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It’s All About Making Adjustments

AS IF LIFE weren’t already difficult for people with chronic illness, these past few months has made it a lot more challenging and anxiety-ridden. Having an impaired immune system already requires limiting activities in public and dealing with concerns about what could happen when exposed to viruses that for most don’t pose the same threats. Enter COVID-19, and while those same threats now apply to all, they are greatly compounded for primary immunodeficiency patients and others with weakened immune systems. But, as Erika Lawrence, PhD, a licensed clinical psychologist, explains in her Therapeutic Helpline column “Coping with Anxiety During the COVID-19 Pandemic” (p.10), “while there are a lot of things you cannot control right now, there are some things within your power to manage.” Dr. Lawrence’s column and other articles in this issue show how adjusting to factors that can affect individuals’ physical and mental health can help to minimize their impact.

Foremost, a number of lifestyle changes can affect the body’s response to infection. Our article “Factors That Can Impact Immunity” (p.22) focuses on ways individuals with compromised immune systems can improve their bodies’ response to a contagion. Yes, they may seem like common sense, but repeatedly washing hands (which, surprisingly, most people don’t do), minimizing stress (which interferes with the production of T cells and white blood cells), getting enough sleep (which makes people more likely to get sick), avoiding environmental hazards such as air pollution and smoking (which affect people’s immune responses), as well as eating well and getting enough exercise are more important than ever.

There are also other adjustments people can make that are detailed in our article “Adjusting to Life with a Chronic Illness: 3 Things to Do After Diagnosis” (p.26). Especially when faced with “shelter-in-place” orders that require social isolation, building a support system is critical. Even if it’s not face-to-face, there are ways to connect with others (friends, family, doctors, etc.) via social media, telephone and other technologies. Again, stress can be minimized with exercise, breathing exercises, meditation, mindfulness and other activities. And, during times of uncertainty, planning for the future in terms of long-term care, property and estate management, retirement savings and more can put people’s minds at rest.

Finally, since individuals with chronic illnesses visit doctors often, now might be the time to embrace telemedicine. As our patient advocate Abbie Cornett explains in her column “The Emergence of Telemedicine During COVID-19” (p.6), this virus is changing the way healthcare will be practiced as patients no longer need to always be seen in person, but rather can opt for care via phone, video chat and direct messaging.

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.
The Emergence of Telemedicine During COVID-19

By Abbie Cornett

The recent outbreak of COVID-19 has forced people around the world to make changes in their everyday lives. But, perhaps one of the most significant modifications in our new normal is how people interact with one another due to the threat of either catching or spreading the virus. This includes family and friends, as well as healthcare providers.

For the last several years, policymakers and healthcare providers viewed the implementation of telemedicine as a way to keep patients with nonurgent health issues out of emergency departments and provide services to areas that are typically underserved. And, while the use of telemedicine has been increasing, it still has not been utilized by the majority of patients or healthcare providers, nor has its use been widely promoted.

But, COVID-19 is changing that. Because this virus has placed additional stress on an already overburdened health system, the Centers for Disease Control and Prevention (CDC) began recommending in March the use of telemedicine to direct patients to the right site of care during the outbreak.

Telemedicine wasn’t traditionally created to respond to a pandemic, but it was quickly apparent it would be a way to reduce exposure to COVID-19 for patients and healthcare providers. Therefore, when CDC called on healthcare facilities to adopt telemedicine to protect patients and staff, many large hospitals raced to implement and scale up these capabilities. In addition, hospitals and public officials urged patients with symptoms of COVID-19 to avoid going to emergency rooms and doctor offices. Instead, patients were encouraged to consult with their doctors over the phone or using video chat or direct messaging.

To encourage the use of telemedicine, many private insurance providers informed their policyholders that telehealth screenings would be covered for free under their plans. Further, Congress approved a $500 million appropriations bill for the use of telemedicine services in an emergency aid package. The bill also waived some Medicare restrictions on payments for telemedicine so seniors could use remote services for COVID-19 treatment.

Undoubtedly, telemedicine reduces the burden on hospitals and healthcare providers particularly during a public health crisis by allowing for initial screenings, nonurgent communication with physicians and triaging of patients. It does have limitations, though. While appropriate for patients who are medically stable, telemedicine does not allow for diagnostic testing such as lab work, nor does it allow clinicians to perform physical exams.

Prior to COVID-19, public awareness of what telemedicine services were available and how they could be used was limited. This virus changed that. Usage of telemedicine apps increased dramatically, and some healthcare apps like Amwell, HeyDoctor by GoodRx and PlushCare are showing tremendous growth. According to Amwell Vice President Holly Spring, “In a matter of days, nearly all of healthcare’s key stakeholders started to direct people in need of doctor consultations and care to telehealth.”

Unfortunately, the dramatic increase in the utilization of telemedicine showed the technology was not fully prepared to handle the massive increase in demand. While some hospitals and large medical practices were set up to provide care in this manner, the majority were not, leaving many providers playing catch-up and rethinking the traditional patient-provider relationship. In addition, medical providers had to learn how to use the technology and figure out billing and payer telehealth policies before they could start seeing patients in this manner.

When life returns to normal and the pandemic is in our rear-view mirror, telemedicine will be here to stay, which is a good thing. It has proven to be an excellent resource for both patients and providers.

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


How do you set goals for your chronic illness?

My goal is simply to wake up in the morning alive. That enough is mission accomplished.
— Peggy SG

I set realistic goals that I have a great chance of achieving. I have many setbacks due to the side effects of the medications I take for myasthenia gravis. Without setting goals and achieving them, it would be more difficult getting through the rough times.
— Judy S

Sometimes my goal is just getting out of the house. I do have a longer-term goal of finishing school. I am becoming a long-term short-term goal-list person. I never thought I would say that. And, it stays on the refrigerator.
— Jane N

Is your infusion nurse properly trained?

I quit doing intravenous immune globulin (IG) infusions at a hospital because some (not all) of the nurses didn’t practice aseptic technique with starting my IV. Also, the patient chairs were crammed side by side, and the chairs and attached trays were never wiped down. I took my own Lysol wipes and did it. Long story short, I got trained to do my own subcutaneous IG treatments, and I happily stay in my clean house and do my own infusions.
— Charlene M

I can’t find a hospital, infusion center or urgent care in my area that has any trained staff to do subcutaneous immune globulin infusions. I had an anaphylactic reaction to Hizentra in November, and my doctor says I cannot have another infusion unless it is in a medical setting. We still haven’t found a facility that will allow me as a patient for infusions. They all claim not to have staff trained in subcutaneous infusions. So, I have been without infusions for three months.
— Lydia LP

Mine was not. This was when I started subcutaneous infusions in my home. She had never done this before, and I had to ask for video help while [she was] here. I ended up with cellulitis on my abdomen. I’m a registered nurse, and this was unacceptable.
— Sandy P

No, my nurse last month said she’d never done an immune globulin infusion before. [She had] never heard of my rare immunodeficiency (common variable immunodeficiency) until she came to do my infusion. When I changed [specialty infusion companies], all the nurses were trained in immune globulin infusions. Such a difference.
— Jerri O

Yes, [they are] trained in all aspects of intravenous immune globulin infusions, including adjusting the rate, which makes a huge difference for post-infusion side effects.
— Laura PP
Abbie » I spoke with a couple of our experts regarding your question. They said they have treated patients with multiple myeloma and high IgA who do well with IG treatment, and there should be no major issues with IG therapy, especially if it is infused subcutaneously (SCIG). With SCIG, the most commonly reported side effects are injection-site reactions, swelling, redness and/or irritation that occur at the needle insertion site and are typically localized. These reactions usually resolve within 24 hours to 48 hours after the infusion is completed. They are more common in the first several infusions, and their incidence normally decreases over time.

