Chronic Illness Benefits
Learning the Positives in Life’s Lessons

How to Discuss End-of-Life Wishes

A Conditioning Program for Building Physical Endurance

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Benefiting from the Positives of Illness

IT MAY SEEM there are only downsides to chronic illness, but those affected by the battles of long-term health conditions often become wiser and stronger people. It’s about learning the positives in the lessons that life hands us each day and putting to use the knowledge we have gained and tools available to us.

In her article, “The Benefits of Having a Chronic Illness” (p.18), Surayyah Morris, an autoimmune disease patient and medication therapy and pain management specialty pharmacist, shares what she has gained from having a chronic condition. Specifically, she outlines seven benefits: 1) learning how to be present in your care and how to advocate for yourself; 2) becoming more compassionate toward others due to your own experiences; 3) discovering the easiest and least-taxing ways to get things accomplished; 4) remaining observant about how caregivers administer treatment to better protect yourself; 5) developing more patience from dealing with time-consuming medical demands; 6) prioritizing what’s most important to accomplish when caring for yourself; and 7) recognizing you are stronger than most people in your life. As Morris explains, those with chronic illness need to give themselves credit about how much they have learned from their journey.

To assist with benefit number three, there are a myriad tools available to help patients deal with their condition and make their lives easier. After a lifetime of dealing with PI and many other chronic illnesses, Ilana Jacqueline (also our 20-Something columnist) shares with us in this first-in-a-series special feature “New Tools and Tech for Patients” (p.24) some of the best new products she has researched to help herself and others in need of pain relief, comfort and safety. In all, she features 16 products for fevers and staying cool, wearable health, health technology, and nausea and pain relief. Our readers are sure to find many of these items very useful to include in your personal healthcare supply arsenal.

Beyond tools, it takes a wise and strong person to discuss the inevitable with family and friends. While most people delay end-of-life discussions, patients are in a unique position to make preparations sooner than later since their illness opens the door to dialogue. In our article “How to Talk Plainly About End-of-Life Wishes” (p.32), we discuss how making preparations for the final days will ensure everyone is on the same page about what kind of care is wanted and all family members needs are fulfilled. We provide helpful tips about how to break the ice about the topic, begin the discussion, items to discuss and what to do when met with resistance.

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.
What Patients Need to Know About Plasma

By Abbie Cornett

IN RECENT YEARS, the demand for plasma protein therapies used to treat rare and chronic conditions has grown at a tremendous rate. The worldwide demand for intravenous immune globulin (IVIG) and subcutaneous IG (SCIG) more than doubled between 2008 and 2016, and it is projected to continue growing at more than 8 percent per year. To keep ahead of this projected growth, manufacturers must expand collection of plasma, which is needed to produce IG products. In 2018, it was projected an additional three million plasma donations were needed to meet demand.

The U.S. Polyvalent IG Market (IVIG/SCIG) from 1986 to 2016

Patients may be surprised to learn it takes between six months and 12 months from the time plasma is donated until it is manufactured into an available IG product. This is because the production of a plasma therapy is a complex multistep process.

Unlike traditional pharmaceutical products that rely on a chemical process, plasma-based therapies rely exclusively on proteins found in human blood plasma. Plasma is the straw-colored liquid portion of blood made up of water, salts and proteins that performs a variety of functions in the body, including clotting and fighting disease. It makes up 55 percent of a person’s total blood volume.

The first step in producing a plasma therapy takes place in a donation center where donors are carefully screened to ensure the quality of their plasma. Once vetted, these individuals donate their plasma through a specialized process called plasmapheresis that separates the plasma from red blood cells and other cellular components that are then returned to the donor. Each individual plasma donation averages about two-thirds of a liter in volume. After an individual’s plasma is collected, it is stored while it undergoes screening to guarantee its quality and safety. The plasma is then pooled with donations from thousands of other donors.

The next step is fractionation, a process that isolates and purifies therapeutic proteins found in the plasma, including those used in the production of IG products. After these steps have been completed, the proteins are ready to be formulated into therapies.

Because of increasing demand and the long lead time needed to bring these products to market, there can be shortages at times. If you are a patient who has been told future treatments with your brand of IG will not be possible, here are some steps you can take:

1) If you are being treated in the home, ask if there is another brand to which you can switch. If no product is available, talk to your ordering physician about another homecare provider.

2) If you are being treated in a physician’s office, ask if there is another brand to which you can switch. If no product is available, look for other sites of care. Specifically, check with the hospital where your ordering physician has privileges. Or, check with a homecare provider.

3) If you are being treated in a hospital outpatient infusion center, go back to your ordering physician and ask about another outpatient center. Or, check with a homecare provider.

4) Consider exploring the subcutaneous route of infusion if it makes sense for your condition.

IG manufacturers are making every effort to address these issues affecting shortages. In fact, every major plasma fractionator is investing in new production capacity, and manufacturers are producing IG products at top capacity. In addition, some new products are coming to market, and one previously removed from the market is being reintroduced.

As the patient advocate for IG Living, I am ready to answer your questions about IG therapy, as well as your concerns if you have been told your product is not available.

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 you seem to get bitten more often than other people?

Yes. I grew up in Alberta and always got bitten by mosquitoes. I moved to Vancouver, Canada, where there are few mosquitoes. I still get bit on occasion, but more often than everyone else. In Alberta, my bites were about the size of half of a pencil eraser. In Vancouver, I have developed an allergy to mosquitoes, with bites that are huge, itchy and painful. IVIG makes no difference. Neither do any other medicines.
— Connie K

More than other people, yes. But, my kids and I swell up a lot and usually need medicine to help the swelling go down. My brother almost died from fire ants.
— Stacey MT

My husband joked and said the reason they are attracted to me is because of all the flavors from the plasma I receive weekly. I’m the only one attacked when I’m outside with my family.
— Debbie B

Yes! But while I was receiving Rituxan to treat my lymphoma, I rarely got bit!
— Debbie K

Has opioid legislation impacted you?

I don’t use any opioids, but many pharmacies and states are limiting prescriptions, so my local pharmacy will not fill more than one prescription for Lyrica because it is listed as a controlled substance. Mail order will fulfill a 90-day supply. I have been on Lyrica a long time, and this is a recent change.
— Lisa P-S

One thing for sure with a chronic disease like mine and infusions every three weeks, there is no way I’m going into oncology without pain killers. I go off to the corner to my local drive-by dealer. I know, sooner or later, the street drugs will kill me if the disease doesn’t first. Why can’t the government understand what pain is all about? Patients with a verified chronic disease deserve to live pain-free without having to turn to drug dealers.
— Peggy Z

Have you been hurt by people’s comments?

I most certainly have. People who are obviously not on board with what goes on in my life, especially my health, I try to ignore. When it happens with someone who is either family or a very good friend, it tends to sting. Then, I remind myself they have to be as frustrated and worried as me from time to time. They do not have to understand my issues, because a lot of the time, I do not understand them either. All I want or need is love, support and encouragement when things are tough. Just knowing there is a support system out there is a gift that cannot be bought.
— Jenny G

Yes. The most hurtful, however, have been comments regarding my daughter’s health. People say I overreact, exaggerate, make things up. She has common variable immune deficiency, and has just started intravenous immune globulin. It’s most hurtful when it comes from family.
— Amanda T
**Question** » What do I do if I am told my brand of IVIG is unavailable due to the nationwide shortage?

*My mother is treated with intravenous immune globulin (IVIG) every month, and she gets very ill if she misses her medicine. I am concerned about the nationwide shortage of IVIG, and I would like to know what steps to take if she is denied treatment.*

**Abbie** » If you are a patient who has been told future treatments with your brand of IVIG will not be possible, here are some steps you can take:
1) If being treated in the home, ask if there is another brand to which you can switch. If no product is available, talk to your ordering physician about another homecare provider.
2) If being treated in a physician’s office, ask if there is another brand to which you can switch. If no product is available, look for other sites of care. Specifically, check with the hospital where your ordering physician has privileges. Or, check with a homecare provider.
3) If being treated in a hospital outpatient infusion center, go back to your ordering physician and ask about another outpatient center. Or, check with a homecare provider.
4) Consider exploring the subcutaneous route of infusion if it makes sense for your condition.

**Question** » What can be done to reduce the side effects of IVIG to treat myasthenia gravis (MG)?

*I have MG, and for the last decade, I’ve been treated with intravenous immune globulin (IVIG) infusions every three to four weeks, after which I experience severe headaches and nausea about 48 hours after the infusions. My new neurologist prescribed acetazolamide (Diamox) starting four days prior and three days after infusions, as well as Depakote (divalproex sodium) for seven days starting the day of infusion. Although my last infusion resulted in a 20 percent decrease in headaches and nausea with this protocol, the headaches still required injections of 30 ml of ketorolac, 50 mg/ml of promethazine (Phenergan) and long-acting lidocaine scalp/eyebrow injections. Additionally, the infusions have been slowed during the last three months to 90, and my neurologist has added Benadryl, Decadron and Zofran prior to starting the infusions. My next infusion is in 11 days, and while the intense headache has subsided, I am still experiencing malaise and brain fog, which makes it very challenging for me to work. These symptoms are also taking a significant toll on the MG; my symptoms are worse than they’ve been in more than five years.

My neurologist says I am experiencing aseptic meningitis, and she is taking steps to change my brand of IVIG. Do you have any other suggestions? Are there other drugs that could be used to treat the symptoms?*

**Abbie** » I spoke with Leslie Vaughan, senior vice president of clinical programs at Nufactor, a specialty infusion company, regarding your question. To reduce your symptoms, she suggests asking your physician to switch brands of your IVIG or slow the rate of your infusion even further. According to Vaughan, some patients can’t tolerate even a rate of 90. She also recommended infusing a large dose over several days or infusing on nonconsecutive days. Lastly, she suggests you ask your physician if he or she would consider you a candidate for Soliris, which is approved for MG and has a much lower side effect profile than IVIG.

**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.
Blood Testing Issues for PI Patients, Part 4

By Terry O. Harville, MD, PhD

NUCLEIC ACID TESTING (NAT) has become the modern assay for identifying microorganisms that may be the cause of an infection. With NAT, a sample in the form of saliva, sputum or lung fluid, nasal secretions, blood, urine, cerebral spinal fluid, pus from an infection site or stool is obtained from the patient. As long as DNA (or RNA for certain viruses) can be extracted from the specimen, the assay can be performed.

NAT generally uses a procedure known as polymerase chain reaction (PCR). PCR uses known sequences of DNA from the microorganism in question. Primers are manufactured using the sequence information that makes them specific to the microorganism. Thus, each microorganism in question will generally have its own unique primer set, which enables PCR to identify the specific microorganisms since primers are able to bind only to the sequence of DNA from the specific microorganism.

After obtaining a specimen, it can take less than three hours to explicitly identify which microorganism is causing the infectious symptoms. This rapid identification determines the best course of therapy that will result in overall better outcomes. Therefore, NAT seems ideal for primary immunodeficiency disease (PI) patients because it provides early and specific identification of the infectious microorganism, as well as earlier introduction of the right course of treatment. This is especially ideal for PI patients when on replacement immune globulin (IG) therapy since serum antibodies are not required to make the diagnosis.

With NAT, however, the physician needs to have a reasonable idea about which microorganisms to look for to be able to test for, detect and correct the offending pathogen. This is the major drawback with NAT. If the physician does not identify the correct reagents for the assay because he or she did not suspect the correct microorganism, the source of the infection will not be detected.

For PI patients, unusual or opportunistic infections may be causing the symptoms. Therefore, the physician should first look for the common causes of the infection, but he or she should also evaluate infectious opportunistic microorganisms that cause problems in PI patients.

In summary, PI patients may not be capable of making antibodies to infectious microorganisms. Because of this, the routine assays for detecting infections using antibodies are unreliable (they can result in a false negative). Further, patients receiving IG replacement therapy are receiving antibodies from the donors of the plasma used to manufacture the IG. Therefore, a test may detect the presence of antibodies to microorganisms with which the patient has never been infected. And, this may result in incorrectly identifying a microorganism as the one causing the infection (resulting in a false positive). Therefore, for PI patients and those receiving IG replacement therapy, special considerations for testing need to be in place to prevent false negative and false positive results.

We will continue with a new topic 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.
LIVING WITH A chronic illness means experiencing unwanted, unpleasant and uncomfortable feelings on a regular basis. For some of us, it’s anxiety, worry or fear about how our illness will change over time, or about whether we will have a “good” or “bad” day when we make plans. For others, it’s feelings of sadness, grief or disappointment about the things we can no longer do or about how our lives have forever changed. For still others, it’s anger or frustration toward our illness, pain or a higher power we may blame for our illness. We may experience all of these feelings on different days, along with occasional shame and guilt.

Think about some of the feelings you find most unpleasant or distressing, the ones that seem to have the most power over you. What have you tried to get rid of them, change them or control them? These are called “away” moves because they provide distraction from unwanted, unpleasant and uncomfortable feelings. We all engage in away moves at times. These range from sleeping, watching TV, browsing the Internet, listening to music, reading, exercising, shopping online and eating junk food, to yelling at/lashing out at others, gambling, drinking or taking other drugs, or physically hurting ourselves.

Which away moves do you engage in the most? Do they make the unwanted feelings go away? If the answer is yes, then you engage in these behaviors because they provide distraction from unwanted, unpleasant and uncomfortable feelings. We all engage in away moves at times. These range from sleeping, watching TV, browsing the Internet, listening to music, reading, exercising, shopping online and eating junk food, to yelling at/lashing out at others, gambling, drinking or taking other drugs, or physically hurting ourselves.

What are these away moves? Do they make the unwanted feelings go away? If the answer is yes, then you engage in these behaviors because they provide distraction from unwanted, unpleasant and uncomfortable feelings. We all engage in away moves at times. These range from sleeping, watching TV, browsing the Internet, listening to music, reading, exercising, shopping online and eating junk food, to yelling at/lashing out at others, gambling, drinking or taking other drugs, or physically hurting ourselves.

Which away moves do you engage in the most? Do they make the unwanted feelings go away? If the answer is yes, then you engage in these behaviors because they provide distraction from unwanted, unpleasant and uncomfortable feelings. We all engage in away moves at times. These range from sleeping, watching TV, browsing the Internet, listening to music, reading, exercising, shopping online and eating junk food, to yelling at/lashing out at others, gambling, drinking or taking other drugs, or physically hurting ourselves.

