or concerns. It’s OK for parents to make special requests such as asking to be notified if other campers in the room/cabin become ill, or for the children to be removed from a cabin where another camper has become ill.

- Parents should ask about the chaperone-to-camper ratio so they can be sure their children will receive the attention they need. Children will receive more individual attention in a program with one adult for every five campers than at a program with one adult for every 15. For children with a chronic illness, a closer adult-to-camper ratio will ensure changes in health don’t go unnoticed.

- It’s a good idea for parents to call their health insurance company to ask whether their children’s insurance is accepted at other hospitals, and what requirements exist should their children need to be seen by a doctor while at camp.

- If children require a special diet, make sure the camp can provide it. If not, ask if any special foods/meals can be brought to camp with the children and prepared for them by kitchen staff.

- While there are fewer viruses circulating in the summer, illnesses such as West Nile virus and Lyme disease, which are spread by insects, are at their peak. Keep disease-spreading bugs away by treating the camper’s clothing with permethrin, which kills ticks and mosquitoes on contact. Follow the instructions for use precisely for the best outcome.

Communicating with Children During Camp

While it’s best for kids to experience camp without too much parental interference, there may be times when communication is necessary. If children will be attending a daytime-only program, parents will have the chance to speak with counselors at drop-off and pick-up times. For overnight camp, parents should inquire if staff members will be readily available by phone or email. Also, parents might want to find out how often they will be able to speak with their children. Some camps have strict guidelines about campers contacting their parents. While this policy helps campers stay focused on their activities, it can be scary for parents of kids with chronic illness. It is important to figure out ahead of time how to get information about the children’s status. It’s OK to request that the camp director, nurse or counselor call parents with updates. Exceptions to the rules can usually be made for chronically ill children, provided there is a plan in place ahead of time.

Day Camps

Depending on the specific health needs of children, an overnight camp may be out of the question. In situations like this, most local school districts, YMCAs or parks and recreation departments offer daytime-only programs for kids according to their specific interests and abilities. Sports camps, engineering camps, art camps and outdoor nature discovery camps are among the many programs offered to children in many communities across the country.

Have a Plan in Place

No matter what camp a family chooses, the opportunity to spend a few days away from home sharing adventures with other children is an experience chronically ill children shouldn’t have to miss out on. Camp is a great opportunity to take a break from medical appointments and therapy sessions, and a time to enjoy just being a kid. If something should go wrong, having a plan in place for such an occurrence will eliminate confusion and anxiety for parents, staff and children. For those who are well-prepared, the week should go by without incident, and the children will return home very “happy campers.”

JESSICA LEIGH JOHNSON is a stay-at-home mom and mother of four kids, three of whom have X-linked agamma-globulinemia. 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


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Move More in the New Year

By Heather Bremner Claverie

THE SIGNS encouraging people to exercise more in the new year are everywhere: Get stronger. Get fitter. Lose weight. But what about individuals with chronic illnesses receiving weekly or monthly treatments such as immune globulin (IG) infusions? Can these people safely join in those sweaty sessions? The quick answer is yes; even moderate amounts of exercise can soothe the body and mind.

Magical Medicine
It’s no secret that pumping iron, jogging or riding a bike can help tone tummies, trim waistlines and improve cardiovascular health. What’s more, studies show regular exercise can also reduce stress and depression and even diminish fatigue and anxiety. Research has even revealed exercise is as effective as antidepressant medication in treating depression. One study conducted by the Harvard T.H. Chan School of Public Health discovered just a 15-minute daily run or walk reduces the risk of major depression by 26 percent.

Reduce Anxiety and Stress
Since patients treated with IG have weakened immune systems, they are particularly vulnerable to stress and anxiety. Too much stress can cause a domino effect, leading to bouts of illness that can last for weeks or months. Thankfully, a little exercise can go a long way to counteract this scenario. It can be anything from a sweaty spin session to a walk around the block or a round of sun salutations in a local yoga class.

The other great benefit of exercise is it can be done anywhere at any time. Any sort of movement that gets the heart pumping and the muscles moving helps. And, these days, with access to online or on-demand exercise classes and home-based products such as the Peloton bike, there are a myriad of options available to all.

For those who don’t have the budget to join a gym or pay for Pilates classes, exercising doesn’t have to cost a cent. Taking a walk around the neighborhood or hiking through a local nature preserve is a free and effective way to reap the benefits of exercise. And, since subcutaneous IG therapy patients can receive infusions at home without supervision, home-based exercise is a great option.

Ward Off Weariness
Fatigue is an issue many IG patients wrestle with during and after treatments. So, something as simple as getting the blood and muscles moving can help infuse energy into anyone’s day. In addition, cramping and muscle aches can be alleviated with simple stretching routines. And, a yoga class can address an array of issues, too, since the practice helps center the mind, body and soul.

The Magical Pill
Humans are constantly searching for that magical pill, that cure-all elixir — something that can ward off fatigue, reduce anxiety and depression, increase life expectancy, melt those love handles and make us happier beings. Well, it seems we have already found it!