Question » Will immune globulin replacement therapy cause severe side effects in patients with polyclonal IgA hypergammaglobulinemia?

I have been diagnosed with specific antibody deficiency, IgM deficiency and polyclonal IgA hypergammaglobulinemia. Despite taking prophylactic antibiotics for the last few years, I continue to be ill. I have been prescribed antibiotics, antivirals or antifungals 64 times in the last three years. My current immunologist and I have discussed immune globulin (IG) replacement therapy, but she is worried that because of my polyclonal IgA hypergammaglobulinemia, I would suffer unacceptable side effects. My IgA levels are in the high 300s to middle 400s. My IgG level is below normal but not overly so. Are the possible side effects to IG therapy extreme, and can they be managed?

Abbie » I spoke with Michelle Greer, RN, senior vice president of Nufactor, a Specialty Infusion Company, and she said the short answer is yes. Gammagard is dosed by the patient’s body weight. The typical dosing is 2 grams per kilogram. However, some physicians may prescribe a little less or a total gram amount, but dosing would still be close to 2 grams per kilogram. Total grams are then divided over a few days, depending on the size of the dose, which is better for tolerability. For example, if the patient weighs 75 kilograms, the total dose of Gammagard would be 150 grams. That patient could then be infused with 50 grams per day for three days, or if the patient is more prone to reactions, he or she could be infused with 30 grams per day for five days.

Question » How is the dose of Gammagard (intravenous immune globulin) calculated for myasthenia gravis patients?

I am a patient with generalized myasthenia gravis, and I would like to know how the dosage for Gammagard is calculated. Is it by body weight?

Abbie » I spoke with Michelle Greer, RN, senior vice president of Nufactor, a Specialty Infusion Company, and she said the short answer is yes. Gammagard is dosed by the patient’s body weight. The typical dosing is 2 grams per kilogram. However, some physicians may prescribe a little less or a total gram amount, but dosing would still be close to 2 grams per kilogram. Total grams are then divided over a few days, depending on the size of the dose, which is better for tolerability. For example, if the patient weighs 75 kilograms, the total dose of Gammagard would be 150 grams. That patient could then be infused with 50 grams per day for three days, or if the patient is more prone to reactions, he or she could be infused with 30 grams per day for five days.

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

ABBIE CORNETT is the patient advocate for IG Living magazine.
Type I Hypersensitivity: ‘True’ Allergic Disease, Part 3

By Terry O. Harville, MD, PhD

IN PREVIOUS issues, we have been discussing the differences between hypersensitivities and allergic diseases, focusing on the “true” allergic disease known as type I hypersensitivity, which is IgE-mediated. We noted that most of the aeroallergens (airborne allergy-inducing proteins) that we may breathe in or that may come into contact with our eyes contain digestive enzymes found in plant pollens, mold spores and animal danders. When this happens, our body and immune system perceive a parasitic attack is occurring (something is trying to eat us) and, thus, turns on the Th2 system of immunity. Th2 immunity evolved to protect us from parasites, but in our “sanitized” world has led to the development of allergic disease (known as the hygiene hypothesis). While this concept works well for aeroallergens that come in contact with the eyes and airways, the response to other allergens is not so easily explained, particularly those involved with the gastrointestinal (GI) system.

There is a condition known as “oral-allergy syndrome,” which is thought to occur from swallowing typical aeroallergens. When an allergic person swallows pollen grains (mold spores or animal dander), activation of mast cells is elicited and allergic symptoms occur in the mouth, esophagus, stomach and possibly further down into the intestines. Thus, the IgE that forms against the aeroallergen has a digestive enzyme that activates the Th2/allergy-provoking immunity for parasite protection, with the mast cell reaction occurring in the GI system rather than the airways and/or eyes. Consequently, GI symptoms may begin due to swelling and irritation of the GI tract. And while this consequence could be mistaken for a food allergy, it is difficult to determine which food item initiates the symptoms.

To further complicate identifying the troublemaker, in some instances, different plants may share similar protein components, with some in pollen grains from one plant and some in parts of another plant that may be eaten. Thus, having a birch pollen allergy can result in allergic symptoms when eating apples. Similarly, ragweed allergy can cause a reaction in some people who eat watermelon. And, since hickory and pecan share allergic proteins, an allergy to one item could be manifested as a reaction to a different item that was ingested. In such instances, there would not necessarily appear to be a connection. Allergic symptoms may range from itchy mouth, urticaria (hives) and swelling of the mouth, lips and face; GI distress, nausea, vomiting and loose stools or diarrhea; and systemic symptoms, including anaphylaxis.

Food allergies are more difficult to explain in regard to the immune system responding to parasites. Most food allergens do not have a pattern suggestive of a digestive enzyme, or we may not yet fully understand how to interpret the pattern profiles. While we have been able to identify the specific allergy-sensitizing protein involved with many food allergies, the reason the immune system responds to these as though they were from parasites has been more difficult to ascertain. For a well-studied food allergy such as with the peanut, we know very precisely the protein components associated with provoking allergic disease. We also know the specific proteins causing the allergic reactions to many other common food allergens. What remains unclear is why they are provoking an allergic reaction since they do not have a recognizable digestive enzyme pattern for eliciting an anti-parasitic immune response. At some point, we hope to be able to find a key connection to the immune system misidentifying these items as though they represent parasites.

We will continue with the topic of hypersensitivity and allergic disease in the next issue.

TERRY O. HARVILLE, MD, PhD, is medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences and a consultant for immunodeficiencies, autoimmunities and transplantation.
Coping with Anxiety During the COVID-19 Pandemic

By Erika Lawrence, PhD, LCP

THE CORONAVIRUS (COVID-19) has been labeled a pandemic. Schools, businesses, restaurants and stores are closed. Many of us have been told to “shelter in place.” This may cause many of you to feel anxious, worried or overwhelmed since so much is out of your control and so much is unknown. You may be wondering: Will things get worse? How long will this last? Can I get tested? Will there be a treatment? What happens if I get sick? What if I run out of groceries or personal care items?

Being immunocompromised leaves you vulnerable to serious complications if you contract the COVID-19 virus. Without a doubt, your anxiety is understandable. Yet, while there are a lot of things you cannot control right now, there are some things within your power to manage:

1) Take a break from the news. News or conversations about the virus are everywhere. You can spend hours reading material online about the spread of the virus or joining social media conversations about it. You may think gathering information will help lower your anxiety. But, it actually has the opposite effect. It increases your anxiety. So, turn off the news alerts, stay off social media and take a break from coronavirus information. If some real new information comes out (if there is suddenly a vaccine), trust me, you will hear about it.