These acts tend to be labeled as good vs. bad, right vs. wrong or positive vs. negative. But, I encourage you to move away from judging your behaviors in this way. Instead, think about these acts as helpful or unhelpful. Why did you choose that behavior? What was the purpose for that behavior? Was it to help get rid of an uncomfortable feeling? Or, was it to move you toward something that matters to you?

For most of us, the away moves may help the feelings go away or lessen them temporarily, and there may be no downside to using them. If that is the case, great! Keep using them. However, for some of us, these away moves do not help. Instead, the away moves make the feelings come back as soon as we stop distracting ourselves, and we can get caught in a cycle of anxiety, distraction, anxiety, distraction, and around and around we go.

Some of these away moves may also have consequences or costs. Ask yourself the following questions: Are there any consequences to engaging in these behaviors? Do they cost a lot of money? Do they lead to problems in relationships or at work? Do they impact health? If the answer is yes to any of these questions, the away moves may not be helpful, and it may be time to consider a different way to manage the unpleasant feelings.

Noticing why we do what we do is a key step toward reducing the influence our feelings have on our behaviors. If these away moves are helping, keep doing them! If they are not, and they are causing additional problems, it may be time to think about alternative ways of responding to these uncomfortable feelings. The alternative is to be willing to experience the feelings, instead of spending all of your time and energy trying to control, change or get rid of them. Try to allow the feelings to be there while you continue living your life. Over time, they will have less power over you.

Of course, this is much easier said than done, but the bottom line is: We can’t control our feelings. We can only control how we respond to them and how much we let them run our lives.

There are hundreds of willingness or acceptance strategies to try for free on contextualscience.org (you may need to join) or by Googling willingness or emotional acceptance techniques.

ERIKA LAWRENCE, PhD, LCP, is director of translational science at The Family Institute at Northwestern University, Evanston, Ill.
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How Insurance Authorizes IG Treatment

By Michelle Greer, RN

OVER THE YEARS, health insurance has drastically changed how it approves payment for immune globulin (IG) therapy. Most of today’s plans not only require approval prior to treatment, but the diagnosis requiring treatment must be proven.

Insurance Medical Policies and Approval Times

Almost all health insurance plans have a medical policy that outlines the approval process for intravenous IG (IVIG) and subcutaneous IG (SCIG) therapy for both U.S. Food and Drug Administration (FDA)-approved and non-FDA-approved indications. While authorization can differ depending on the site of care (hospital outpatient setting, physician office or home), the medical policy applies to the approval and payment of treatment regardless of where care is rendered. To avoid any patient misunderstanding, the IG therapy provider should explain coverage and any patient responsibility up front.

It must be note that differences in medical policies among health plans can be frustrating when coverage is changed for any reason. However, since most health plans post their medical policies on their websites, any provider in or out of network can access these policies to ensure patient treatment meets the plan’s criteria.

Medical polices are written by a committee based on clinical trial and other study literature showing the efficacy of treatment. However, because conditions treated with IG are rare, there is sparse consistent data showing treatment efficacy. And, while most FDA- and non-FDA-approved indications are clear, as the use of IG therapy continues to expand, it is difficult to get approval for other less-documented indications.

The approval process timeline also varies between plans (Table 1). Depending on the plan and the clinical documentation needed to receive authorization, the process can take days or even weeks. If the situation is urgent or life-threatening, an expedited authorization process can be requested.

IG Approval Requirements

For IG to be approved for treating a primary immune deficiency (PI), three qualifiers are required by all plans: 1) a low immune globulin (IgG) level, 2) failure to mount a response to vaccination and 3) history of infection. Citing a low IgG and a history of infections is not enough. There must be proof the immune system cannot mount a response to pneumococcal pneumonia, diphtheria and/or tetanus vaccinations. All brands of IVIG and SCIG have labeled indications for PI, and all plans’ medical policies outline how IG for PI is covered. However, there are differences among plans for how IG is covered. Some plans will state the IgG level must be below a certain level, but those levels vary from plan to plan as well. Other plans require two IgG levels to be low. These specifics are outlined in the plans’ policies.

For a secondary immune deficiency as seen in certain blood cell cancers such as chronic lymphocytic leukemia (CLL), the vaccination requirement is not necessary. CLL is an FDA-approved diagnosis for only one brand of IVIG. However, all medical policies address CLL, and some address additional blood cell cancers such as multiple myeloma.

Medical policies also address IG treatment for autoimmune disorders.

Table 1. IG Approval Process Timeline

<table>
<thead>
<tr>
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<th>Immune globulin therapy is ordered.</th>
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<tr>
<td>1</td>
<td>Referral for treatment is made.</td>
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<tr>
<td>2</td>
<td>Insurance coverage is verified.</td>
</tr>
<tr>
<td>3</td>
<td>Clinical documentation is submitted to health plan.</td>
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<tr>
<td>4</td>
<td>Case is reviewed by health plan, and decision to approve or deny is made.</td>
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<tr>
<td>5</td>
<td>If approved, care is initiated. If denied, there is an appeal process.</td>
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<td>6</td>
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But, again, coverage criteria for these diagnoses vary from payer to payer. Some payers may cover a certain diagnosis, while another may not. An example of this is multiple sclerosis. In general, the diagnosis must be proven, and the criteria for treating the diagnosis with IG should be included when submitting a request for approval (Table 2).

Prior Authorization and Predetermination
If a commercial health plan or Medicare Part D states no prior authorization is required, a provider can request a predetermination, which is essentially asking the plan to review the patient clinical information and provide a prior authorization. In the absence of either prior authorization or predetermination, a provider administering therapy risks nonpayment. If a plan denies IG treatment after treatment is rendered, a provider can bill the patient. To avoid this untenable situation, properly completing the prior authorization process is vital. No doubt, the lengthier approval time needed to receive a predetermination when prior authorization is not required is in the best interest of both the provider and patient.

Medicare Part B provides limited coverage for IVIG and SCIG in the home setting. While Part B covers many different types of conditions in the physician office or hospital infusion suite, coverage at home is limited to 24 primary immune deficiency diagnoses. Medicare Part B does not have a prior authorization or predetermination review process. Criteria for coverage is outlined in a policy called a local coverage determination. A patient’s diagnosis must be clearly outlined in the physician’s medical record with supporting laboratory testing and a description of the condition. Medicare covers 80 percent of charges for IG therapy provided in the infusion center or the home. A supplemental or Medigap plan may cover the remaining 20 percent of charges. Medicare may choose to review all of the supporting clinical information at the time of claim processing to ensure the patient meets its coverage criteria.

Understanding and Communication Is Essential
In summary, all health insurance plans have written medical policies outlining coverage for IG therapy. However, each health plan’s policy is unique, so it’s important for the ordering physician to be well-versed in what a specific plan will cover and to clearly communicate to patients up front whether they will be required to pay for any portion of the treatment.

Table 2. Criteria for IG Treatment Approval

- **Chronic inflammatory demyelinating polyneuropathy**: EMG, nerve conduction studies, history and physical showing course of symptoms and neurological exam. Some payers require a nerve biopsy.
- **Multifocal motor neuropathy (MMN)**: EMG and nerve conduction studies. Some payers want a conduction block, even though studies show not all people with MMN have this. Some payers require positive anti-GM1 antibody laboratory results, even though not all patients will test positive. History and physical showing course of symptoms and neurological exam. Because MMN is often misdiagnosed as amyotrophic lateral sclerosis, the history and physical should show a firm diagnosis rather than a suspected one.
- **Myasthenia gravis (MG)**: History and physical outlining condition and current status. MG is typically approved for exacerbation only so clinical notes should show symptoms of exacerbation. Medications tried and failed, not tolerated or contraindicated should also be described.
- **Myositis**: EMG, history and physical outlining symptoms, any myositis-related antibody testing, muscle enzyme laboratory results such as CK, ALT and AST, and muscle biopsy. Medications tried and failed, not tolerated or contraindicated should also be described.
- **Stiff person syndrome**: EMG, history and physical outlining symptoms and anti-GAD antibody laboratory testing. Medications tried and failed, not tolerated or contraindicated should also be described.
- **Autoimmune blistering diseases (pemphigus and pemphigoid)**: History and physical outlining symptoms and skin biopsy/immunofluorescence. Medications tried and failed, not tolerated or contraindicated should also be described.

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

Medicines

FDA Approves Grifols’ Xembify to Treat PI Patients

The U.S. Food and Drug Administration (FDA) has approved Grifols’ Xembify (immune globulin subcutaneous 20%) to treat primary immunodeficiency (PI) patients 2 years old and older. The product will be launched in the U.S. in the fourth quarter of this year. Grifols is also planning to sell the product in Canada, Europe and other global markets.

“This approval reinforces Grifols’ longstanding commitment to patients and healthcare professionals by expanding our product portfolio to better serve individuals with primary immunodeficiencies,” said Joel Abelson, president of the company’s commercial bioscience division. “We are pleased to offer patients living with this challenging chronic disease another important treatment option.”

Research

Informatics Could Help Diagnose PI and Save Money

A recent study shows population-wide informatics can help in detecting primary immunodeficiency disease (PI) and improve outcomes, thus saving money. In the study, researchers assessed individual risk for PI by analyzing diagnostic codes and pharmacy records from 185,893 members of a pediatric health network. Relevant infection-associated diagnostic codes were weighted and enumerated for individual members allowing for risk score calculations (risk vital sign). At-risk individuals underwent further assessment by chart review and reanalysis of diagnostic codes 12 months later. Findings showed of the original cohort, 2,188 (1.2 percent) individuals were identified as medium-high risk for having a PI. This group included 41 subjects who were ultimately diagnosed with PI. An additional 57 medium-high risk patients had coded diagnoses worthy of referral. The researchers concluded early identification of the 98 patients with confirmed or suspected PI could represent an annual cost savings of up to $7.7 million.


Public Awareness Campaign

Jeffrey Modell Foundation Releases PI Awareness Documentary

Jeffrey Modell Foundation (JMF) cofounders Fred and Vicki Modell in conjunction with an award-winning film production team have produced “Do Something: The Jeffrey Modell Story” now available on DVD, digital and video-on-demand platforms. The film’s goals are to spread awareness about primary immunodeficiency disorders (PIs), to educate those who are unfamiliar with these rare conditions and to provide a strong sense of community for patients and their families who may feel underrepresented or misunderstood.

The film features the Modell’s story about their son, Jeffrey, who succumbed to PI at 15 years of age. Prior to Jeffrey’s death, he begged his parents to “do something,” which they were determined to accomplish by searching for answers about the disease that took his life. Driven by loss, their story is one of resilience and perseverance, guided by Jeffrey’s memory and the hope to help other children.

JMF is encouraging PI patients and their families to help create the greatest awareness of PI by sharing the film with friends. The documentary is available on iTunes, Amazon, Google Play, YouTube, Vimeo, Vudu, Vubiquity, DirecTV, InDemand and DVD.

For additional information, visit dosomethingdoc.com. Individuals can also stay informed and connected on the foundation’s Facebook page at www.facebook.com/info4pi and the documentary’s Facebook page at www.facebook.com/dosomethingdoc.
IN THE NEWS

Research

New Gene Therapy Treatment Offers Possible Cure for SCID-X1

Researchers at St. Jude Children’s Research Hospital have cured infants with X-linked severe combined immunodeficiency (SCID-X1) using gene therapy involving a re-engineered virus. In the clinical trial developed by St. Jude’s Brian Sorrentino, the study’s senior author who led groundbreaking gene therapy research before his death in November at 60 years old, researchers used a modified version of HIV that can’t cause AIDS to deliver the correct gene into the DNA of eight newly diagnosed SCID-X1 infants’ blood stem cells, replacing those that do not function correctly. Two days prior to that, the infants received low-dose busulfan, an agent used in chemotherapy to help make space for donor stem cells to grow in the marrow. The majority of patients were able to leave the hospital within a month. And, all patients are developing normally so far, and none have incurred a life-threatening infection. In addition, none have developed leukemia, which was an outcome of previous gene therapy attempts for SCID-X1.

“While longer follow-up is needed to assess any late effects of treatment, these results suggest most patients treated with this gene therapy will develop a complete durable immune response without side effects,” said co-author Mort Cowan, a University of California at San Francisco professor of pediatrics.

The only other viable treatment for SCID-X1 is a bone marrow transplant, but patients must have a matched sibling donor, and fewer than 20 percent of patients usually do. Instead, they tend to rely on blood stem cells from donors who are not family, a situation that is better than no treatment, but often leads to marked side effects.


Patent

ADMA Biologics Receives U.S. Patent for Treatment and Prevention of Pneumococcal Infections

The U.S. Patent and Trademark Office has issued a patent to ADMA Biologics for methods of treatment and prevention of S. pneumonia infection. Specifically, the patent encompasses the method of preparing immune globulin (IG) via harvesting plasma from S. pneumonia-vaccinated healthy adult human donors and pooling that harvested plasma as the source for manufacturing a hyperimmune anti-pneumococcal IG containing elevated opsonic antibodies to a plurality of S. pneumonia serotypes. The patent also encompasses the prepared anti-pneumococcal IG, the method of treating S. pneumonia infection and the method of providing immunotherapy using the hyperimmune anti-pneumococcal IG.

The patent, which extends to March 2037, will enable ADMA to protect its proprietary rights while attracting collaborators interested in the development, marketing and commercialization of the needed therapeutic for treating and preventing infection of immune-compromised, immunodeficient and elderly patients who are poorly responsive to available S. pneumonia vaccines.

“This will be the first patent to issue in ADMA’s immune globulin program tailored specifically to anti-pneumococcal hyperimmune globulin compositions and treatment modalities,” said Adam Grossman, president and CEO of ADMA. “As stated in the National Foundation for Infectious Diseases, it is estimated that about one million U.S. adults get pneumococcal pneumonia each year, and as many as 400,000 hospitalization from pneumococcal pneumonia occur annually in the U.S. and about 5 percent to 7 percent of those who are hospitalized from it will die despite the widespread use of multiple vaccines for disease prevention.”