HEATHER BREMNER CLAVERIE is a contributing writer for IG Living magazine.
Roll Out Tranquility

Designed by yogis, the long-lasting Manduka yoga mats are available in a rainbow of colors, styles and prices. Individuals who haven’t practiced many downward dogs can hop on a wallet-friendly beginner mat, while long-time yogis may opt for one of the Pro Series versions. The company also offers a special mat for hot yoga devotees. An online quiz will help people locate which mat is the best one for them. $37 and up; www.manduka.com/pages/collections-yoga-mats

A Vision of Fitness

Without leaving the comfort of home, less than two feet of wall space can be converted into a personal fitness studio. While it looks like a simple, elegant mirror, the Mirror serves as much more. It’s a cardio class, a yoga studio or a boxing ring. The Mirror can be easily installed in any room in the home. With more than 10,000 on-demand classes and more than 20 types of workouts ranging from kickboxing to yoga, there’s something for everyone. $1,495 or $42/month; www.mirror.co

Shopping Guide to Exercise Products and Accessories

Rein ’em In

Brooks sports bras help minimize bounce and discomfort, allowing women to enjoy high-impact sports from running to tennis. With features like odor-resistant material, lightweight perforated cups and adjustable straps, these bras give supportive shape while keeping everything in place. They’re also available in cup sizes up to E. $34 and up; www.titlenine.com/category/title-nine-brand-partners/brooks-sports-bras.do

Sweat to Your Own Beat

The Peloton bike has become the ambassador of sorts for home exercise equipment, turning that image of the dust-covered bike into a relic. With on-demand classes, a slim profile and a price tag that rivals the cost at brick-and-mortar facilities, this smart bike delivers the convenience of a 24-hour gym. $2,245 or $58/month; www.onepelotono.com/bike

Sweat in Style

Comfort, durability and performance are all essential when it comes to workout clothes, and with Lululemon’s full line of gear, it’s possible to hit all those targets. From high-rise yoga leggings to sweat-wicking running shorts, the Canadian-based athletic apparel company has something for every exercise enthusiast. shop.lululemon.com

Pump Some Iron

The AmazonBasics Neoprene Dumbbell Set and Stand is ideal for resistance training. Available in a variety of sizes and colors, the set also includes a stand to neatly stack the weights. The neoprene coating allows for a secure grip, and the weights are made for both indoor and outdoor use. $28.49; www.amazon.com/AmazonBasics-20-Pound-Dumbbell-Lettering-Renewed/ dp/ Neoprene+Dumbbell+Set+and+Stand
Women’s Health in Autoimmune Diseases
Author: Shefali Khanna Sharma
Publisher: Springer

This book focuses on conveying autoimmune disease expertise to gynecologists and other clinicians to help them approach the treatment of each disease in a pragmatic manner. Each chapter reviews the current literature on treatments for autoimmune diseases, especially under special circumstances such as pregnancy, rating disease severity and providing practical guidelines based on the current state of knowledge. How autoimmune diseases affect fertility, and how to best prepare patients with these diseases for pregnancy is also addressed. In addition, the book explores important issues concerning autoimmune diseases such as lupus nephritis, vasculitis, Sjogren’s syndrome, antiphospholipid syndrome and systemic sclerosis in women and their potential effects on unborn children.

Taking Back Your Health and Happiness: Hope and Healing from Chronic Pain, Fatigue, and Invisible Illness
Author: Marie Anne June L. Tagorda
Publisher: Morgan James Publishing

This book is written to help those who suffer from chronic pain discover the source of their pain to achieve self-healing and happiness. As a nurse for more than 10 years, Marie Anne June L. Tagorda has seen the effects of illness on people and their loved ones. And, having had her share of chronic pain and invisible illness, she shares her process for achieving self-healing and happiness by helping readers learn the source and meaning of their chronic pain and illness, how to deal with their condition, and how to effectively communicate with loved ones to include them in their care and not be burdened by it.

Living a Healthy Life with Chronic Conditions: Self-Management Skills for Heart Disease, Arthritis, Diabetes, Depression, Asthma, Bronchitis, Emphysema and Other Physical and Mental Health Conditions
Authors: Kate Lorig, DrPH, David Sobel, MD, MPH, Diana Laurent, MPH, Marion Minor, PT, PhD, Virginia Gonzalez, MPH, and Maureen Gcht-Silver, OTD, MPH
Publisher: Bull Publishing Company

The goal of this book is to help people with chronic illness explore healthy ways to live with physical or mental conditions. By showing readers how to become active self-managers through problem solving, goal-setting and action planning, while also presenting the basics of healthy eating, exercise, relaxation and emotional empowerment, Living a Healthy Life with Chronic Conditions offers readers the chance to take back one’s life and enjoy it to the fullest extent possible while living with chronic illness.