2) Talk to someone. We have all been asked to engage in “social distancing,” which means staying at least 6 feet away from others. People who are immunocompromised have likely had to practice social distancing at some point, although it probably didn’t last as long as it looks like it will now. Social distancing can easily lead to social isolation and loneliness, which can increase depression or even worsen physical health. Without a doubt, it is important to stay connected to other people right now. Facetime or Skype with others every day. Set up virtual dinners with friends or family members. Even simply texting can help you stay in touch with others.

Speaking to a professional may also be an important step to consider. Almost all therapists and medical doctors have been given permission to conduct telehealth and teletherapy right now, meaning they can meet with you online. Most insurance companies have also agreed to cover teletherapy. Call your insurance company to see if you can reach out to a therapist or doctor for help.

3) Practice being present in the moment. When people are anxious, it is easy for us to get caught up in our heads rather than being present in the moment. If you catch yourself doing this, try using the “STOPP” technique:

    • Stop: Pause for a moment.
    • Take a breath: Notice your breathing as you breathe in through the nose and out through the mouth.
    • Observe: What thoughts are going through your mind right now? Where is your attention? What are you reacting to? What sensations do you notice in your body?
    • Pull back, and add some perspective: Ask yourself, “What is another way I can look at this situation?” “What advice would I give a friend?” “Is this thought a fact or an opinion?” “What is a more reasonable explanation?”

Practice what works: What can you do right now that would be helpful? What is the most helpful thing for you, others and for the situation? What can you do that fits with your values? Where can you focus your attention right now?

4) Treat yourself and others with compassion, empathy and kindness. This is a very stressful situation. It is much harder to be thoughtful before speaking in times of stress. Take time to unwind. Meditate, color in an adult coloring book, bake, read a book, do yoga.

5) Remind yourself what you can do. It may feel as if your life and health are spiraling out of your control right now. So, remember the important things are still under your control: Washing your hands. Looking out for a fever or a dry cough. Staying at least 6 feet away from others. Not going anywhere with more than 10 people.

And, remember: Everyone reading this magazine right now is connected to you. We are all going through this together.

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

Author’s note: This article was written in March when everything closed down and shelter in place went into effect. It’s likely things may have changed dramatically by the time this article is published.

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CLINICAL BRIEF

Understanding Autoimmune Disorders

By Ronale Tucker Rhodes, MS

TODAY, THE growing number of individuals diagnosed with an autoimmune disorder (AD) is alarming. The National Institutes of Health (NIH) estimates more than 23 million Americans live with an AD, whereas the American Autoimmune Related Diseases Association (AARDA) says that number is 50 million.¹² To put these numbers in perspective, the AD prevalence equals heart disease and cancer combined.¹ What’s more, each year, the incidence rate of ADs is increasing.

What Are ADs?
ADs are conditions in which the immune system attacks its own body tissues. They consist of a host of diseases, many of which are very rare, widely scattered over a number of medical specialties, and usually grouped by body system (digestive, joints, metabolic) rather than as a category with common causal mechanisms (immune system attacks own tissues).⁴

What Causes AD?
While it is not definitively understood what causes AD, researchers suspect environmental factors like infections and exposures to chemicals and solvents, rather than from genetic susceptibilities, are interfering with the immune system’s ability to distinguish self from non-self.¹⁵ In addition, the Western diet is suspected as a trigger because it is high in fat, sugar and processed foods that cause inflammation. Another theory is the hygiene hypothesis, which suggests that because kids aren’t exposed to as many germs today as in years past, their immune system may overreact to harmless substances.³ And, for women specifically, some clinical scientists suspect estrogen may be the culprit, causing a woman’s immune system to produce more antibodies and increasing the likelihood that her body will turn on itself.⁷

Who Is Affected by AD?
ADs affect women at a rate of about two to one compared with men (6.4 percent of women vs. 2.7 percent of men), and the disease often starts during childbearing years (ages 14 through 44). Some ADs are more common in certain ethnic groups. For instance, lupus affects more African-American and Hispanic people than Caucasians. And, certain ADs such as multiple sclerosis and lupus run in families with a susceptibility.⁵

ADs can affect people of any age depending on the disorder/disease.⁹ Indeed, ADs tend to arise early in life. For instance, type 1 diabetes starts in childhood or adolescence, and rheumatoid arthritis and systemic lupus erythematosus (SLE) start during the 20s and 30s. However, these conditions often worsen and lead to complications later in life when they become more noticeable.⁹

Symptoms of AD
Symptoms of AD depend on the disease pathology. There are two types of ADs: systemic and localized. Systemic ADs tend to spread to various organs, from the skin to the kidneys, as well as the heart. Localized ADs have an effect on a particular body organ like the thyroid, liver or adrenal glands. ADs can have an effect on any body part since they can afflict joints, blood vessels, red blood cells, connective tissues, muscles or even endocrine glands (such as the pancreas or thyroid).¹⁰

General symptoms of AD have the maximum probability of emerging in the very beginning, whereas other unique abnormalities might appear later. In adults, each AD has its own peculiar set of symptoms, yet many share similar features such as muscle aches, joint pain, signs of inflammation (redness, heat or pain) and flu-like symptoms. Fatigue is a defining symptom of many ADs.¹¹ In children, the first very common symptoms are dizziness, slight fever, fatigue, dry mouth or eyes, weight loss, diffuse joint pain and skin rashes.¹⁰

Diagnosing AD
Diagnosing an AD is a notoriously exhausting journey, with patients seeing on average five doctors over three and a half years before receiving a diagnosis.⁴ Furthermore, diagnosis can be tricky because symptoms often come and go,
making it difficult to pinpoint the problem unless the physician happens to know the individual has a family history of AD. In many cases, it’s necessary to follow a patient for a while so the disease will manifest itself.13

A rheumatoid factor test is one blood test primarily used to help pinpoint a diagnosis. A positive test result indicates a high level of rheumatoid factor was detected in the blood, which is associated with AD.13 But, it’s important to know that blood tests that look for autoantibodies can yield positive results even when someone doesn’t have an AD.14 For instance, a number of other diseases and conditions can raise rheumatoid factor levels, including cancer, chronic infections, inflammatory lung diseases, mixed connective tissue disease, Sjögren’s syndrome and SLE. In addition, some healthy people (particularly older adults) have positive rheumatoid factor tests, although it’s not understood why. Even some people who have rheumatoid arthritis (one of the most common forms of AD in adults) have low levels of rheumatoid factor.13

Other blood tests used to pinpoint a diagnosis include the anti-nuclear antibody, anti-cyclic citrullinated peptide antibodies, C-reactive protein and erythrocyte sedimentation rate tests.13