The U.S. Food and Drug Administration has approved Amgen’s romiplostim to treat pediatric patients ages 1 year and older with immune thrombocytopenia (ITP) for a minimum of six months and who have had an insufficient response to corticosteroids, immune globulin therapy or splenectomy.

Approval was based on two double-blind placebo-controlled clinical trials in pediatric patients 1 year and older with ITP for at least six months. In one study, patients whose disease was refractory or relapsed after at least one prior ITP therapy were randomized to receive romiplostim or placebo. Durable platelet response (at least six weekly platelet counts greater than or equal to $50 \times 10^9/L$ during weeks 18 through 25 of treatment) was achieved in 22 patients (52 percent) who received romiplostim and two (10 percent) on the placebo arm. Overall platelet response, defined as a durable or a transient platelet response, was achieved in 30 (71 percent) and four (20 percent) patients, respectively. Patients who received romiplostim had platelet counts greater than or equal to $50 \times 10^9/L$ for a median of 12 weeks, compared to one week in patients who received placebo. The results for all three endpoints were statistically significant, with p-values all less than 0.05.

In the other study, patients diagnosed with ITP at least six months prior to enrollment were randomized to receive romiplostim or placebo. Fifteen patients who received romiplostim achieved a platelet count of greater than or equal to $50 \times 10^9/L$ for two consecutive weeks and an increase in platelet count of greater than or equal to $20 \times 10^9/L$ above baseline for two consecutive weeks during the treatment period. No patient receiving placebo achieved either endpoint.

In pediatric patients, the most common adverse reactions included contusion, upper respiratory tract infection and oropharyngeal pain.


University of California, Los Angeles (UCLA) researchers led by Donald Kohn, MD, have created a method for modifying blood stem cells to reverse the genetic mutation that causes immune dysregulation, polyendocrinopathy, enteropathy, X-linked (IPEX), a life-threatening autoimmune disease. IPEX is caused by a mutation that prevents a gene called FoxP3 from making a protein needed for blood stem cells to produce immune cells called regulatory T cells that keep the body’s immune system in check.

Without regulatory T cells, the immune system attacks the body’s own tissues and organs.

In the study, researchers used viral vectors to deliver normal copies of the FoxP3 gene to the genome of mice blood stem cells so they produced functional regulatory T cells. All of the mice in the study were free of IPEX symptoms shortly after the treatment. The researchers also put their IPEX-targeting vector into human blood stem cells and then transfused those cells into mice without immune systems, which caused the stem cells to produce regulatory T cells that turned on the vector. “It’s exciting to see how our gene therapy techniques can be used for multiple immune conditions,” said Dr. Kohn, a professor of pediatrics and microbiology, immunology and molecular genetics at the David Geffen School of Medicine at UCLA. “This is the first time we’ve tested a technique that targets an autoimmune disorder, and the findings could help us better understand or lead to novel treatment for other autoimmune conditions such as multiple sclerosis or lupus.”

According to Dr. Kohn, to treat humans with IPEX, blood stem cells would be removed from the bone marrow of patients, the FoxP3 mutation would be corrected in the lab using the IPEX-targeting vector, and patients would receive a transplant of their own corrected blood stem cells, which would produce a continuous lifelong supply of regulatory T cells.

Dr. Kohn is also the principal investigator in a clinical trial testing the use of patients’ own genetically corrected blood stem cells to treat sickle cell disease. He also led another study that used a similar technique to cure 40 babies with severe combined immunodeficiency.

IN THE NEWS

The board of directors of the Plasma Protein Therapeutics Association (PPTA), the world’s leading trade association that represents more than 750 human plasma collection centers in North America and Europe, as well as the manufacturers of lifesaving plasma protein therapies, named Amy Efantis as its president and chief executive officer effective Jan. 16.

Efantis most recently served as vice president of global public policy and government affairs at Biogen. Previous to her role with Biogen, she held roles with Boehringer Ingelheim, PhRMA, and worked on Capitol Hill as a congressional legislative director, and, prior to this role, advised Rep. Thomas Barrett (D-WI) on various House Energy and Commerce Committee issues, primarily in the healthcare policy areas.

“I am thrilled and honored to be joining the PPTA and the vital efforts of the plasma industry,” said Efantis. “To represent companies whose mission is serving patients with rare diseases that can be treated through the development of plasma-derived products is a stewardship role that I embrace with humility and a deeply felt passion.”


Associations

Research

Octagam Safe and Effective for Treating CIDP

In a retrospective study that investigated the efficacy and safety of sucrose-free intravenous immune globulin (IVIG) Octagam (Octapharma AG) in patients with chronic inflammatory demyelinating neuropathy (CIDP), researchers found the results were consistent with the efficacy and safety of IVIG reported in randomized controlled studies.

In the study, data from 47 patients who received at least one dose of Octagam were collected from the records of 11 centers in France. Efficacy was assessed using the Overall Neuropathy Limitation Scale (ONLS), and safety was evaluated using adverse event rates. Of the 47 patients, data from 24 patients who were IVIG naïve (n = 11) or had stopped IVIG more than or equal to 12 weeks before initiation of Octagam therapy (washout group; n = 13) were included in the efficacy analysis. At four months post-initiation of Octagam treatment, 41.7 percent of patients had improved their functional status (decrease of greater than or equal to one ONLS score) with a significant change in the ONLS score from baseline (- 0.42; p = 0.04; signed test). Functional status was reduced in only two patients: one patient in the IVIG-naïve group and one in the IVIG-washout group. All 47 patients were included in the safety analysis, which showed Octagam was well-tolerated, with a frequency of 0.04 adverse events per Octagam course. The most common adverse drug reaction was headache.

According to the researchers, a long-term prospective study of Octagam in patients with CIDP is warranted.


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The Benefits of Having a Chronic Illness

A lot of positive lessons come from having a chronic illness; they just need to be acknowledged.

By Surayyah Morris
THOSE OF US with chronic illness are in a daily battle against our bodies, making it sometimes difficult to see the good in things. But, here are a few reminders of what we can learn and how we benefit from our experiences of dealing with chronic illness.

Advocacy

If there is one thing to be learned from having a chronic condition, it’s how to advocate. Having someone advocate for us is very reassuring. But, eventually, we realize that those who fight so hard for us when we’re too weak or unable are actually teaching us to do the same for ourselves.

Pay close attention to what your advocate is saying to the doctor. Listen to the types of questions he or she asks. Watch how your advocate remains assertive to help the doctors provide you with what you need. Throughout your experience, you’ll develop these skills, and you can begin advocating for yourself and feel confident doing it. So, tell your story. What do you feel? How are your daily activities affected? How can the issue be improved or changed? Who else can you speak to about your concerns? Let others know you are serious about being present in your care. Surround yourself with people who will stand by your side to advocate with you and support you.

Compassion

Having compassion not only forwards our kindness to make someone else feel better, but also serves us in a way that helps us to be more mindful and present in our own lives. We are more understanding of others in their situations because we’ve likely experienced some type of mistreatment during our journey. Being mistreated is not a good feeling for anyone, but it is especially discouraging when it comes from those who are supposed to help. Whether or not we have a chronic condition, we need people in our lives who care and show it through their words and actions.

Not everyone you encounter will go out of their way to provide good care, but when that one nurse finds a warm blanket when you forgot yours for a half-day infusion, you are beyond grateful. When the doctor listens to your concerns and responds with empathy instead of dismissing you with a psychosomatic spiel, you gain more trust and respect for that person. Appreciate these moments. Pay it forward and extend your hand to others. Even a simple smile goes a long way. Best of all, you don’t lose anything!

Our experiences teach us that everyone may not be genuinely compassionate and caring, but we can be that empathetic person for someone else. In short, treat others the way you want to be treated.

Improvisation

On bad days, we develop extraordinarily convenient ways to get things done. When our bodies decide to take control, we have to take control back. No strength to cook? There’s an app for that. Haven’t cleaned the house in weeks? There’s an app for that, too. Need to use the bathroom but can barely get out of bed? There isn’t an app for that, but there is a bedside commode. Ice packs getting warm too quickly? There isn’t an app for that either; however, you can freeze several ice packs and keep them in rotation or place a mini fridge/freezer where it’s most convenient.

The point is that whatever you would typically struggle with, I guarantee you will find the easiest, simplest, least-taxing way to get that task accomplished. This may take some trial and error, but you’ll be more than proud of yourself for having tricks up your sleeve in a variety of situations.

Having patience allows us to get what we need, accomplish tasks successfully and solve problems efficiently.

Observant

When it comes to our health, we cannot be careless. No matter how much we trust the people involved in our care, we must do our best to remain attentive. During one of my first infusions, I had my attention on the television when I broke out in hives and my throat began to swell. I was having an allergic reaction to a medication that was inappropriately administered by my nurse. From that day forward, I always ask to physically verify what I’m being administered before allowing anyone to touch me. I also make sure I’m not watching television.
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.

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

- 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.
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.

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
Ensure all hands are properly washed and technique is clean. If the doctor comes in with multiple people, ask about the role of each individual and what purpose they serve in your care. Medical mistakes are easy to make despite having processes in place to prevent them. Be alert, oriented and fully present. Ask questions. Most importantly, though, read! The information you receive is in writing most of the time. It seems tedious, but read all the documents, and be sure you understand them. Information in black and white may not always be explained to you, even if it should be. If you do not understand something, ask for clarity. Have someone with you as a second set of ears and eyes to make sure you’re safe and being treated well — to act as your personal lookout!

**Patience**

Most of us with chronic illness might as well set up camp and live at the doctor’s office. Sometimes, we observe a healthy person becoming impatient when experiencing long wait times or when unexpected problems arise. But for those of us with chronic conditions, our experience with multiple doctor visits with multiple doctors in multiple places and for a variety of reasons gives us a different perspective. At minimum, we deal with insurance claims, disability forms, accommodation requests and the pharmacy. Because we usually don’t have a choice, at the very least, we learn to wait. Over time, waiting builds patience. Having patience allows us to get what we need, accomplish tasks successfully and solve problems efficiently.

This outlook translates into positives because in addition to having more patience with wait times, you’ll also have more patience when dealing with people. Waiting for authorizations, referrals or pharmacy benefits to process can be frustrating and time-consuming, but knowing how to prepare ahead of time can also minimize stress.

**Prioritization**

When more than one provider is managing our health, things become overwhelming. A whirlwind of information is often provided to us with no clear direction of what to tackle first.

When the to-do list begins to grow, you will learn what’s most important to accomplish based on your situation. For example, I used to think my newly diagnosed brain aneurysm was going to be the driving factor in how my life moved forward. Then, I realized my aneurysm wasn’t causing me any problems. Yes, it’s very much present, but it isn’t affecting my health like my other conditions. Being able to improve my health by giving priority to my primary condition as opposed to giving energy to something that isn’t immediately affecting me has allowed me to be a little less stressed and lot less worried. Try to prioritize your concerns from most to least important so managing your health becomes easier overall.

**Strength**

We all have bad days, but those days come and go. Now, you may be reading this after a terrible day that seemed bad from start to finish. But remember, you are stronger than most of the people in your life, I promise. You fight a battle with your body daily, you remain mentally strong, you take care of yourself without compromising the care of others, and you wake up the next day to do it all over again. That is so difficult! But despite the pain, inconvenience and unpredictability, you are able to adapt, and that takes more strength than anyone will ever know. A stranger once told me fear means we are afraid of something we have already been through and survived. Don’t be scared to show your strength.

**Give Credit Where Credit Is Due**

Don’t let the circumstances of life trouble you. Look at all you’ve learned so far. Continue to learn and grow and be the person you need to be for you. The next time you find yourself down about your condition, give yourself some credit, and acknowledge how much you have learned through this journey!

**SURAYYAH MORRIS**, PharmD, is an autoimmune small fiber neuropathy patient from Central Florida. As a medication therapy management and pain management specialty pharmacist, she enjoys supporting patients with chronic pain and chronic conditions to help find balance and improve quality of life.

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Don’t be scared to show your strength.
WE ALL NEED a little extra help sometimes. Whether it’s tracking our medications, enhancing the strength of them or finding new ways to cope with chronic illness, sometimes retail therapy really is the answer! We hunted for the best in new tools and tech for patients and here’s what we found to help you get through what you have to manage.*

Managing Fevers and Staying Cool

- FeverMates Cooling Patches; $4.18 (FeverMates.com)
  You’ve probably seen mentholated products for fevers or migraines, but who wants to smell like chemicals or feel that icy-hot burn on their face? FeverMates Cooling Patches by Mediband offer an alternative with their medication-free, no-drop, no-slip stick-on patches. These peel-and-stick pads go directly on your forehead (or the back of your neck) and instantly begin cooling you down. No scent and no residual stickiness means these are a great on-the-go remedy. While not reusable, one patch lasts approximately eight hours and a pack of six pads sells for only $4.18. I’ve found them useful for high fevers, migraines, hot flashes and unbearably hot summer days. Take care to use only as directed, and remember that because this is not a medication, it won’t solve the root cause of a fever, so use these as a supplementary tool to keep you cool when you’re feeling on fire!

- FeverMates Stick-On Temperature Indicators; $5.62 (FeverMates.com)
  If you’ve ever had to wake a sleeping child to take a temperature, you know how awful it can be for the both of you. Avoid the tantrum and relieve your anxiety by using one of these child-friendly stick-on thermometers. Made in colorful, fun animal designs, these stickers go right on your child’s forehead and give a continuous temperature reading. There are several black circles on each

*Editor’s note: Statements have not been evaluated by the U.S. Food and Drug Administration. These products are not intended to diagnose, treat, cure or prevent any disease.
sticker. If one of the circles reads “N” it means your child’s temperature is normal. If it’s higher than normal, the digits will appear in one of the other circles. When dealing with immune deficiencies, temperature readings are imperative during a cold, flu or infection.

- **Thermo by Withings; $99.95 (Withings.com)**
  It’s important to keep track of your body temperature when you have an immune deficiency. Even the slightest increase in your “normal” can be cause for concern. That’s why Withings has created Thermo, a Bluetooth-enabled temporal thermometer that allows you to record your temperature throughout the day and track it on your phone. It’s more sanitary than oral thermometers, can track up to eight users, syncs with your Wi-Fi and its batteries last up to two years! This is a must-have for parents or adults dealing with chronic illness who want to use technology to take control of their health.