Children’s Health and Illness Recovery Program (CHIRP): Clinician Guide (Programs That Work)
Authors: Bryan D. Carter, William G. Kronenberger, PhD, Eric L. Scott, PhD, and Christine E. Brady, PhD
Publisher: Oxford University Press

Derived from six decades of combined experience from the authors, CHIRP is an interdisciplinary cognitive behavioral and family systems-based treatment program designed to maximize the independent functioning of teens with chronic illness. The guide is a detailed outline for implementing this manualized treatment protocol over the course of 12 sessions and provides clear guidance on the philosophy, pragmatics and art of working with this challenging pediatric population. It is designed to accompany the CHIRP Teen and Family Workbook.
Download the IG Living eBook today—now available for iPad, Nook and Kindle!

“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
### Ataxia Telangiectasia (A-T)
- **WEBSITES**
  - A-T Children’s Project: www.atcp.org

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

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

### Guillain–Barré Syndrome (GBS)
- **WEBSITES**
  - GBS/CIDP Foundation International: www.gbs-cidp.org
  - The Foundation for Peripheral Neuropathy: www.foundationforpn.com
- **ONLINE PEER SUPPORT**
  - GBS Support Group: www.gaincharity.org.uk
  - GBS/CIDP Foundation International Discussion Forums: forum.gbs-cidp.org/forum/main-forum

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

### Kawasaki Disease
- **WEBSITES**
  - American Heart Association: www.heart.org/HEARTORG/Conditions/More/ CardiovascularConditionsofChildhood/Kawasaki-Disease_UCM_308777_Article.jsp#.T1T2boePWE0
  - Kawasaki Disease Foundation: www.kawasaki.org
  - KidsHealth: www.kidshealth.org/parent/medical/heart/kawasaki.html

### Mitochondrial Disease
- **WEBSITES**
  - United Mitochondrial Disease Foundation: www.umdf.org
  - Mitochondrial Association: www.mitoaction.org

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

### Multiple Sclerosis (MS)
- **WEBSITES**
  - All About Multiple Sclerosis: www.mult-sclerosis.org/index.html
  - Multiple Sclerosis Association of America: mymsaa.org
  - Multiple Sclerosis Foundation: www.msfocus.org
  - National Multiple Sclerosis Society: www.nationalmsociety.org
- **ONLINE PEER SUPPORT**
  - Friends with MS: www.FriendsWithMS.com
  - MS World’s Chat and Message Board: www.msworld.org

### Myasthenia Gravis (MG)
- **WEBSITES AND CHAT ROOMS**
  - Myasthenia Gravis Foundation of America (MGFA): www.myasthenia.org
  - Genetic Alliance: www.geneticalliance.org

### Myositis
- **WEBSITES**
  - The Myositis Association: www.myositis.org
  - International Myositis Assessment and Clinical Studies Group: www.niehs.nih.gov/research/resources/imacs
- **ONLINE PEER SUPPORT**
  - The Cure JM Foundation: www.curejm.org
  - Myositis Association Community Forum: tmcommunityforum.ning.com
  - Myositis Support Group – UK: www.myositis.org.uk

### Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)
- **WEBSITES**
  - PANDAS/PANS Advocacy and Support: www.pas.care
  - PANDAS Network: www.pandasnetwork.org
  - Midwest PANS/PANDAS Support Group: www.midwestpandas.com

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

### Peripheral Neuropathy (PN)
- **WEBSITES**
  - Neuropathy Action Foundation: www.neuropathyaction.org
  - Western Neuropathy Association: www.pnhelp.org
  - Neuropathy Alliance of Texas: neuropathyalliance.org
  - The Foundation for Peripheral Neuropathy: www.foundationforpn.com

### Primary Immune Deficiency Disease (PI)
- **WEBSITES**
  - Immune Deficiency Foundation: www.primaryimmunedeficiency.org
  - Jeffrey Modell Foundation: www.info4pi.org
  - The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/Pages/index.aspx
  - American Academy of Allergy, Asthma & Immunology: www.aaaai.org
  - International Patient Organisation for Primary Immunodeficiencies (IPOD) — UK: www.ipodi.org
  - New England Primary Immunodeficiency Network: www.nepin.org
  - Rainbow Allergy-Immunology: www.uhospitals.org/rainbow/services/allergy-immunology
- **ONLINE PEER SUPPORT**
  - IDF Friends: www.idffriends.com
  - Jeffrey Modell Foundation Facebook Page: www.facebook.com/JMFworld
  - IDF Peer Support Program: www.primaryimmunedeficiency.org/idf-peer-support-program
  - Michigan Immunodeficiency Foundation: www.michigan-immunodeficiency-foundation-monroe

### Scleroderma
- **WEBSITES**
  - Scleroderma Foundation: www.scleroderma.org
  - Scleroderma Research Foundation: www.srfcure.org
  - Johns Hopkins Scleroderma Center: www.hopkinsscleroderma.org
- **ONLINE PEER SUPPORT**
  - International Scleroderma Network: www.sclero.org/support/forums/a-to-z.html

### Stiff Person Syndrome (SPS)
- **WEBSITES**
  - Scleroderma Foundation: www.sclero.org
  - Scleroderma Research Foundation: www.srfcure.org
  - Johns Hopkins Scleroderma Center: www.hopkinsscleroderma.org
  - Stiff Person Syndrome: www.stiffpersonsociety.net
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