Treating AD

Despite the growing number of ADs, scientists are still in the dark regarding interventions that can help cure them. While some may resolve, usually spontaneously and for unknown reasons, most do not. But, ADs can go into remission with therapy. The main goal of therapy is to suppress disease flares and extend periods of remission, if remission is attainable.14 Treatment for ADs include medication, physical therapy, exercise, nutrition and, for some, complementary and alternative medicine (CAM). Medications prescribed depend on the disease, its severity and symptoms. These include nonsteroidal anti-inflammatory drugs such as ibuprofen and aspirin to relieve mild symptoms; prescription drugs to relieve more severe symptoms such as pain, swelling, depression, anxiety, sleep problems, fatigue or rashes; medicines that replace vital substances the body no longer makes such as insulin, hormones and enzymes; corticosteroids to decrease inflammation and reduce the activity of the immune system; biologics (immune-suppressing drugs) to control inflammation and help control disease process and preserve organ function; plasmapheresis to remove antibodies from the bloodstream, thereby preventing them from attacking their targets; and immune globulin (plasma protein replacement therapy).11,13

When an AD affects joints, muscles and bones, physical therapy can help to reinforce muscles and help individuals move body parts more easily. Eating a well-balanced diet and getting regular exercise can also help individuals feel better. In addition, physicians may suggest supplements to replace insulin, hormones or vitamins.10

While it’s hard to know if CAM therapies will work for ADs, some people do opt to try them. Some examples of CAM therapies include herbal products, chiropractic, acupuncture and hypnosis. However, there are limited studies on these therapies, and it’s important for patients to discuss them with physicians since some products can cause health problems or interfere with medicines.16

The Future of AD

The National Institute of Allergy and Infectious Diseases has made it a priority to study ADs because “the chronic and debilitating nature of these diseases, which can lead to high medical costs and reduced quality of life, is a burden on patients and also affects their families and communities.”11 Indeed, based on data from the last decade, estimates of the total AD financial burden are around $100 billion.7 It can only be hoped, then, that continued study will improve outcomes for the millions of people affected by ADs.7

RONALE TUCKER RHODES, MS, is the editor of BioSupply Trends Quarterly.

References

**Medicines**

**Hizentra Granted Orphan Drug Exclusivity as Maintenance Therapy for CIDP**

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

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

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

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**Research**

**Recombinant IVIG Technology Could Reduce Global Shortage**

With a global shortage of immune globulin (IG), GigaGen is developing a recombinant intravenous IG (IVIG) product that could prevent future shortages. IG can be subject to supply shortages because it contains antibodies harvested from plasma provided by thousands of donors to treat patients to help them fight off infections, as well as disorders of the muscles and nervous system. However, a recombinant product could overcome the challenges of plasma-based products.

GigaGen’s recombinant polyclonal IG technology allows the company to capture receptors from the immune system and express them recombinantly. To date, the company has “captured millions-diverse DNA libraries from B cell and plasma cell repertoires from human donors and used these libraries to manufacture recombinant IgG proteins.” David Johnson, GigaGen’s CEO, says the products remain consistent from batch to batch and contain high titers of antibodies, which reduces infusion times for patients. The company’s recombinant IVIG therapy has been validated in mouse models and is being advanced in collaboration with other leaders, including Grifols.

According to Johnson, the development of its recombinant polyclonal IG technologies is “fairly standard.” However, he said the company is still in the learning phase, and he anticipates it will take two more years to complete the process of scaling, at which time he hopes the products will be in clinical trials by 2021 and will then proceed through the regulatory phase.

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The Immune Deficiency Foundation (IDF) has launched a new website to support parents of babies diagnosed with severe combined immunodeficiency (SCID). The site, www.scidcompass.org, offers parents an in-depth explanation of SCID; detailed information on treatment options; advice on how to care for their child after treatment; and access to support systems where they can meet other families.

The website is part of a broader project, the SCID Compass Program, funded by a two-year grant from the U.S. Health Resources and Services Administration, whose goal is to improve outcomes for infants with SCID by:

- Increasing awareness and knowledge about SCID;
- Linking families, especially those living in medically underserved areas, to services; and
- Developing long-term follow-up strategies for infants identified through newborn screening.

To develop and evaluate the website, IDF partnered with parents of children with SCID, grassroots support groups and healthcare professionals with experience working with families affected by SCID. Other partners included the Association of Public Health Laboratories, a professional association responsible for supporting newborn screening programs; the Genetic Alliance, a health advocacy organization; and RTI International, a nonprofit research organization.

“Scidcompass.org is truly a product of teamwork and is presented in an accessible format that we hope parents will find clear, engaging and helpful as they navigate their journey living with SCID,” said Heather Smith, chairperson of the SCID Compass Steering Committee and president of SCID, Angels for Life, a nonprofit support group for parents of children with SCID, which works closely with IDF.

The launch of the website coincides with the one-year anniversary of all 50 states implementing screening for SCID in their newborn screening protocols.

“The website is a natural next step now that newborn screening for SCID is implemented in the U.S. Now, more than ever, babies are being screened, diagnosed and treated for SCID. That means a probable increase in the number of SCID cases and a greater need for information,” said John G. Boyle, president and CEO of IDF.

ADMA Biologics Receives BioNJ 2020 Innovator Award for ASCENIV

ADMA Biologics, a commercial biopharmaceutical company dedicated to manufacturing, marketing and developing specialty plasma-derived biologics for the treatment of immunodeficient patients at risk for infection and for the prevention of certain infectious diseases, has been awarded the BioNJ 2020 Innovator Award in recognition of the development and approval of ASCENIV (immune globulin intravenous [human] slra 10% liquid) by the U.S. Food and Drug Administration in 2019.

“We are honored to be recognized by BioNJ with this prestigious award,” said Adam Grossman, president and chief executive officer of ADMA. “The robust life sciences community in New Jersey provides ADMA with talented and dedicated healthcare professionals who are contributing to the launch and commercialization of ASCENIV, our novel, proprietary immune globulin product, which we believe can help appropriate patients in the U.S.”

“The purpose of the BioNJ Innovator Award is to celebrate the dominant role New Jersey plays in the healthcare landscape and to highlight the vision and innovation contributed by these recipients,” said Debbie Hart, president and chief executive officer of BioNJ. “We are delighted to present ADMA with the 2020 Innovator Award for its success in bringing ASCENIV to market to help immune deficient patients.”
**Important Safety Information**

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

If you are at high risk of blood clots, your doctor will prescribe Hizentra at the minimum dose and infusion rate practicable and will monitor for signs of clotting events and hyperviscosity. Always drink sufficient fluids before infusing Hizentra.

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

Hizentra is a prescription medicine used to treat:
- Primary immune deficiency (PI) in patients 2 years and older
- Chronic inflammatory demyelinating polyneuropathy (CIDP) in adults

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

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

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

Immediately report to your physician any of the following symptoms, which could be signs of serious adverse reactions to Hizentra:
Reduced urination, sudden weight gain, or swelling in your legs (possible signs of a kidney problem).

Pain and/or swelling or discoloration of an arm or leg, unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, or numbness/weakness on one side of the body (possible signs of a blood clot).

Bad headache with nausea; vomiting; stiff neck; fever; and sensitivity to light (possible signs of meningitis).

Brown or red urine; rapid heart rate; yellowing of the skin or eyes; chest pains or breathing trouble; fever over 100°F (possible symptoms of other conditions that require prompt treatment).