- **SkinCool Ice Rollers; $25 (Hansderma.net)**
  Have you ever had a headache or a fever so bad you’ve considered dunking your head in a bucket of ice? I have a solution that’ll deliver a little less shock to the system while still cooling you down in a hurry. Hansderma’s SkinCool Ice Rollers live in your freezer and can be used to roll over your face, neck or entire body! They deliver a cooling, comfortable massage with little effort, and their broad-barreled rollers do all the work so you don’t have to. Quit cleaning up wet washcloths and sticky ice-packs and roll on to cool down with this handheld roller ice pack.

- **Snuz Mattress; $500 (TWIN) (SleepChoices.com)**
  When your immune system is at an all-time-low, it’s imperative your hours spent getting a restful night’s sleep are at an all-time high. With so much hype in the online mattress world, it can be hard to tell if the mattress you order will be a perfect fit. If you’re concerned about overheating, Snuz might be your perfect fit. The company’s foam mattress allows cool air that enters through its knit fabric to travel up through special grooves to allow your body to cool while you sleep. Snuz is CertiPUR-US-certified and made without ozone depleters, PBDE flame retardants, mercury, lead, other heavy metals, formaldehyde or chlorofluorocarbons, and it is safe and environmentally friendly.

**Wearable Health**

- **Skineez Hydrating Compression Socks; $19.99 (GetSkinEez.com)**
  Whether you’re stuck in bed for too long or just deal with chronically poor circulation, you can get your blood moving with these compression garments from Skineez. No one wants to wear uncomfortable shapewear when they’re already feeling faint. Skineez uses a unique fabric that slides on comfortably and breathes. All the benefits of compression and none of the discomfort. Skineez are imbed with shea butter to moisturize, apricot kernel oil to increase elasticity, rose hip oil to rejuvenate, retinol to soothe and vitamin E to nourish skin. The company’s unisex compression socks are easy to put on and comfortable enough to wear all day. They also sell compression leggings and gloves for joint pain.

- **MediBand Write-On Medical ID Bracelets; $6.40 (Mediband.com)**
  Mediband knows it’s difficult to find appropriate medical ID jewelry for rare diseases, so they created a write-on band that allows wearers to write the name of their condition themselves. Their ultra-light and comfortable silicone band IDs ensure against incorrect medical treatment, give vital information to others when you can’t speak for yourself and are 100 percent hypoallergenic. If you upgrade to their Mediband Plus product, your bracelet will give first responders a secure website and unique code to get even more details about your health, including recent medical records.
• WeatherX Earplugs for Pressure Headaches; $11.95 (WeatherX.com)

Changes in weather can cause extreme discomfort for patients with chronic ear infections. WeatherX offers natural relief with its pressure filtering earplugs. Their ceramic inner filter helps regulate air flow in and out of the ear canal, and their four-ring design offers a snug and comfortable fit. These earplugs are made out of soft, cleanable and hypoallergenic silicone available in both child and adult sizes. Users suggest slipping them in at the first sign of discomfort. WeatherX even offers its own downloadable app to get weather pressure alerts to remind you when to slip them in.

• Bioscarf; $44.99 (Bioscarf.com)

Embarrassed to wear a mask in public? Protect your personal space with the first scarf with air pollution, allergy, cold and flu protection built in. If you’re concerned about air pollution, allergies or contagious illnesses like pneumonia, strep, influenza or tuberculosis, Bioscarf literally has you “covered.” These stylish accessories come in black, white, green and camo, and as part of their PlusOne Program, a portion of the proceeds go to giving scarves to patients living in high-risk areas with toxic air pollution.

New in Health Technology

• Wynd Plus Smart Personal Air Purifier with Quality Sensor; $199.95 (shop.hellowynd.com/products/wynd)

Have you ever walked into a room and immediately thought: “Oh no, I better hold my breath!” If you suffer from asthma and allergies, you probably have. With Wynd Plus, you can not only check the air quality wherever you go, but improve it with this tiny, portable air purifier. Wynd Plus creates a bubble of clean air around you by removing dust, smoke, allergens and pollution from the air. It captures particles as small as 0.3um (1/210th the average thickness of human hair!). Wynd Plus breaks down into two pieces: the air purifier that comes with its own kickstand for easy placement and a detachable tracker that displays a different color based on the quality of the air. Leave the device in a place with poor air quality and Wynd Plus cleans it up!

• Nima Starter Kit; $289 (www.NimaSensor.com)

Anyone with a gluten or peanut allergy knows that just because a product says it’s peanut- or gluten-free doesn’t always mean it actually is. That’s why Nima created a compact food tester that takes the first bite for you! This amazing, portable device allows users to take a sample of their meal, pop it into Nima’s sensor and know for certain whether the food is allergen-free. This has to be one of the coolest new products out there that truly gives patients some control over their diet and their lives. An absolute must-have for anyone who fears eating outside their own kitchen!

• TimerCaps 4 Pack; $19.95 (Timercap.com)

Don’t guess when the last time you took your medication was, know for sure with TimerCap! The lids on these pill bottles count down to the exact second you last took your medication and helps solve that age-old concern about forgetting to take your meds. These simple, yet innovative pill caps have been featured on “The Today Show” and in The Wall Street Journal, and they come in all shapes and sizes. They help patients keep track of their medication schedules and help stop prescription abuse and deter unwanted openings. In addition, if you’re concerned about keeping pain medication at home, the TimerCap can act as a strong deterrent to thieves since patients will know the bottle has been opened at the wrong time.
Nausea and Pain Relief

- Moji Roller; $69.99 (GoMoji.com)

Rolling out sore muscles with self-massage techniques can help patients stay comfortable when pain strikes. Moji has taken the experience to a new level by creating a line of microwavable massage tools. Their heated foam rollers and massage balls can fit in any standard microwave and stay warm for hours, allowing for extensive relief anytime. With added heat, Moji Rollers help to relax sore muscles, enhance healing, speed recovery, increase circulation and boost endorphins.

- Quell 2.0 Starter Kit; $299 (www.QuellRelief.com)

Can pain be quelled by neurotechnology? That’s what some patients are saying after using Quell Relief for 30 days. This 100 percent drug-free, FDA-cleared device is a prescription-strength nerve stimulation powered by a Bluetooth-enabled device that straps around your calf. Quell is safe to wear for 60 minutes at a time up to three times a day to help quiet pain throughout the body. Using the app, patients can adjust the intensity or stop the device at any time. It can be worn while active or sleeping. It is up to 10 times more powerful than the average over-the-counter TENS unit devices.

- PainPod 3; $479 (www.painpodusa.com)

PainPod has combined advances in science waveform, electrotherapy and microcurrents with the latest in neurological understanding and advanced bioengineering to create a comfortable, easy-to-wear device that conquers pain without medication. The device is currently being used throughout the world by occupational therapists and medical rehabilitation centers to reduce pain throughout the body. While the device can be used with traditional pads that stick to pressure points, it can also connect to custom gloves, socks, foot pads and even a belt that can help reduce abdominal pain. While all three units (PainPod 3, PainPod XPV and PainPod Mi) are portable, they are easily one of the most expensive electronic pain devices on the market. But, according to athletes Michael Jordan, Bruce Lee and Jack Nicklaus, these devices really do work.

- Tummydrops; $10.99 (Amazon.com)

Designed by a gastroenterologist, Tummydrops are the new go-to for getting through nausea. When you’ve maxed out on prescription antiemetics, these all-natural lozenge candies are an excellent next step. Made with ginger and peppermint, they taste delicious and are vegan, gluten-free, organic and kosher. Each drop is individually wrapped for easy storage in your purse or pocket. New flavors include sweet blackberry ginger.

- Relief Band; $174.99 (www.reliefband.com)

You’ve probably tried those silicone nausea bands sold at the grocery store, but nausea relief just went to the next level with ReliefBand 2.0. This fascinating little device looks like a smartwatch but zaps queasiness like a TENS unit. Find the pressure point on your wrist using the included instructions, apply a small drop of gel, place the device over the gelled area and fasten snugly. You can adjust the amount of stimulation to the area depending on the severity of your nausea. This device is not for the faint-hearted, it delivers a seriously distracting buzz — maybe just distracting enough to quell your nausea.

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Building Physical Endurance with a PI and/or AD

By Matthew D. Hansen, DPT, MPT, BSPTS

Most people can participate in a physical endurance conditioning program, but it is advised for those with a PI or AD to start slowly and progress steadily.

POOR PHYSICAL ENDURANCE or “the time span between the beginning of physical activity by an individual and the termination because of exhaustion” is often a familiar complaint of people living with a primary immunodeficiency disease (PI) or autoimmune disorder (AD). To be sure, fatigue is not an uncommon complaint for the population at large, and there are many potential contributing factors, including poor sleep, diet, infection, low blood sugar, anemia, some pharmaceutical medications and a number of physical disorders.

Under the premise of improving physical endurance, cardiorespiratory function (or how well the heart, lungs and muscles work together) “to use oxygen to produce energy for movement” is usually the topic of discussion. Although the benefits of exercise are generally established and rarely disputed, it’s startling how many people believe they are an exception to the rule and exercise would somehow be more detrimental to their health than not doing anything at all. This is simply flawed perception. When people view themselves performing aerobic exercise, it might be running a marathon, jumping rope, Zumba or other strenuous activities that cause them to pour sweat out of every pore and gasp for air. But, aerobic exercise can be light to moderate in intensity, too. For some, vacuuming the floor is an aerobic exercise and, by applying the principles of cardiorespiratory training, can improve physical endurance over time. The point is, almost anyone can safely improve cardiorespiratory endurance.
Underlying Principles of Aerobic Exercise Prescription

Exercise is normally prescribed according to the parameters of frequency, intensity and time (FIT). In other words, exercise volume equals F times I times T. Once someone understands the principles of exercise prescription and possible exercise adaptations, it’s easy to see the wide array of possible activities to increase cardiorespiratory endurance.

Frequency and time (duration). It is generally recommended adults participate in aerobic exercise three days to five days a week, with activity continuing at least 20 minutes to 30 minutes to impact cardiorespiratory conditioning. However, some studies suggest sessions of at least 10 consecutive minutes can be performed intermittently throughout the day to effectively acquire the recommended daily duration; and even sessions that are less than 10 minutes in duration may be of benefit in extremely deconditioned individuals (though more evidence is needed to support their effectiveness).

The American College of Sports Medicine (ACSM) states most adults should participate in moderate intensity exercise for 30 minutes to 60 minutes (more than 150 minutes a week), in vigorous intensity exercise for 20 minutes to 60 minutes (more than 75 minutes a week) “or a combination of moderate and vigorous intensity exercise daily to attain the recommended targeted volumes of exercise.” However, ACSM also points out that “less than 20 minutes of exercise per day can be beneficial, especially in previously sedentary individuals.”

Intensity. The overload principle of exercise indicates a minimum intensity (or threshold) must be reached and surpassed to result in physiologic changes. Most exercise and sports science physiologists use someone’s maximal volume of oxygen uptake (VO2max) over time to measure gains with cardiorespiratory function. However, the minimum threshold of intensity for benefits appears to vary based on several factors, including current level of cardiorespiratory function, age, health status and genetics. So, establishing an exact threshold for exercise intensity, particularly for someone working out on their own at home, can be tricky.

The easiest, although not the most accurate, way to determine workout intensity for cardiorespiratory benefits is by establishing a target heart rate (HR).

Target Heart Rate

To figure out the max/peak HR without population-specific formulas or by taking an exercise or chemical stress test (both which could be contraindicated for some conditions), a participant can simply subtract their age from 220. For example, for someone who is 69 years old, their estimated max/peak HR would be 220 minus 69, which would be 151 beats per minute (bpm).

Once the maximum heart rate is known, the target HR zone is calculated. This is the range in which exercise participants will try to maintain their HR to improve cardiorespiratory endurance over time. The American Heart Association recommends a general target HR of:

- 50 percent to 70 percent of maximum HR for moderate exercise intensity
- 70 percent to 85 percent of maximum HR for vigorous exercise intensity

Because of factors unique to individuals, it’s not uncommon to misestimate exercise intensity when using this simple formula. Therefore, instead, a more accurate target zone can be calculated by using someone’s HR reserve (HRR):

1) Calculate maximum heart rate as before (220 minus age).
2) Figure resting HR by counting bpm when relaxed and sitting down.
3) Subtract resting HR from maximum HR to get HRR.
4) If aiming for moderate exercise intensity, multiply the HRR by .50 (50 percent), and add the resting HR to this number.
5) Multiply HRR by .70 (70 percent), and add resting HR to this number.

It is generally recommended adults participate in aerobic exercise three days to five days a week, with activity continuing at least 20 minutes to 30 minutes to impact cardiorespiratory conditioning.
6) The exercise participant’s HR should be between the two numbers calculated in steps four and five. This is the training zone HR for moderate intensity exercise.

Reusing the example of someone who is 69 years old with a max/peak HR of 151 bpm (220 minus 69), let’s say we check his or her resting HR and find it to be 68 bpm. This number is then subtracted from his or her max/peak HR of 151 to get the HRR (151 minus 68 equals 83 bpm).

If we were creating an aerobic exercise program at a moderate exercise intensity, we would multiply the HRR of 83 by .50, add the resting heart rate of 68 back to this number and round to the nearest whole number (83 times .50 plus 68 equals 110 bpm). This is the low end of the recommended training zone HR. The same steps would be repeated by multiplying the HRR by a factor of .70 instead of .50 to get the high end of the training zone HR (83 times .70 plus 68 equals 126 bpm). The targeted training zone HR for this individual, then, would be between 110 bpm and 126 bpm.

The recommended percentage of HRR varies somewhat by source. For example, ACSM recommends 40 percent to 59 percent HRR for moderate aerobic exercise and 60 percent to 89 percent HRR for vigorous intensity exercise, with light aerobic exercise as low as 30 percent to 39 percent HRR proving beneficial to individuals who are deconditioned. The essential warm-up and cool-down periods of an aerobic exercise routine typically take five minutes to 15 minutes each, and are performed at approximately 50 percent of the conditioning period stimulus intensity (i.e., at a level that allows the participant to hold a conversation without much difficulty).

Many exercise machines now have built-in HR monitors, or participants can purchase a portable pulse oximeter at a reasonable cost to take their pulse through their finger. However, the tried-and-true manual method of checking the pulse over the carotid or radial arteries also works. If taken in this manner, the pulse should be checked with the index and third finger placed over the artery while counting the number of beats for 15 seconds. At the end of 15 seconds, multiply by four to calculate bpm.

**Modes of Aerobic Exercise**

The most effective types of aerobic exercise are those that involve the use of large muscle groups and are rhythmic in nature. The following is a short list of activities to get people started:

- Walking/dancing
- Swimming
- Biking
- Aerobics
- Climbing stairs
- Hiking
- Jogging
- Aqua aerobics
- Rowing
- Skiing
- Elliptical exercises
- Running
- Sports
- Stationary bike

When training for a particular activity, the principle of specificity indicates the physiologic adaptations needed are specific to the type of exercise performed. In other words, if a person wants to become a better-conditioned swimmer, his or her most effective training exercise is swimming, although the benefits of cardiorespiratory endurance certainly do carry over between all activities of life.

**Progression of Aerobic Exercise**

There are three stages of aerobic (cardiopulmonary) conditioning (because the parameters that are often presented with each stage should also take into consideration individual factors, they are not included here):

- Initial conditioning stage: This typically lasts for the first four weeks to five weeks of training. However, if the exercise
routine is interrupted for significant periods, it may last longer.

Improvement stage: Presuming continued adherence to the exercise routine, this stage usually lasts four months to five months.

Maintenance stage: This tends to begin after the first six months of training. Further gains in cardiopulmonary function may not be significant, although continuing aerobic exercise at least three days a week will help to maintain current levels of fitness.

ACSM suggests “a gradual progression of exercise volume by adjusting exercise duration, frequency and/or intensity is reasonable until the desired exercise goal (maintenance) is attained. This approach of ‘start low and go slow’ may enhance adherence and reduce risks of musculoskeletal injury and adverse cardiac events.”

It’s important to note that volume should not be increased by more than one factor at a time. For example, if a participant feels he or she can increase the duration of workouts, the intensity should only be increased once the person is confident about participating in longer sessions. I usually recommend taking at least one full week to acclimate to a change so participants aren’t only successful with a new workout, but they can also recover from it successfully.

Special Considerations

It was long believed exercise — at least strenuous exercise — could temporarily suppress immune function. However, if this is the case, it appears the period of vulnerability, as suggested by a reduction in white blood cells circulating in the blood, is short-lived and only occurs “after an excessive amount of prolonged, high-intensity exercise.”

The theory suggests that this “open window” of increased risk of infection lasts between three hours and 72 hours. However, a widely cited 2018 review study published in *Frontiers in Immunology* concluded “regular physical activity and frequent exercise are beneficial or, at the very least, are not detrimental to immunological health.” The authors further noted that “limited reliable evidence exists to support the claim that vigorous exercise heightens risk of opportunistic infections.”

Although it’s probable that being active is likely to be beneficial versus detrimental to immune function, some modes of aerobic exercise may be contraindicated for people with specific medical conditions. For example, someone who is known to have a low platelet count should not participate in contact sports. There is also danger in overtraining, particularly for those living with ADs that are characterized by chronic inflammation. Stressful exercise can increase inflammation and make symptoms worse. Too much too soon can also result in musculoskeletal injuries.

Start Slow and Steadily

Almost anyone can improve physical endurance, but rather than exacerbating symptoms of a PI or AD, it’s best to both start and continue to improve one’s cardiopulmonary function slowly and steadily.

Patients should begin by consulting with their doctor, and consider utilizing the expertise of an exercise physiologist, personal trainer or physical therapist to build a program. If they are just beginning an exercise program, or if they have other precautions that would contraindicate exercising at a higher intensity, the objective should still at least initially be at the lower end of their HR target zone, with the goal of building up gradually.

Just “staying in the game” is winning with aerobic training, because once training is discontinued, cardiopulmonary function will decrease by approximately 50 percent within four weeks to 12 weeks. Individuals are advised to choose activities that are fun and/or are functional and need to be done anyhow. If someone is consistent in performing them, and consistent in applying the principles of aerobic exercise prescription, the benefits will be felt, and physical endurance will improve.

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References

HERE’S A QUESTION we are not asked often but should consider: If someone were to ask your family members, friends and healthcare providers what your end-of-life wishes are, would any of them know the answer?

End-of-life matters are a conversation 95 percent of people in the United States say they want to have, and 92 percent say they believe is very important. Yet, only 32 percent of Americans have actually approached the subject with the people they believe they want to be involved in such decisions. What’s more, only 37 percent have put their end-of-life wishes in writing for friends and family to access when needed.

Conversely, what are your friends’ and family members’ hopes and desires for how they want to spend their final years and months? Would you be able to help them live their final season in comfort, with gladness and even with peace? If you don’t have the answers to any of these questions — and even if you are avoiding the topic altogether — statistics show you are clearly not alone. But, as more people understand the basics of advance care planning, the statistics can change for the better.

In his seminal book on end-of-life matters, Being Mortal: Medicine and What Matters in the End, Atul Gawande, MD, MPH, writes, “You may not control life’s circumstances, but getting to be the author of your life means getting to control what you do with them.” Thankfully, there are ways to make this difficult conversation and all of its resultant logistics more approachable.

**When to Discuss End-of-Life Wishes**

The best time to discuss your beliefs and values regarding your or a family member’s end-of-life care is well before a health crisis strikes. Working through such matters early reduces anxiety, emotional exhaustion and other stressors. (If you doubt this, imagine attempting to prepare a detailed, well-considered plan for anything, in addition to navigating a long-term emergency or difficulty.) Thinking through and discussing things beforehand allows ample time to address issues carefully so end-of-life plans can be most beneficial and peaceful for all.

And, lest anyone believe advance care planning is only for older people, it’s not. No one knows when they could face illness or grave injury. And, no one knows when their loved...
ones will, either. Whether you are a young mother, a middle-aged empty nester, a grandparent or a single young person, having some idea of how you want your end-of-life choices to be handled is invaluable for you and others should something devastating occur.

The common reluctance to address this issue is more than understandable, though. Talking about end-of-life choices and wishes is something that can elicit sorrow and angst, among many other feelings. Any time we are reminded of mortality, whether it’s our own or a loved one’s, the threat or reality of lingering illness, or the pressing matters of managing finances, estates and legal matters, we can feel overwhelmed. And, for good reason. But it’s even more uncomfortable for everyone not to discuss it.

Because so much rests on having an end-of-life dialogue, it is something we will all want to face despite any discomfort. And, we won’t regret doing so. Without it, relationships, emotions and finances could be unnecessarily strained, and you will not receive the type of care you hope for. The lives of everyone involved can be made less stressful and even more enjoyable because simple preparations are in place for the future to mitigate grief, crises, uncertainties and frustrations when the final season ushers in.

Preparing for the Discussion

If you feel nervous about talking about such an important subject, understand that not everything rides on one conversation. End-of-life decisions and wishes are typically discussed — and even discovered — through a series of conversations over time. So, when you begin the discussion, you do not necessarily have to have everything planned out that moment. It’s often a process. And before you begin, you might want to prepare yourself first.

For some people, writing their thoughts in letter or outline form is helpful for organizing the points they want to make. For others, role-playing with a trusted friend, spiritual advisor or therapist can be helpful. No matter the method, the point is to give yourself a chance to understand and shape your thoughts, desires and emotions.

To begin, ask yourself what concerns you most about addressing the subject with loved ones and healthcare providers. If you can identify these things, you can begin to better understand and address them in productive ways.

And give yourself grace if you feel anxious. This is a difficult subject for virtually everyone.

Some Main Points to Consider

Advanced end-of-life care planning touches on multiple aspects of a person’s social, familial, spiritual and healthcare worlds. While there are many facets to consider, here are some main points to discuss and prepare for. Of course, you may add additional questions or thoughts, if necessary. But, this list gives you a place to start.

People Involved in My Care

• Who will be my primary beneficiary?
• Who will be my primary caretaker?
• Who will have power of attorney, if necessary?
• What kind of bedside manner do I hope for in a physician? (On a more humorous note, would I prefer Dr. House or Nurse Nightingale? To each his or her own!)

• What do I want my healthcare providers to know about me personally, if anything? What about my other caretakers?
• How do I want hospice or other healthcare providers to interact with me? For example, do I want them to chat jovially with me? Say only the minimum? Not ask questions about my personal life?

Medical Treatment

• What types of medical treatment do I want, and when? For example, strictly palliative or more aggressive?
• What about obtaining a do-not-resuscitate order?
• How much do I want to know about my medical condition? Should doctors tell me everything, or do I want to be unaware of much?
• How much should my friends and family know about my condition? How should they interact with me regarding a diagnosis?
• What is important to me with regard to dignity and comfort in my care?

The best time to discuss your beliefs and values regarding your or a family member’s end-of-life care is well before a health crisis strikes.
Legal and Financial Affairs
• Who do I want to receive my most cherished possessions, and how will they obtain them?
• Who is handling my will?
• Who will be given what from the estate?
• Do I want a living will?
• Which charitable or for-profit organizations do I want to donate my vehicles, home and other assets to?

Spiritual
• How will my faith traditions be carried out during my last days and at my memorial service or funeral?
• Do I want a spiritual leader of my faith to visit me and carry out rites or ceremonies?
• What type of service do I want to commemorate my life? Favorite music to be played, for example? A poem or passage of scripture to be read?
• Do I not wish to have a memorial service or funeral?

Breaking the Ice
While a single conversation is rarely sufficient, the first one regarding end-of-life decisions can at least broach the subject and lessen initial discomfort. Find a time to mention things when the people you are discussing it with are not hurried or distressed. You might approach by saying something simple such as, “I need your help with something I’ve been considering,” or “I’ve been thinking about the future and could use your insight.”

Significant family life events such as a wedding, funeral or retirement party can also serve as a springboard to mention your desires. You could also bring it up while preparing or revising a will or working on estate planning. Holidays, although they can be stressful for some, might also serve as an opportunity since the family is often together. And, if another friend or family member is facing end-of-life decisions, that opens the door to conversation as well.

Because your thoughts about end-of-life care could evolve over time, be sure to amend your verbal or written plans should a significant change occur. Keeping your loved ones apprised is important, and they might appreciate it as well. You could even ask, should they have their own plans, whether they have new ideas.

Anticipating Disagreement and Resistance
It’s important to consider the possibility that you and a loved one or healthcare provider could disagree on something while discussing end-of-life wishes. Or, perhaps more common, it’s possible you’ll be met with deflection or other resistance.

For example, a common way people deflect advance care planning when it feels uncomfortably negative is to say, “We don’t need to do that. We’ll be able to handle it all.” Or, “You can beat it if you get sick.” It’s helpful to consider how you will respond. Saying something as simple as, “I want to be prepared anyway,” or, “This will be useful because it will relieve stress and help us to invest in one another, instead of scrambling for a plan when the time is shorter.”

Or, if you are approaching a loved one to discuss a plan for their own care, they might respond with hostility or passivity. “I don’t want to talk about it,” “I don’t need anything special” or “It’s too painful to think about” are common responses from resistant loved ones. A helpful way to address these comments might include saying, “We care about you and want you to be comfortable. It’s more painful for all of us not to discuss it in the long run, because we won’t know what to do.” Or, perhaps, “We understand your frustration. It’s a difficult topic. We still want to help you plan because we want to know we are making the best, most helpful decisions for you when the time comes.”

If healthcare providers disagree, perhaps a social worker, counselor, chaplain, mediator or other official can help to ease the friction and help foster better understanding between parties.

It’s Never Too Early
It’s never too early to begin the discussion. Professional advance planning services such as Five Wishes are also available for those who want a prescripted directive approach and helpful checklists. Remember, few ever regret the planning, even if initiating the discussion was uncomfortable. An advance care plan is worth the awkwardness of the first conversation.

MEREDITH WHITMORE is an English professor and freelance journalist in the Northwest.

References
By Ronale Tucker Rhodes, MS

IN 2014, THE Crohn’s and Colitis Foundation of America estimated 1.6 percent of Americans (more than five million) have inflammatory bowel disease (IBD), which includes Crohn’s disease (780,000) and ulcerative colitis (907,000). Researchers estimate six to 15 new cases of Crohn’s are diagnosed per 100,000 people each year.¹

Crohn’s is a chronic IBD characterized by inflammation of any part of the gastrointestinal (GI) tract, from the mouth to the anus; however, it more commonly affects the small intestine where it joins the beginning of the large intestine (or colon).² Crohn’s can also affect the eyes, skin and joints.¹

The disease affects both adults and children. The prevalence of Crohn’s in 2014 was approximately 241.3 per 100,000 adults 20 years and older, and 57.8 per 100,000 in children and adolescents under 20 years old. Sadly, these numbers have continued to increase. In 2004 and 2005, 43 children and 214 adults per 100,000 people had Crohn’s. In 2008 and 2009, these estimates rose to 48 children and 236 adults per 100,000 people.¹

Crohn’s is more common in females, and Caucasians and Ashkenazi Jews develop it at a higher rate than other ethnicities; however, the rate of diagnosis for African Americans is approaching that in whites.¹ ¹¹ Interestingly, children are twice as likely to be diagnosed with Crohn’s than ulcerative colitis.³ Crohn’s is also more common in the North than in the South, and the number of people diagnosed is higher in the Northeast and Midwest than in the South or West.¹

Crohn’s can be treated, but it cannot be cured. And, it can significantly degrade a patient’s quality of life and may have a high financial burden. In a 2008 review, direct medical costs in the U.S. were $18,022 to $18,932 per patient per year¹ at an annual cost of $3.6 billion. More than one-third of expenses are due to medications (with biologicals generally the most expensive) and hospitalizations (31.4 percent of cost).¹

Causes of Crohn’s Disease

The cause of Crohn’s is not definitively known. It is believed to be an autoimmune disease, but recent research suggests that rather then the immune system attacking the body itself, thus causing chronic inflammation, the immune system may instead be attacking a harmless virus, bacteria or food in the gut.⁴