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

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

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

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

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

You can also report side effects to CSL Behring’s Pharmacovigilance Department at 1-866-915-6958.
Initial U.S. Approval: 2010

BRIEF SUMMARY OF PRESCRIBING INFORMATION

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

WARNING: THROMBOSIS

See full prescribing information for complete boxed warning.

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

INDICATIONS AND USAGE

HIZENTRA® is indicated for:
* Treatment of primary immunodeficiency (PI) in adults and pediatric patients 2 years and older.
* Maintenance therapy in adults with chronic inflammatory demyelinating polyneuropathy (CIDP) to prevent relapse of neuromuscular disability and impairment.

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

For subcutaneous infusion only.

DOSE FORMS AND STRENGTHS

- 0.2 g per mL (20%) protein solution for subcutaneous injection

CONTRAINdications

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

WARNINGS AND PRECAUTIONS

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

ADVERSE REACTIONS

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

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

DRUG INTERACTIONS

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

Based on March 2018 revision
Research

HyQvia Can Be Tailored to Adults Requiring Alternative Rates, Ramp-Up or Dosing Regimens

A retrospective study has found HyQvia (subcutaneous immune globulin 10% with recombinant human hyaluronidase) can be tailored to adults requiring alternative rates, ramp-up and/or dosing regimens, and it may be especially well-suited to children. The study was designed to evaluate the clinical experience of treating patients with primary immunodeficiency disease (PI) with HyQvia regimens outside of package insert recommendations, as well as in pediatric patients. Data were abstracted from 38 patient records (317 HyQvia infusions), including five patients younger than 16 years old, from seven U.S. immunology clinics.

In the study, 37 patients received HyQvia regimens different from prescribing guidelines. The most notable variations included shorter ramp-up periods, use of two infusion sites rather than one, and slower than maximal infusion rates to mitigate local adverse events. The medical volume infused for single-site doses ranged from 75 mL to 200 mL, and doses split between two sites ranged from 100 mL to 750 mL. The most common type of regimen variation was a condensed ramp-up phase (shorter schedule, higher doses), with 96 percent (24/25) of patients completing ramp-up. The most common ramp-up schedule was three infusions (one at 25 percent to 45 percent, another at 50 percent to 75 percent and the final at 100 percent of target dose) spread over two weeks to four weeks.

The researchers found a shorter ramp-up schedule did not appear to increase the number of adverse events compared to standard ramp-up schedules. According to the researchers, for patients with adverse events, slower infusion rates and the use of two infusion sites may improve medication tolerability. Four of five pediatric patients reported no adverse events, and only one discontinued, stating a fear of needles.


Research

Study Finds Alternative IVIG Dosing Methods Save Millions

A recent study has found intravenous immune globulin (IVIG) dosing optimization using alternative dosing weights can save up to $3.89 million. In the study, the researchers analyzed all IVIG doses administered to adults age 18 years and older from January 2011 to January 2016, which used total body weight (TBW) and height at the age of administration to calculate prescribed dose (g/kg), ideal body weight (IBW) and adjusted body weight (AdjBW). The researchers then analyzed three dosing methods: use of AdjBW if TBW is less than 120 percent of IBW, AdjBW for all doses and IBW for all doses. Outcomes included potential IVIG use averted, direct drug cost savings and reductions in outpatient infusion times for each method.

A total of 9,918 doses were administered to 2,564 patients over five years, representing an average usage of 75,994 grams per year. If dosing methods one, two and three had been used, the annual use of IVIG would have decreased by 21.9 percent (16,658 grams per year), 24.2 percent (18,371 grams per year) and 35.9 percent (27,252 grams per year), respectively, translating into average annual cost differences of $2.37 million, $2.62 million and $3.89 million and average annual outpatient infusion time savings of 841 hours, 920 hours and 1,366 hours, respectively. According to the researchers, IVIG dosing optimization through use of alternative dosing weights represents a significant source of waste reduction and cost reduction.

IN THE NEWS

Research
Higher IgG Trough Levels Are Attained with Weekly SCIG Therapy Than with Monthly IVIG Therapy in PI Patients

In a systematic review and meta-analysis conducted to compare the relationship between IgG dosing and trough IgG levels with overall infection incidence in primary immunodeficiency disease (PI) patients receiving intravenous immune globulin (IVIG) and subcutaneous IG (SCIG) therapy, researchers found SCIG attained a higher trough level, which was associated with lower infection rates, compared to monthly IVIG, which demonstrated no relationship between trough level and infection rates.

The review looked at 24 observational studies published from January 2010 through June 2018 in Medline, EMBASE, Cochrane, Central and Scopus. Eleven of these compared IgG trough levels among SCIG and IVIG patients that showed for every 100 mg/dl increase in the trough, a linear trend of decreased incidence rates of infection was identified in SCIG patients, but no similar trend was identified in trough levels versus infection rates for patients receiving IVIG.


Resource
IDF Publishes Sixth Edition of Patient & Family Handbook for PI Disorders

The Immune Deficiency Foundation (IDF) has released the *IDF Patient & Family Handbook for Primary Immunodeficiency Diseases, Sixth Edition*, which provides comprehensive information about the diagnosis and treatment of primary immunodeficiencies (PI) and life management. The book contains general medical information and is intended to provide individuals and families living with PI tools to enhance the communication process and understand the information they receive from their healthcare team. It can be downloaded for free at primary-immune.org/publication/patients-and-families/idf-patient-family-handbook-primary-immunodeficiency-diseases-6th.

Resource
IgNS Creates COVID-19 Resource Guide and FAQ for Patients and Providers

The Immunoglobulin National Society (IgNS) has created the *COVID-19 Resource Guide and FAQ* to support clinicians and patients during the pandemic. According to IgNS, the guide is intended to be used as a supplement to making clinical decisions. It is not meant to replace clinical judgment and patient-specific decisions, established clinical guidelines or policies created by individual organizations.

Research

AARDA Is Partnering with AutoimmuneMom

The American Autoimmune Related Disease Association (AARDA) is partnering with AutoimmuneMom. For the last seven-plus years, AutoimmuneMom has created a digital community through the hard work and dedication of Katie Cleary. Now, AARDA will host the content that has brought together people with autoimmune disease through stories of how autoimmune disease impacts motherhood and pregnancy, general autoimmune information, personal stories and doctor recommendations. AutoimmuneMom will continue to maintain its social media presence, but all website content will be hosted on AARDA.org.

“I’m honored and grateful that AARDA will now hold all of AutoimmuneMom’s personal autoimmune journey stories and research posts, and will continue providing the research, storytelling and community that are so important to all of us living with autoimmune [diseases]. I can’t think of a better organization to partner with, and I am proud to contribute to AARDA’s vital role in the autoimmune community,” said Cleary.


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Factors That Can Impact Immunity

The body’s response to infection can be improved by incorporating lifestyle changes one step at a time.

By Matthew D. Hansen, DPT, MPT, BSPTS

MANY FACTORS CAN impair immunity, some of which are beyond our control or the full understanding of the scientific world. However, the effectiveness of the body’s defenses against disease is a spectrum that may be impacted by several key factors influenced by healthy living habits. Therefore, those who live with compromised immune systems should not sell themselves short by thinking there isn’t anything they can do to stop infection. There are always things people can do to better help keep germs out and to improve the body’s response to a contagion.

Handwashing and Sanitization

Though not technically a factor that affects immunity directly, proper handwashing, as reported by the Centers for Disease Control and Prevention (CDC), is the most effective action individuals can take to reduce the spread of infectious diseases. While this should be a no-brainer, it’s amazing how many different germs people are exposed to in an average day. It’s also astonishing how many people don’t wash their hands when or how they should.
Many studies have been conducted on handwashing during the last two decades, and sadly, the results are very similar. Anywhere between 10 percent and 35 percent of subjects surveyed or directly observed didn’t wash their hands at all after using the bathroom!

One study conducted at Michigan State University observed more than 3,700 people after using public restrooms. Of all studies conducted on handwashing, the Michigan State findings demonstrated among the highest compliance rates for subjects at least making a token washing of their hands. That’s what one might expect of a higher-educated population, right? The study found “only” 7 percent of women and 15 percent of men did not wash their hands after using the bathroom. However, a closer look at the results reveals just 50 percent of men and 78 percent of women used soap. The average handwashing time: 6 seconds! Only 5 percent of people washed their hands long enough to kill infectious germs. Surveys have demonstrated that even fewer people self-report washing their hands before preparing and eating food, and after handling money and performing other activities that involve high exposure to microbes than they do after using the bathroom.

My brother and I used to think my Aunt Diane was paranoid for always opening public doors with a clean tissue or with her sleeve. I chuckled at the comedian Howey Mandel for refusing to shake someone’s hand and fist bumping instead, thinking maybe it was part of his bit. But, my wife and I began modeling both Diane’s and Howey’s behavior a long time ago, and we insist our kids do the same. I’ll still shake hands or give a high five, but as soon as I can discreetly sanitize my hands afterward, out comes the bottle, and I’m very careful not to touch my eyes, nose or mouth — or anything that could come in contact with them — in between.

CDC indicates it takes 15 seconds to 20 seconds of vigorous handwashing to effectively kill germs. The proper way to wash is to wet the hands with clean running water and apply soap. The hands should be rubbed together to make a lather and scrubbed well, including between the fingers, under fingernails and the back of hands. You may have heard the suggestion to sing the “Happy Birthday” song twice (with a regular rhythm) to assure sufficient time. After you’re done washing, rinse hands under running water and dry with a clean towel or air dryer (damp hands are much more likely to spread bacteria). I usually use my knuckles to turn the paper towel dispenser if it isn’t automatic. I do the same, or use my elbow, to press public elevator buttons.

Other universal precautions that should be considered in addition to handwashing, particularly if someone with a compromised immune system is around others who they suspect to be infectious or if they are infectious, include wearing medical gloves and/or a face mask.

**Stress**

If reducing stress were only as simple as handwashing, would we do it? According to the American Psychological Association, 75 percent of Americans reported experiencing at least one symptom of stress in the past month. When stressed, individuals’ adrenal glands produce hormones called adrenaline and cortisol. These hormones have many effects on the body to help it prepare for a fight-or-flight response. The short-term response on the immune system is actually a boost for three to five days. However, if people persist in worrying about the stressor, even after the immediate concern may have resolved, the cortisol begins to interfere with the number of T cells and white blood cells produced by the body, which then affects the efficiency of the body’s immunological response.

Studies have demonstrated that subjects exposed to stress are more susceptible to colds and other infections. Stress itself does not actually make people sick; however, when it compromises the immune system, that person is likely to get sick more often and take longer to recover. In addition to this direct impact on the body’s immune system, stress can influence other factors (e.g., sleep patterns, nutrition and activity levels) that, in turn, can distress the immune system as well.

Individuals should find activities that help to reduce stress, and learn how and where to apply them as needed. Activities can include exercising, walking/hiking, meditating or performing...
yoga, praying, listening to music, writing in a journal or art — anything that works in a pinch that they can look forward to in order to decompress.

**Sleep**

Experts generally agree that the optimal amount of sleep for adults is seven hours to nine hours per night. However, according to a widely referenced 2013 Gallup Poll, 40 percent of respondents received less than the minimally recommended seven hours. CDC published similar findings in 2016 from a review of collected data that indicated “a third of American adults are not getting enough sleep on a regular basis.”

Sleep experts generally agree that the optimal amount of sleep for adults is seven hours to nine hours per night. However, according to a widely referenced 2013 Gallup Poll, 40 percent of respondents received less than the minimally recommended seven hours. According to Eric J. Olson, MD, of the Mayo Clinic, studies show people who don’t get quality sleep or enough sleep are more likely to get sick after being exposed to a virus such as a common cold virus. Lack of sleep can also affect how fast you recover if you do get sick. These studies have shown sleep deprivation may decrease the production and release of small protective proteins called cytokines that are needed to combat infection and inflammation. Additionally, infection-fighting T cells are reduced, as is their ability to attach to infected cells during periods when people don’t get enough sleep.

As with stress, there are a number of motivators for someone experiencing poor sleep; however, given the fact that the average amount of sleeping has dropped by one-and-a-half hours to two hours over the past century, it would suggest many of the problems are self-induced. Video games and binge-watching shows were not pastimes 100 years ago. In fact, before the electric age, people tended to retire for the night and rise in the morning with the sun, or soon after. Trying to go to bed and wake up at the same time every day is still a good practice.

Getting rid of distractions and practices that could interrupt sleep are also important. For instance, the bedroom should be comfortable, dark and as quiet as possible. That might mean having to address concerns with a bed partner who snores or grinds his or her teeth. It almost definitely means there shouldn’t be a TV or computer on in the room, even if the volume is muted. Some people can’t take caffeine any time after noon, or they find it difficult to sleep at night at a reasonable time if they take a nap after a certain hour.

If people still can’t sleep due to insomnia or other health complications, they should meet with their physician or other healthcare specialist to consider their options such as prescribed or over-the-counter medications and relaxation or cognitive behavioral therapy techniques.

**Environmental Factors**

A plethora of suspected environmental factors affect people’s immune responses, and an army of research is seeking to confirm or refute those factors’ role. What can definitively be said is the prevalence of many medical diagnoses associated with the immune system is increasing.

Although we know genes play a role in many of these diseases, not all have been proven to have a genetic link. What’s more, it appears the disease process in some genetically susceptible people may still be triggered — or at least exacerbated — by environmental factors. In this context, environment includes the air people breathe, what people take into their bodies, the industrial chemicals that surround people and other potential hazards that people are exposed to as part of daily life in modern society.

There are many hypotheses regarding the consequences of air pollution, processed foods, plastics, contaminated water and other possible environmental offenders, but the purpose here is not to elaborate on theories, as convincing as some of them may be. Instead, individuals should conduct their own research, paying close attention to the sources of information. Oftentimes, many of the articles available on the Internet are sponsored, if not commissioned, by legal groups heading class action lawsuits, or other factions that may possess a bias.