There are several known risk factors for Crohn’s. One is genetics. Approximately 20 percent of people with IBD have another family member with IBD, and families frequently share a similar pattern of disease. In fact, between 5 percent and 20 percent of people with IBD have a first-degree relative with IBD. When both parents have IBD, the risk of their children developing Crohn’s is 35 percent.³

Age is also a risk factor, primarily affecting young people, with most people diagnosed before age 30. A diet high in fat and/or processed foods increases the odds of getting Crohn’s. And, the
environment may play a role. For instance, rates of Crohn’s are higher in developed countries, urban areas and northern climates. Finally, the bacteria Mycobacterium avium paratuberculosis and a type of E. coli are linked to Crohn’s.3,4

A risk factor easy to control is smoking. While smoking doesn’t cause Crohn’s, it can make the disease more severe and raises the odds of needing surgery. Nonsteroidal anti-inflammatory drugs such as ibuprofen and naproxen can also make Crohn’s worse.4

Symptoms of Crohn’s

Symptoms of Crohn’s can range from mild to severe and can even be followed by periods of no symptoms that can last weeks to years. Common Crohn’s symptoms include frequent and recurring diarrhea, rectal bleeding, unexplained weight loss, fever, abdominal pain and cramping, fatigue and a feeling of low energy, and reduced appetite. Other symptoms can include mouth sores, pain or drainage near or around the anus due to inflammation from a tunnel into the skin (fistula), constipation and an interruption in menstrual cycle in women.3 In addition, as mentioned previously, Crohn’s can also cause inflammation in areas outside the GI tract such as the eyes, skin and joints. Such symptoms are called extraintestinal.5

For those with mild-to-moderate Crohn’s, symptoms mostly include frequent diarrhea, abdominal pain (with the ability to walk and eat normally), and no signs of dehydration, high fever, abdominal tenderness, painful mass, intestinal obstruction or weight loss of more than 10 percent. Moderate-to-severe symptoms include frequent diarrhea, abdominal pain or tenderness, fever, significant weight loss and significant anemia such as fatigue, shortness of breath, dizziness and headache. Very severe symptoms include high fever, persistent vomiting, evidence of intestinal obstruction (blockage) or abscess (localized infection or collection of pus), and more severe weight loss.3

Symptoms also depend on the type of Crohn’s disease, which is determined by where the disease is located in the GI tract. There are five types of Crohn’s:1

1) Ileocolitis affects the end of the small intestine (the ileum) and the large intestine (the colon). Symptoms include weight loss, diarrhea and cramping or pain in the middle or right lower abdomen.

2) Ileitis affects the ileum, and symptoms are the same as for ileocolitis.

3) Jejunoileitis is characterized by patchy areas of inflammation in the upper half of the small intestine (the jejunum). Symptoms include mild to strong abdominal pain and cramps following meals, as well as diarrhea.

4) Gastroduodenal Crohn’s disease affects the stomach and the beginning of the small intestine (the duodenum). Symptoms include loss of appetite, weight loss, nausea and vomiting.

5) Crohn’s colitis affects the colon. Symptoms include diarrhea, rectal bleeding and disease around the anus (abscess, fistulas, ulcers).

Complications of Crohn’s

Two types of complications can arise due to Crohn’s, including local complications that affect just the intestines and systemic complications that affect the whole body (also known as extraintestinal complications).

Local complications include abscess (a pocket of pus that can form on the walls of the intestine or near the anus), bile salt diarrhea (that occurs because the body can’t process the fat), fissure (a painful tear in the lining of the anus that can cause bleeding during bowel movements), fistula (sores or ulcers that can turn into openings), malabsorption and malnutrition (that occurs when Crohn’s has persisted for a long period and the body is no longer able to make the most of what is eaten), small intestinal bacterial growth (when the bacteria in the gut is higher in the digestive tract) and strictures (narrowed, thickened areas of the intestines).4

A host of systemic complications can occur. These include three types of arthritis (peripheral, axial and ankylosing spondylitis), skin problems (pyoderma gangrenosum, skin tags and mouth ulcers), bone loss (most often caused by steroids), vitamin D deficiency, eye problems (episcleritis, scleritis and uveitis), kidney problems (kidney stones, uric acid stones, hydronephrosis and fistulas), liver problems (fatty liver disease, gallstones, hepatitis and pancreatitis) and physical development problems (growth failure and delayed puberty).4

People who have Crohn’s disease in their large intestine may be more likely to develop colon cancer; however, receiving ongoing treatment to ensure the disease stays in remission reduces the chances of developing colon cancer.2

Managing Crohn’s

Crohn’s is treated with a combination approach. The goal of treatment is to reduce the inflammation that triggers signs and symptoms using preventive care, including medication, surgical procedures and diet.

Medications include anti-inflammatory drugs such as
mesalamine (Asacol, Lialda, Pentasa), olsalazine (Dipentum) and sulfasalazine (Azulfidine), and immune system modifiers such as azathioprine (Imuran) and methotrexate (Rheumatrex). However, the latter can take up to six months to work, and they have a higher risk of causing infections that can be life-threatening. Antibiotics such as ciprofloxacin (Cipro), metronidazole (Flagyl), vancomycin (Vancocin) and antiprotozoal (Alinia) are also commonly prescribed. In addition, corticosteroids, a more powerful type of anti-inflammatory, such as prednisone (Solu-Medrol) and budesonide (Entocort) can be used in conjunction with other therapies; however, side effects can be severe if used long-term.4,6

Moderate to severe Crohn’s disease can also be treated with biologics when other treatments haven’t worked. Unlike other therapies for Crohn’s, biologics aggressively target the particular proteins that cause inflammation in the GI tract. There are three types of biologics to treat Crohn’s: TNF (tumor necrosis factor)-alpha inhibitors, integrin blockers and interleukin blockers — and all are costly.

TNF-alpha inhibitors are sold under the brand names Remicade, Humira and Cimzia, and are given in prefilled pens or syringes that can be self-administered. Because these drugs can’t solely block the immune system from attacking its own tissue while leaving the natural immune responses intact, they increase susceptibility to other diseases and infections, and they can sometimes increase the risk of developing certain cancers.

Integrin blockers include natalizumab (Tysabri) and vedolizumab (Entyvio), which work by interfering with the process of white blood cells attaching to the lining of the intestines to reduce inflammation and relieve other symptoms. Severe and even fatal side effects have been associated with integrin blockers.

Ustekinumab (Stelera) is the only interleukin inhibitor approved by the U.S. Food and Drug Administration to treat Crohn’s. It works by targeting two specific proteins (interleukin-12 and interleukin-23) that are thought to cause inflammation and are found in high levels in people with Crohn’s. Stelera can also increase the risk of infections.7

The Crohn’s and Colitis Foundation estimates two-thirds of people with Crohn’s disease will need surgery to treat complications of the disease or when medications don’t help.4 Surgical procedures include anastomosis (the diseased part of the bowel is removed and the two healthy ends are joined together), ileostomy (connecting the intestine to the skin of the torso, which has an opening where waste products can be collected in a special pouch), small bowel resection (removal of the small bowel), fistulectomy (removal of the fistulous tract), strictureplasty (removal of scar tissue that has built up in the intestinal wall), colectomy (removal of all or part of the colon) and proctocolectomy (removal of the rectum and all or part of the colon).4,6

Dietary modifications, especially during flare-ups, can help reduce disease symptoms and replace lost nutrients. Doctors recommend making changes to diet such as avoiding carbonated drinks; avoiding popcorn, vegetable skins, nuts and other high-fiber foods; drinking more liquids; eating smaller meals more often; and keeping a food diary to help identify foods that cause problems.3

Preventive care is an essential aspect of lifelong disease management. According to a U.S. population-based study that used 2015 and 2016 National Health Interview Survey results, adults with IBD are more likely than adults without IBD to receive preventive care services such as receiving medical advice about smoking cessation and healthy diet, receiving colon cancer screening in the past year, getting an HIV test, receiving the pneumococcal vaccine and flu vaccine in the past year, and having a tetanus vaccine in the past 10 years.8

### Crohn’s Disease vs. Ulcerative Colitis

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<thead>
<tr>
<th>Crohn’s Disease</th>
<th>Ulcerative Colitis</th>
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<tbody>
<tr>
<td>• Inflammation may develop anywhere in the GI tract, from the mouth to the anus</td>
<td>• Limited to the large intestine (colon and rectum)</td>
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<tr>
<td>• Most commonly occurs at the end of the small intestine</td>
<td>• Occurs in the rectum and colon, involving a part of the entire colon</td>
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<td>• May appear in patches</td>
<td>• Appears in a continuous pattern</td>
</tr>
<tr>
<td>• May extend through entire thickness of bowel wall</td>
<td>• Inflammation occurs in innermost lining of the intestine</td>
</tr>
<tr>
<td>• About 67 percent of people in remission will have at least one relapse over a five-year period</td>
<td>• About 30 percent of people in remission will experience a relapse in a year</td>
</tr>
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</table>

However, in 2017, the American College of Gastroenterology published clinical guidelines addressing preventive care in patients with IBD in response to data that suggest IBD patients do not receive preventive services at the same rate as general medical patients. According to the guidelines, patients with IBD often consider their gastroenterologist to be the primary provider of care, but to improve care, health maintenance issues need to be co-managed by both the gastroenterologist and primary care team. Therefore, the guidelines recommend gastroenterologists inform primary care providers of the unique needs of IBD patients, especially those on immunomodulators and biologics or being considered for such therapy. In particular, documentation of up-to-date vaccinations are crucial as IBD patients are often treated with long-term immune-suppressive therapies and may be at increased risk for infections, many of which are preventable with vaccinations. Health maintenance issues addressed in the guideline include identification, safety and appropriate timing of vaccinations; screening for osteoporosis, cervical cancer, melanoma and nonmelanoma skin cancer; identification of depression and anxiety; and smoking cessation.9

Looking Ahead

Due to the vast and growing numbers of people suffering from Crohn’s disease, research to treat and manage the condition is prolific. In fact, there are more than 1,000 studies listed on ClinicalTrials.gov researching medications for Crohn’s. In addition, many organizations are supporting ongoing research initiatives.

The Crohn’s and Colitis Foundation is at the forefront of accelerating research about the disease. Every five years, it convenes leading scientists to update the foundation’s IBD research agenda and identify new priorities. The foundation is currently supporting two major initiatives. The Microbiome Initiative seeks to develop greater understanding of the role of gut microbes (bacteria, viruses, etc., normally found in the intestines) in digestive health and IBDs. And, the Genetics Initiative is a collaborative effort to better understand genes and their functions, and the chain of biological events that result in IBD.10

The foundation is also supporting five studies, including:10

- The Pediatric Risk Stratification Study, which is looking to identify disease prognosis by identifying measurable risk factors for the complications of severe disease;
- IBD Partners, a comprehensive Internet-based registry designed to study thousands of patients with Crohn’s disease or ulcerative colitis under a single research initiative;
- Clinical Research Alliance, a network of major medical centers and smaller facilities collaborating on clinical studies of the management and treatment of IBD;
- The Autoimmune Disease in Pregnancy Study, which is being conducted to learn how Crohn’s and other autoimmune diseases affect the outcome of pregnancies; and
- The PIANO (Pregnancy in Inflammatory Bowel Disease and Neonatal Outcomes) study, which is investigating whether there is a higher rate of adverse events in a prospective national sample of women from the U.S. with IBD who are being treated with biologic drugs.

Two breaking clinical trials results are showing promise:

- In May, SetPoint Medical released results of its successful proof-of-concept study evaluating its bioelectronic medicine approach to treat Crohn’s, which showed clinically meaningful reductions in disease activity in the majority of patients, along with improvements in mucosal healing. The study, which was conducted across five centers in Europe, treated 16 biologic-refractory patients with active Crohn’s disease. All 16 patients were implanted with a vagus nerve stimulating (VNS) device to deliver proprietary doses of electricity designed to activate the innate inflammatory reflex to produce a systemic anti-inflammatory effect and help regulate the immune systems. Patients were separated into two cohorts: The first were washed off their biologic drugs and received only VNS monotherapy, and the second continued their biologic drugs, to which they had inadequate clinical response, in addition to adjunctive VNS therapy.

At 16 weeks, enhanced clinical response (with Crohn’s Disease Activity Index (CDAI) score improvement of 100 or more points) was observed in eight of 16 patients, with four patients achieving CDAI remission (CDAI below 150). On average, levels of serum biomarkers associated with inflammation were reduced compared to baseline, while an anti-
inflammatory cytokine, IL-10, increased from baseline, indicating pharmacodynamic activation of the inflammatory reflex. Patient-reported outcomes indicated a significant improvement in quality of life for seven of the 16 patients who had previously been refractory to biologic therapy. Over the course of the study, 10 patients had an improvement in their autonomic balance, the ratio of sympathetic to vagal tone as measured by heart rate variability, with the shift toward values typically observed in the healthy population.15

• Also in May, results of a study showed people with Crohn’s who worked with their gastroenterologists to earlier detect and address worsening symptoms were more likely to stay out of the hospital and reduced their annual medical cost by $6,500 per person. In the 24-month study conducted by SonarMD and Blue Cross and Blue Shield of Illinois, researchers compared a group of 176 Crohn’s disease patients who were enrolled in SonarMD’s solution to a matched control group that was not. SonarMD contracts with payers to work directly with subspecialists in their network to provide value-based care for patients with high-beta (symptomatic, chronic and likely to deteriorate rapidly, leading to complications, hospitalizations and highly variable per-capita costs) conditions. Its clinical staff uses technology to connect with patients, calculate risk and coordinate care. “The results of this research study demonstrate how an innovative approach to patient engagement and monitoring can improve access to care, health outcomes and affordability,” said Derek Robinson, MD, vice president and chief medical officer at Blue Cross and Blue Shield of Illinois.12

**Crohn’s Impact on Patients**

Crohn’s disease takes a toll on patients both physically and mentally. A study conducted to understand the impact of Crohn’s on various aspects of daily life from the perspective of patients living with the disease found a need for more patient education and more collaborative relationships between patients and providers regarding treatment decisions. Study participants stated Crohn’s caused fear and embarrassment, they were reluctant to share the full impact of the disease with family and providers, and they relied on their provider for treatment decisions. Many participants accepted a new state of normalcy if their current medication helped their most bothersome symptoms without providing sustained remission. Participants receiving biologic therapy generally were more informed, more satisfied and more likely to adhere to treatment regimens.13

More recently, studies have found anxiety and depression are two to three times more likely to occur in IBD patients compared with the general population. In one study, researchers looked at the data of 432 IBD patients within the Intestinal Diseases Natural History Database at the Pennsylvania State College of Medicine in Hershey, Pa. Of these, approximately 44 percent were found to have significant scores on a scale for anxiety or depression, with a majority of them (59 percent) female. About 20 percent had both anxiety and depression. The study also found patients with anxiety and/or depression reported their symptoms to be more severe compared to those who did not have anxiety or depression.14

Fortunately, as more is being discovered about how to manage symptoms of Crohn’s, the search for the cause of the disease continues. It is understood to be an autoimmune reaction, and treatment of the disease is reliant on medication, surgery and diet. Moreover, the American College of Gastroenterology’s recently introduced guidelines for Crohn’s now recognize the additional need for screening for depression and anxiety in these patients. Given Crohn’s increasing prevalence and profound impact on patients, further studies will hopefully yield a more complete understanding of it, leading to more effective treatment. ■

RONALE TUCKER RHODES is the editor of IG Living magazine.