Though the exact consequences of many environmental risk factors may not be well-established, there are some that are. The hazards of smoking and, to a lesser degree, second-hand smoke are well-known. The government website www.smokefree.gov states that the high levels of tar and other chemicals in cigarettes can make the immune system less effective at fighting infections and leave it more susceptible to acquiring autoimmune diseases.
Nutrition and Supplements

Many people are under the impression that at the first inkling of feeling sick, they can pop a vitamin C with 1,000 percent of the recommended dietary allowance (RDA) or some other special supplement to nip a cold in the bud. Though the placebo effect can be a factor in feeling better, the Cleveland Clinic maintains, “A truly healthy immune system depends on a balanced mix of vitamins and minerals over time. With some exceptions, it’s best to get your vitamins and minerals from your food rather than in pill form.” Vitamins and other natural elements important to preventing and fighting infection include vitamins C, E, B6, A and D, folate, iron, selenium, zinc and protein.

Multivitamins and other vitamins certainly have a place, but guess where most of the 900 percent excess in RDA often goes if the body is already at capacity? I’m weary of high-priced supplements that claim to be superior because they have a proprietary formula. I also warn people they should become familiar with the origination and quality of their supplements because some have been demonstrated to not include what they say they contain, or they are contaminated with other ingredients.

If there’s one common ingredient individuals should try to ingest less of on behalf of their immune systems and other bodily functions, it’s refined sugar. Sugar provides fuel for bacteria, fungi and yeast. When white blood cells are exposed to sugar in high levels, their ability to fight bacteria decreases. Sugar also depletes vitamin C and B vitamins in the body, triggers inflammation, raises cortisol (see discussion on stress), and can lead to insulin resistance and type 2 diabetes, impairing the immune system further.

Unfortunately, many foods high in refined sugar are also low in nutrients needed for the immune system. People shouldn’t be fooled by marketing ploys that try to make sugar sound healthy (e.g., organic cane juice, agave nectar, palm sugar). These sugars are broken down in the body essentially the same as table sugar. Agave, often pitched as a healthier alternative to sugar, is up to 90 percent fructose, which can be metabolized only in the liver. High fructose intake can lead to diabetes and other complications.

Exercise

If you’ve read any of my other articles, you know that exercise is one of my favorite topics. According to the government website www.medlineplus.gov, we do not know exactly if or how exercise increases your immunity to certain illnesses, because none of the theories have yet been proven. We do, however, understand that exercise helps to decrease stress hormones (e.g., cortisol), improve sleep, increase general fitness and generally help participants feel better about themselves and life.

One Step at a Time

In fact, that’s the best part about all of this information. Science is finding that if something is good for the immune system, it usually has far-reaching implications for well-being too. So, people shouldn’t give up on the influence they can have on their health. Sometimes it doesn’t start with healthy, but healthier. Healthy living takes time, but it’s something everyone can start or continue to do one step at a time.

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

References
Preparing for what may come after being diagnosed with a chronic illness can help individuals adjust to their new lives.

By Brenda Kimble

**SHOCK. DESPAIR.** Worry. That’s what people often feel after being diagnosed with a chronic illness. But there’s hope, too, and relief. Why? Because now you know what is wrong. And, even though it might take time to realize it, there’s power in knowing what it is that’s been plaguing your body for such a long time. All those symptoms? All those doctor visits? All those blood tests? Now, they amount to something: a diagnosis.

Living with a chronic illness is hard, and sometimes it can feel downright impossible. But in this fight, you’re certainly not alone. By 2020, an estimated 157 million people in the United States will be living with a chronic illness. The key word, of course, is “living.” Making it through days, weeks, months and — yes! — years is more difficult when you’re doing it while sick. But whether your illness is visible or invisible, whether it affects you daily or flares up every so often, whether you’re in remission or fighting for it, living while chronically ill is something you can do. You just need to make a few adjustments. Following are three things to do now that you’ve been diagnosed.
**Build a Support System**

Even the most independent people can’t go it alone all the time. You know the phrase “it takes a village”? Well, that doesn’t only apply to children. It makes perfect sense for you, too, and every other adult out there. We all need help from time to time, especially when we’re ill.

With a chronic illness, there are times of sickness ahead. If your symptoms are well-managed now, it can be tempting to push aside this necessary step to living with a chronic illness. I urge you not to wait. Get a support system in place now, whether you’re feeling healthy or not.

Who should be members of your support system?
- Doctors, including specialists and a general practitioner
- A mental health counselor, since the incidence of depression and anxiety increases in those with chronic illnesses
- Friends and family whom you feel comfortable asking for help such as cooking meals, running errands, cleaning the house and providing moral support
- You might also want to enlist the help of a physical therapist, a nutritionist, a financial planner, a patient advocate or a social worker.

**Manage Stress**

The impact of stress on the immune system is well-documented. Numerous studies show stress puts strain on organs and body systems, increases inflammation in the body, triggers symptomatic flares and worsens mental health.

From impacting digestion to raising blood pressure, there are a variety of ways stress damages the body. Each of these effects prevents the immune system from defending the body against foreign invaders and healing the damage caused by normal wear and tear. Minimizing stress is essential to maintaining health.

But how do you minimize stress when you’ve just received your diagnosis? How do you do it up to you. There are lots of techniques to help you manage your stress level.

A bit of good news: Research is beginning to show these destressing strategies also improve other areas of mental and physical health.

You can try a variety of different things such as breathing exercises, yoga, an exercise regimen, meditation, mindfulness practices and therapy. Spending time outside and participating in physical activity and creative outlets have all been shown to reduce stress levels in both the general population and in the chronically ill.

To reap the benefits, begin exploring your options now. Put a routine in place, and make note of it. Share it with others so that if you forget about it, they can gently remind you it’s time to take it up. That way, when you’re beginning to feel stressed or depressed, you can turn to your chosen practice and begin to breathe a little easier once again.

**Plan for the Future**

There are hundreds of different chronic illnesses and many subtypes with different projected outcomes, so what your future looks like will be different from what another’s future will look like.

To figure out what your future could hold, first sit down with your medical team or the specialist you work with to manage and treat your chronic illness. It might be wise to have your significant other at this meeting so you can discuss the possibilities together.

Then, sit down with the decisionmakers in your family. That could be a spouse, a child, a parent or even a close friend. You need to have an honest conversation — or a series of conversations — about your wishes for the future, along with contingency plans in case your health deteriorates.

Here’s a short list of topics you should consider now before they become an issue down the road:
- Fertility and reproduction
- Childcare
- A living will
- Long-term care and caregiving responsibilities
- Power of attorney
- Property and estate management
- Retirement savings and financial investments
- Senior care plans for your parents and your significant other’s parents

**A Next Step**

Adjusting to life with a chronic illness is all about ensuring that if you get sick — or sicker than you are right now — you always have a next step in place. That way, once life happens, you can move forward without worrying about what you need to take care of first. Tackle it bit by bit, and you’ll find yourself ready for whatever lies ahead.

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Making Medications Work Best For Patients

Understanding the various factors that affect medications can help patients ensure their prescriptions are providing the intended therapeutic effects.

By Surayyah Morris

MEDICINE WAS CREATED to diagnose, treat, cure and prevent disease. The ultimate goal is to achieve a desired effect with as few side effects as possible. And, while many factors can affect this process, there are many ways to overcome the challenges associated with taking medications.