**References**

ELLAS CASANO WAS diagnosed with idiopathic thrombocytopenic purpura (ITP) when she was 7 years old. ITP is an autoimmune disease that causes the body to destroy its own platelets, putting patients at risk for bleeding or injury. Ella’s diagnosis requires her to receive intravenous immune globulin (IVIG) infusions every eight weeks, a prospect that was initially daunting for a young child. But, in a classic “turn-lemons-into-lemonade” scenario, her childhood fear of the IV bag became the inspiration for an invention that is now bringing comfort to children around the country. We chatted with Ella’s mom, Meg Casano, to learn more about her entrepreneurial daughter and her innovative idea: Medi Teddy.

**IG Living:** Tell us a little bit about Ella’s condition; when did you know something was wrong?

**Meg:** When Ella was younger, we started to notice huge bruises on her body. One day after climbing a rope ladder, the entire insides of her legs and thighs were a huge dark bruise. Every morning, she would have blood on her pillow because her nose and gums would be oozing. Her bruises seemed unusual based on her activities. Her pediatrician suggested we have her blood work checked, and we found out her platelet count was only 13,000.

**IG Living:** How long did it take to get a diagnosis?

**Meg:** I became concerned about the bruising when she was 2 years old. We actually saw three pediatricians in two states seeking answers, only to be told she was just “an active kid.” I finally found our current pediatrician who really listened to me and checked her blood. It was a great lesson for me to trust my gut and not be afraid to get second (and third and fourth) opinions! We believe she may have had ITP for several years before she was actually diagnosed at age 7.

**IG Living:** What is her treatment plan with IVIG?

**Meg:** It will likely change over time, but for now she gets IVIG approximately every eight weeks at our local infusion center. She does experience migraines from the IVIG, so she gets a steroid taper for six days after each infusion to help mitigate those, but she still misses two to three days of school after each infusion.

**IG Living:** How is Ella’s health today?

**Meg:** Between infusions, she is a normal kid! She’s active in her school and church community, and she is a great student. The IVIG lasts for about four weeks, and then her platelets begin to dip very low again over the next few weeks. Right now, she only gets infusions when her platelet count is below 10,000. So, there are several weeks every cycle when she is walking around with dangerously low platelet counts and is restricted in her activities.

**IG Living:** What inspired the creation of Medi Teddy?

**Meg:** Ella had to make an invention for school, and she wanted to create something unique. She liked the idea of coming up with something that could help her and other kids like her, so she started brainstorming ideas. For Ella, one of the scariest parts of infusions is seeing the big bags of medicine hanging on her pole. It’s funny because most adults focus on the IV insertion; we forget the equipment itself can be very overwhelming. Ella wanted to hide the bags of medicine, and that’s how Medi Teddy was created!
She made the first prototype by cutting up a stuffed penguin and using a hot glue gun to make a little pouch on the back but later decided a teddy bear was a more natural fit with the name.

**IG Living:** Tell us about your fundraising efforts.

**Meg:** We started a Go Fund Me account with the hope of raising $5,000 from our local community so Ella could have 500 Medi Teddys manufactured. We figured we’d be driving them around in the back of my car and begging kids to try them out. We had no idea Medi Teddy would become almost instantly accepted worldwide and more than quadruple our fundraising effort. What was so amazing to us was the sheer number of people who donated. It wasn’t like there were a few huge donors; there were thousands of people who donated small amounts and, together, it added up to more than we ever dreamed.

**IG Living:** What is the goal with the Medi Teddy nonprofit?

**Meg:** Our goal continues to be to give Medi Teddys to children who would like one so they feel more comforted during IV infusions.

**IG Living:** What advice do you have for other families facing chronic illness?

**Meg:** Oh, this is a hard one. I happen to have Crohn’s disease myself and get bimonthly infusions, too, so it’s been really hard to see my daughter have to go through the same thing. Chronic illness can wear you down if you let it, and this is something I have struggled to get better at with time. I tell myself and Ella it’s OK to have hard days, take days off and go easy on ourselves. It’s OK to not feel like that “fighter” everyone thinks you are every once in a while. Just be sure you don’t stay in that place; shake it off, get help if you need to and find some things to focus on that you feel good about, because that always helps. And, I think for both of us, Medi Teddy has been incredibly therapeutic. We’ve heard from thousands of people going through the same things we are, and it has created the most incredible feeling of connection.

**IG Living:** What are Ella’s hopes for the future?

**Meg:** Right now, she says she wants to be a doctor, specifically a hematologist, so she can help other kids with blood disorders like her. I know she would be really good at that. ■

Editor’s note: To purchase a Medi Teddy or to make a donation to have a Medi Teddy sent to a child, visit medi-teddy.org.

**Trudie Mitschang** is a contributing writer for IG Living magazine.
Find the Joy in Each Day
By Stacy Oliver

I was feeling a bit exasperated. I had received some sourdough bread yeast years ago, but it had to be fed every week. It was starting to weigh on me. Like having another pet to feed. I just didn’t want to do it anymore. My husband said: “Throw it out.” What? “We can always get more if you want to start again. In the meantime, don’t do it. It shouldn’t be stressful. Only do what makes you happy.”

I paused for a second and threw the sourdough starter out. I felt so relieved. Like a weight was taken off my shoulders. Time is precious, and I was going to follow his advice. No one knows how much time they have, but when you’re told you are dying and you feel it happening, it’s a game-changer.

Last August, I was diagnosed with multiple system atrophy-C (MSA-C). The doctors have never seen such rare neurological cases in one person. I have continued to decline. It is harder for me to type, walk, get up and concentrate. I have erratic blood pressure, and it takes quite a while for my day to start so I don’t pass out. On that lovely note, I’ve decided this is my last article. Thank you for letting me share my thoughts on intravenous immune globulin all these years. It has truly been a privilege. However, it’s time to move on. I have some things I want to do. People to see. I can’t have any steadfast deadlines anymore.

I’m going to follow my husband’s advice and each day find the joy and do what makes me happy. Whether I’m in a wheelchair or bedbound, before then and even more so, I’m going to cherish the moments:
The breeze on my face;
Being in the water;
Listening to classical music;
Food, while I can eat and taste;
Moving my body.

If I may give one last word of fun advice: Eat the doughnut, be crazy, wear the purple wig, put sugar sprinkles on everything, write people “just because,” say “I love you” and be in the now.

All we have is now.
I wish you well. I hope we meet again. Until then, I’ll be making soapy bubbles in the air with wands and petting my dogs.

Editor’s note: Sadly, Stacy’s contribution to our magazine ends with this issue so she can devote all her energy to battling her illness and cherishing life’s moments. The staff and readers of IG Living will fondly remember Stacy and her columns that were so often humorous and full of sage words of advice.

Stacy Oliver was diagnosed in 2008 with multifocal motor neuropathy. When she isn’t writing her book, herding three pit bulls or trying to put eyeliner on straight, she is working on her super secret identity as Neuropathy Girl, who will one day save the world after an infusion and a nap.
I HAVE BEEN making dream boards of my hopes and goals and sticking them to my bathroom wall since I was a teenager. I keep them there to remind me of my goals and all the big, shiny things I want out of life. But, instead of bringing me the excitement and motivation I am looking for, they sometimes make me a little sad to think of the big beautiful life I want but can’t have because of my disease.

So, I’m flushing these boards down the toilet, and I am Marie Kondo-ing all the pseudo motivation that does not spark joy. Instead, I’m going to tell myself all the things I’ve been failing at so I can remind myself to change my bad habits. After all, there are so many things I’m reminded I can’t do, but I can make an effort to change other things within my control.

Here are three of my chronically bad habits:

1) Not consistently being in therapy. I wrote about mental health in a previous column, but whether you are a parent, caregiver, sibling or spouse of a patient with a chronic illness, or you are the patient with a chronic illness, therapy can be a useful tool for getting through bad days and celebrating good days. When I found the right therapist and began going on a regular basis, I was better. Not better in the sense that my health was better, but in the sense that I had strengthened by ability to cope with disappointments and frustrations about a disease that will likely always be a part of me. I haven’t always been consistent in making my mental health a priority, but moving forward, I’m going to accept the help I need to get me through the hard times and give me tools to practice in the interim to prepare me for what’s ahead.

2) Relying too much on technology rather than people. In my early 20s, I was dying for independence. But, with a chronic illness, even the most monotonous daily tasks were challenging during flares. I was over the moon when I realized I could use apps to do a lot of the heavy lifting for me. It gave my caregivers a break, and I didn’t have to beg for rides when I couldn’t drive or get food because I physically couldn’t make it to the grocery store. However, I started relying on these services so much that I didn’t accept my friend’s and family’s offers to get out of the house or to do these things together. Just because I could Uber to appointments didn’t mean it wasn’t better for me emotionally to have a friend present during tough appointments. While I don’t want to burden my loved ones, when they offer out of the kindness of their hearts, I can do more to accept.

3) Mercy for me. The last habit I’m promising to break is consistently blaming myself for everything that goes wrong with my disease: the infections, surgeries, flares. I never had control over these things in the first place. What I actually had were choices. Rarely were any of those choices “good” or “right,” they were simply executive decisions made for survival. Ripping into myself when something goes wrong that I can’t influence or direct isn’t productive, and showing myself some mercy in the face of an already impossible task is the kindest thing I can do.

Have you uncovered some chronically bad habits of your own? What will you be doing to break the cycle moving forward?  

ILANA JACQUELINE is a 29-year-old dysautonomia and primary immune deficiency disease patient from South Florida. She’s been writing professionally since 2004 on everything from health and wellness to celebrities and beauty. Her blog www.letsfeelbetter.com is both a personal collection of anecdotes about life with chronic illness, as well as a resource for patients of all ages.
CHILDREN WHO suffer from chronic illness such as a primary immunodeficiency (PI) are no strangers to hospitals. Whether they visit monthly for infusions or have to stay for longer periods while recovering from a serious infection or surgery, these children generally spend more time in the hospital than the average child. But, just because they’ve grown up around doctors and nurses and have spent countless hours walking hospital halls, there is no need for these kids to be bored while waiting for their next appointment or to be fearful of an upcoming procedure.

What is a child life specialist? As early as the 1920s and 1930s, hospitals began to develop play programs for children — both patients and visitors. In 1955, the first Child Life and Education division was created by Emma Plank at Cleveland City Hospital to meet the unique needs of children in a hospital setting. Today, with more than 400 child life programs in North America alone, what began as a play program has turned into “a quality benchmark of an integrated patient- and family-centered healthcare system, a recommended component of medical education and an indicator of excellence in pediatric care.”

While it’s typical for larger hospitals specializing in pediatric care to include child life programs, these services are recommended and offered in smaller community hospitals with pediatric units, as well as in walk-in clinics, emergency departments and physician offices.

The purpose of a child life program is to address the psychosocial concerns surrounding children and their families during a hospitalization or other healthcare experience. While caring for these infants, children and adolescents, child life specialists teach coping skills and other ways of minimizing the adverse effects of hospitalization and stress-inducing medical procedures — all while focusing on the development and well-being of the patients.

Child life specialists, working alongside the children’s healthcare teams and their families, use a combination of therapeutic play and modes of expression (such as art, drawing, sculpture, collage, dance, movement and language) as primary tools to make unfamiliar circumstances seem less overwhelming. Play is an essential component of a child life program and of the child life professional’s role, and is adapted to address the unique needs of each patient. We already know playing benefits children developmentally while being a normal and familiar activity for them, but play in a hospital setting is particularly beneficial for children who are anxious or struggling to cope with stressful circumstances. One specific way a child life specialist might intervene in a stressful situation is to distract a child with a game on an iPad while he or she is having blood drawn. My son learned to play Fruit
Ninja with his child life specialist while having medical procedures performed in the hospital.

Medical play is another tool used by child life specialists to help prepare children for upcoming procedures that may frighten them or with which they’re unfamiliar. When my son Andy was in the hospital for an extended period of time, he had a double-lumen line, or Hickman catheter, placed in his chest. His child life specialist brought him a doll with the exact same tubes sewn into its chest, and used the doll to demonstrate exactly what would happen when the nurses came to draw blood from the tubes or infuse medicine through them. This helped Andy to be prepared for the procedures by removing the fear of the unknown.

At many of the nation’s larger hospitals, the child and family services programs and arts programs are joining forces to make young patients’ lives better. Boston Children’s Hospital has what they call the Creative Arts Program, which is a collaboration between Child Life Services and the Art Program that provides “engaging, innovative and uplifting arts experiences that support patients and families throughout their hospital experience.”

Benefits of child-focused hospital programs. More than 50 years of research and experience shows children who participate in programs that provide developmentally appropriate information, encourage them to ask questions and express their emotions, allow them to form a trusting relationship with a healthcare professional, and prepare them for surgery experience fewer negative symptoms than children who do not receive this preparation. Not only is their anxiety reduced and a more positive experience is shared by patients and their families, but research shows preparation and coping interventions actually decrease the need for sedation in procedures such as MRIs, resulting in lower health risks for children and cost savings for both the hospital/facility and families.

A survey of parents and patients at Boston Children’s Hospital revealed the arts program decreases patients’ perceived pain and significantly decreases both patient and family anxiety. According to the parents surveyed, 75 percent said their child had reduced anxiety, and 60 percent said their child had less perceived pain. Of the children surveyed, 71 percent reported reduced anxiety, and 59 percent reported less perceived pain. One parent surveyed said (about the Creative Arts Program), “The program helps children feel more relaxed in an atmosphere that is not always without pain and sometimes fear of the unknown. It brings some normalcy to their stay.”

The therapeutic play that is such a huge part of child life programs has been shown to have noticeably positive results in children’s emotional well-being both before and after medical procedures and hospital stays. Play as a therapy tool, including medical play, has been found to help children cope with their medical experience and reduce any emotional stress resulting from it. Research has shown therapeutic play interventions can help reduce physiologic responses such as palm sweating, excessive body movement, tachycardia (heart rate that exceeds normal resting heart rate) and hypertension.

Helping children prepare for an upcoming surgery, hospitalization or diagnostic/therapeutic procedure is another beneficial element of a child life program. It is estimated 50 percent to 75 percent of children develop significant fear and anxiety before surgery, and this risk increases with factors such as younger age, anxious temperament, negative experiences with past medical procedures, and parents’ heightened anxiety level. In fact, there is a direct correlation between a child’s anxiety level pre-operation and his or her post-operative behavior and recovery time, including an increased need for pain killers after surgery and delayed discharge from the recovery room.

If your child has a surgery or medical procedure in his or her future, it would be worth asking what kinds of programs the hospital provides for children and their families. Once a child establishes a relationship with a professional such as a child life specialist, that relationship can continue for years and extend to younger siblings as well. Even during short check-ups or follow-up appointments for our kids, the child life specialist may pop in just to say hi and see how things are going. This continuous relationship can make the hospital seem a lot less intimidating and a little bit more like home.

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

References
The Ins and Outs of Port Maintenance

By Heather Bremner Claverie

**FOR PATIENTS WITH** chronic conditions requiring treatment with intravenous immune globulin (IVIG), needles and in-office infusions are often a part of the monthly calendar. But for some patients — whether they have difficult-to-access veins, need long-term venous access or just don’t want to deal with the inconvenience of constant needle sticks — implanted infusion ports are a possible option. Also referred to as a central venous access device, an implanted infusion port is a convenient tool that improves patient care by enabling caretakers to infuse medicines easier with fewer needles in the process. The small device is placed under the skin — usually in the upper chest, but it can also be placed in the arm or abdomen — and is attached to a catheter that enters a large vein.

**Why a Port?**

Convenient and comfortable, many patients find infusion ports more convenient and less painful. Patients can swim, bathe and exercise freely while fitted with one. However, there are some inherent dangers associated with the devices. If a port is not maintained correctly, complications such as blood clots and infections can seriously jeopardize patients’ health. Infections, including life-threatening bacterial infections that travel from the device to the bloodstream, can arise if the port is not cleaned properly. This is why the decision to obtain a port is a collaborative one between patients and their prescribing physicians. In some cases, prescribers may not allow patients with a primary immunodeficiency to have a port due to this increased risk of infection.

**Tips for Care**

Physician or home care agencies may have differing protocols for port maintenance. Therefore, the following tips serve as general information and are not a substitute for maintenance and care as ordered by treating physicians. And, unless patients are adequately trained in port maintenance and access, all care should be provided by a trained clinical professional such as a home care or infusion center nurse.

For self-care, washing one’s hands can help prevent the spread of most pathogens. According to the Centers for Disease Control and Prevention (CDC), hands should be washed for at least 20 seconds with antibacterial soap before and after an infusion, whenever a caregiver enters the room and before and after preparing food. Patients should also wash their hands often, and especially before and after handling the port.

In addition, sterile techniques should be used whenever accessing or de-accessing the port. This means donning sterile medical gloves whenever touching the port, and cleaning the skin around the site prior to port access. The port site should be assessed daily for any signs of infection such as redness, swelling, tenderness or fluids. When accessed, the needle and dressing may be left in place for up to seven days. But, the dressing should be changed at any time it becomes damp or visibly dirty. Dressing changes should be completed by a nurse until patients are trained and competent in maintaining sterile technique.

**Flushing the Port**

Ensuring the port is flushed according to the guidelines established by the manufacturer of the port and/or the protocol prescribed by prescribers is essential to maintaining a functioning port. The frequency of this flushing can vary depending on the source. According to a 2017 *Journal of Infusion Nursing* article, an eight-week interval is ideal: “Guidelines for maintenance flushing of ports not in use provided recommendations of flushing every four to eight weeks, but no evidence existed to support optimal intervals.… The eight-week interval for maintenance port flushing was well received by patients who appreciated the reprieve from the monthly flush visit. By using the maximum interval recommended in the guidelines instead of the conservative four-week interval, the patient experience was improved without an increase in patient risk for occlusion.”

To properly flush the port, a 10 mL or larger syringe should be used (a smaller syringe could cause the catheter to burst). All ports must be flushed with a saline solution before beginning the infusion and then, once complete, flushed again with saline. Some ports may require a final “lock” with heparin, a medication used to decrease blood clotting.

**Opting for Convenience and Comfort**

Fewer needles, less visits to the doctor office and fewer restrictions give patients the ability to carry on with their lives even while in the midst of treatments. For individuals who need frequent infusions, an implanted port is a potentially life-changing option. 

**HEATHER BREMNER CLAVERIE** is a contributing writer for *IG Living* magazine.
**Numb It**

A topical anesthetic cream such as Ebanel Numb520 can be used at the port site to numb the area before inserting a needle. The anesthetic is comprised of equal parts of lidocaine and prilocaine. These creams work by blocking nerve signals on the area of the body it is placed on. The cream will begin its numbing power about 15 minutes after application, and it is most effective after being on the skin for two hours to four hours. These creams are available over the counter. $16.50; [amazon.com](http://amazon.com)

**Combat Infections**

The Central Line Dressing Kit can minimize the possibility of infections. This kit streamlines the sterilization process and includes everything needed to lessen the risk of infection, including transparent tape, alcohol swab sticks, one skin wipe, one blue face mask with ear loops, one pair of medium aloe touch latex-free gloves, gauze, dressings and one Chloraprep application with insert. $124.98 for the set; [amazon.com](http://amazon.com)

**Prep and Clean**

Current guidelines recommend cleaning the port site with chlorhexidine in alcohol solution. One option is the Chloraprep Sponge Applicator that contains 2% chlorhexidine in alcohol and is a rapid-acting antimicrobial that kills a broad array of microorganisms and remains active for 48 hours after cleansing. It’s effective against gram-positive and gram-negative bacteria, including Methicillin-resistant Staphylococcus aureus, vancomycin-resistant Enterococci, Clostridium difficile, Acinetobacter and most viruses and fungi. $79 for a box of 25; [amazon.com](http://amazon.com)

**Shopping Guide to Infusion Aids**

**Implant Power**

BD’s PowerPort Implantable Port combines reliable venous access with power-injection capability. Power-injected contrast-enhanced computed tomography scans produce enhanced images, improving the ability to track tumor markers or perform pulmonary embolism studies. The device’s lightweight, small and flexible design make it ideal for patient comfort and ease. Prices vary based on coverage. Contact a doctor for more information. [www.portready.com/ports.php](http://www.portready.com/ports.php)

**Prepped Syringes**

Prefilled flush syringes are convenient and help reduce the risk of medication errors. They contain saline, which helps keep the infusion port clean, and heparin, which helps prevent blood clots from forming. Saline is required for the maintenance of all infusion ports, while some ports do not require maintenance with heparin. These prefilled syringes are an alternative to vial-based flushing systems and require a prescription. [www.bd.com/en-us/offers/capabilities/syringes-and-needles/pre-filled-flush-syringes](http://www.bd.com/en-us/offers/capabilities/syringes-and-needles/pre-filled-flush-syringes)

**Protect Patients**

A total of 50 patients die every day in U.S. hospitals due to bloodstream infections, according to CDC. There are several dressings that contain integrated chlorhexidine. One option is the BioPatch Disk with integrated chlorhexidine gluconate proven in multiple trials to reduce the incidence of catheter-related bloodstream infections in central venous and arterial catheters. This dressing provides protection around the insertion site for up to seven days. $382.69 for a case of 40 (4 boxes of 10 disks); [amazon.com](http://amazon.com)
Pulmonary Manifestations of Primary Immunodeficiency Diseases, 1st Edition
Editors: Seyed Alireza Mahdaviani and Nima Rezaei
Publisher: Springer

This book provides a broad overview of the respiratory manifestations associated with primary immunodeficiencies (PIs) such as infections, chronic inflammation, autoimmunity, lymphoproliferation, allergic manifestations, and rare forms of cancer. Since the most common site of involvement in PI is the lung, pulmonologists (pediatrics or adult), internists and general practitioners may be among the first to recognize the pattern of pulmonary disorders leading to diagnosis of PI. With the prevalence of lung infections and disease so high in PI patients, respiratory professionals will find this book to be an essential resource for diagnosing, managing and referring PI-related pulmonary disorders in clinical practice.

Identity Theft: Rediscovering Ourselves After Stroke
Authors: Debra E. Meyerson and Danny Zuckerman
Publisher: Andrews McMeel Publishing

Identity Theft follows Stanford professor Debra Meyerson’s journey to recovery from a severe stroke that initially left her physically incapacitated and unable to speak. In addition to providing realistic expectations for the hard work needed to regain everyday capabilities, Meyerson focuses on the less frequently documented emotional journey in recovery. According to Meyerson, virtually every survivor is haunted by questions like: “Who am I now?” and “How do I rebuild a meaningful and rewarding life?” after losing so much of what they had before — capabilities, careers and jobs, relationships and more. This is a book full of hope for survivors — from stroke or other injuries — as well as their families and support networks.

Hashimoto’s Food Pharmacology: Nutrition Protocols and Healing Recipes to Take Charge of Your Thyroid Health
Author: Izabella Wentz, PharmD, FASCP
Publisher: HarperOne

Dr. Izabella Wentz, who was diagnosed with Hashimoto’s thyroiditis in 2009, is one of the pioneering experts in lifestyle interventions for treating the disease. Trained as a clinical pharmacist, Dr. Wentz was surprised at the lack of knowledge about lifestyle interventions for Hashimoto’s and autoimmune conditions, so she decided to take on lifestyle interventions as a personal mission in an effort to help herself and others with the same condition. This book is written for those who are ready to take charge of their own health and need a friendly guide that can provide the tools and confidence to optimize their nutrition, as well as delicious recipes that don’t require them to spend all day in a kitchen. Through this powerful nutrition guide, you can discover how to save time in the kitchen, use food as medicine and recover your health.

Maldynia: Multidisciplinary Perspectives on the Illness of Chronic Pain
Editor: James Giordano
Publisher: CRC Press

Maldynia: Multidisciplinary Perspectives on the Illness of Chronic Pain is about chronic pain that has progressed to a multidimensional illness state. Taking a comprehensive approach that covers science, humanities and culture, this book emphasizes the need for researchers, clinicians and caregivers to regard the ways in which chronic intractable pain becomes illness and affects a patient’s biological, social and psychological states, as well as his or her sense of self. Edited by neuroscientist and neuroethicist James Giordano, the book contains 17 insightful chapters representing medicine, neuroscience, psychology, philosophy, ethics, history, art and the ministry.
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
- The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/Pages/index.aspx

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 National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/Pages/index.aspx
- The Jeffrey Modell Foundation: www.info4pi.org
- Michigan Immunodeficiency Foundation: www.michiganimmunodeficiencyfoundation.org

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

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

Mitochondrial Disease
WEBSITES
- United Mitochondrial Disease Foundation: www.umdf.org
- Mitochon: 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.multi-sclerosis.org/index.html
- Multiple Sclerosis Association of America: mymsaa.org
- Multiple Sclerosis Foundation: www.msfocus.org
- National Multiple Sclerosis Society: www.nationalmssociety.org

Myopathy 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
- The Cure JM Foundation: www.curejm.org
- 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.pandanetwork.org
- Midwest PANS/PANDAS Support Group: www.midwestpandasm.com

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

Primary Immune Deficiency Disease (PI)
WEBSITES
- Immune Deficiency Foundation: www.primaryimmune.org
- American Academy of Allergy, Asthma & Immunology: www.aaaai.org
- American Autoimmune Related Diseases Association Inc.: www.aarda.org
- Genetic Alliance: www.geneticalliance.org
- Living with Stiff Person Syndrome (personal account): www.livingwithspss.com
- Stiff Person Syndrome: www.stiffpersonsindrome.net

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

Primary Immune Deficiency Disease (PI)
WEBSITES
- Immune Deficiency Foundation: www.primaryimmune.org
- Jeffrey Modell Foundation: www.info4pi.org
- The International Pemphigus and Pemphigoid Foundation: www.pemphigus.org
- The Foundation for Peripheral Neuropathy: www.foundationforpn.com

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- Immune Deficiency Foundation: www.primaryimmune.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 Immune Deficiencies (IPOPI) — UK: www.ipopi.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.primaryimmune.org/idf-peer-support-program
- Michigan Immunodeficiency Foundation: www.michiganimmunodeficiencyfoundation.org

Scleroderma
WEBSITES
- Scleroderma Foundation: www.scleroderma.org
- Scleroderma Research Foundation: www.srfcure.org
- Johns Hopkins Scleroderma Center: www.hopkinsscleroderma.org
- The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/Pages/index.aspx

ONLINE PEER SUPPORT
- Scleroderma Support Forum: curezone.com/forums/r.asp?f=404
- International Scleroderma Network: www.sclero.org/support/forums/a-to-z.html

Stiff Person Syndrome (SPS)
WEBSITES
- American Autoimmune Related Diseases Association Inc.: www.aarda.org
- Genetic Alliance: www.geneticalliance.org
- Living with Stiff Person Syndrome (personal account): www.livingwithspss.com
- Stiff Person Syndrome: www.stiffpersonsindrome.net
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E: customerservice@fffenterprises.com

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