Patients who take medications such as intravenous immune globulin know that the side effects can be just as bad as the condition the medication is treating. It can seem as if the medication that helps one problem causes five more. Indeed, it can often be difficult to balance effective treatment with intolerable side effects. To help with this balancing act, following are some tips to assist with becoming a master of risk-benefit decision-making that should be applied accordingly, always ensuring doctors are informed of any changes. But, first, let’s begin with a crash course on how the body affects medicine and vice versa.

How the Body Affects Medicine and Vice Versa

Pharmacokinetics, the process that occurs from the time the drug enters the body until the time it leaves the body, is known as ADME (absorption, distribution, metabolism, elimination/excretion):

- Absorption is the movement of a drug from its site of administration into the blood.
- Distribution is the movement of a drug from the blood into the tissues and into cells.
- Metabolism, also known as biotransformation, is the enzyme-mediated change in the structure of a drug that occurs when the drug enters the tissues and cells.
- Elimination/excretion is the movement of the drug as its metabolites out of the body.

Pharmacodynamics is the term used to describe what a drug does to the body. Drugs act by mimicking or blocking the body’s own regulatory processes. Therefore, they can only alter the rate of those processes; they cannot provide new functions. Because drug responses are not entirely predictable, it is important for prescribers to know the patient’s body to determine the intensity of the drug’s effects, which involves knowing the pharmacokinetics. Pharmacodynamics, however, does not always end when the drug physically leaves the body. Drugs can have an effect even after being eliminated/excreted.

Mastering the Risk/Benefits of Drugs

How can this information be used to patients’ advantage? Following are a few ways to help patients understand and adjust some common life practices to make medications work for them and not against them.
Other medications. Other medications are one of the most common complications that can alter the effectiveness of a drug. Drug-drug interactions occur when one medication has an effect (either negative or positive) on another. For example, if drug A is taken at the same time as drug B, drug A may inhibit the metabolism of drug B, causing too much of drug B to remain in the body, which potentially increases its action and side effects. Contrarily, if drug A induces the metabolism of drug B, too little of drug B will remain in the body, which will inhibit its optimal therapeutic effect.

Combining medications can produce both a desired or undesired effect. For example, a desired effect results when two drugs work together to lower blood pressure if one medication is not working well enough. An undesired effect may occur when taking two medications that cause drowsiness or sleepiness, which can increase risk of falls, inattentiveness or make a person too tired to function.

An important part of the pharmacist’s job is to understand how medications react when taken together. And, while there are minor interactions that are not significant for most people, they may be significant for some.

Food and beverages. Meals and beverages affect some medications, which means medication may be better absorbed on an empty stomach versus a full stomach based on how the medication is affected by gastric contents or where in the body the drug is absorbed to be optimally effective. Medications best taken on an empty stomach are usually better to take in the morning before a meal with a glass of water. Medications best taken with a meal should be taken during or immediately after the meal.

The prescription label should have in bold print whether the medication should be taken with or without food. It’s important for patients to read the entire label to understand the important information they need to know.

Grapefruit juice can inhibit how some drugs are metabolized by increasing their therapeutic levels. An increase in grapefruit juice concentration can cause more of the prescription medication to be available for the body to absorb and use. Consequently, individuals may experience worse side effects and a more intense therapeutic effect than normal. There is no desirable amount of grapefruit juice to consume to delay or avoid this reaction; it should be avoided entirely when taking a medication that interacts with it.

The prescription label will indicate if the medication should not be consumed with grapefruit juice.

Time of the day. Some medications may be better taken at a certain time of day (morning/midday/evening) based on how certain body processes work or how long it takes the medication to metabolize. For example, the human body synthesizes cholesterol at night, rendering cholesterol medications such as statins taken at night more effective. As another example, diuretics prescribed for blood pressure, which cause more frequent urination that can be better managed by simply changing the time of day they are taken. Taking diuretics in the morning will prevent individuals from running to the bathroom all night long if they are taken too late in the day.

If a prescription causes more frequent bathroom use than usual, ask the pharmacist or doctor if it is OK to take the medicine earlier in the day.

Temperature and storage. Medications are greatly affected by temperature. If any medication is subjected to temperatures outside of its recommended storage temperature, it will likely begin to degrade or become so altered that it will be ineffective. To prevent this, prescriptions should be kept at the recommended temperature according to its package insert or medication guide:

- Room temperature: 68°F to 77°F (20°C to 25°C)
- Refrigerator: 35°F to 46°F (2°C to 8°C)
- Freezer: -58°F to +5°F (-50°C to -15°C)

If prescriptions are sent by mail, they should be removed from the mailbox as soon as possible. If prescriptions are required to be stored at less than room temperature, they should be packaged with an ice pack/cold pack. If they are not packed with an ice pack/cold pack, the pharmacy should be notified of this error so it can send medicine to replace what has been possibly compromised.

Contrary to popular belief, the bathroom cabinet is not the best place to store medications because bathroom temperature and humidity levels vary, which is the quickest way to decrease medication effectiveness. Instead, they...
should be stored in a cool, dry area away from heat sources or in a safe place in the bedroom.

**Formulations and pill burden.** If prescriptions are to be taken by mouth in pill form (such as a tablet or capsule), a request can be made of the physician to prescribe a different formulation for those who have difficulty swallowing. A different formulation could include a liquid, a sublingual tablet (melts under the tongue), an orally disintegrating tablet (melts on top of the tongue) or even a patch (sticks to skin so medicine can be absorbed). It should be noted that not all medications come in different formulations, and there are also other less common formulations available.

Aside from requesting an easier formulation, the pill burden can be decreased by asking the pharmacist or doctor if there is a combination medication available for two or more medications. A combination medication includes two to three separate medications that are safely combined into one pill. Combined medications are not common, but it is still worthwhile to see if one is available.

**Lifestyle.** This may take some extra thought and preparation, but it is still very much worth the effort. Calculate the time it takes to do an infusion — from preparing the medication to removing the needles and placing Band-Aids on the injection site. Let’s assume it takes three hours. If patients know they typically experience post-infusion blues (fatigue, headache, nausea, etc.), they can use this to strategize when deciding which day to infuse. If the infusion is usually performed on a workday or school day, a late night can be sacrificed by starting the infusion around bedtime, so once it is finished, sleep can follow immediately after to, hopefully, eliminate any undesired blues. If there aren’t any post-infusion issues, the infusion could be performed early in the morning.

The best advice for infusions when traveling is to do them before leaving home. If patients must infuse while traveling, medication should be prepared in advance. However, the pharmacist or doctor should be consulted.

Medications/supplies should always be kept on the person, not in checked baggage. A medication bag is a free carry-on when flying.

**Social activities.** Certain lifestyle habits can affect how medications work. These include:

- **Smoking:** Cigarettes cause many health problems, but they are also an inducer when taken with certain medications. This means the medication will clear the body faster and ultimately be less effective than intended.

- **Alcohol:** Although alcohol can safely be consumed with some medications, it’s not safe for all. When medication interacts with alcohol, there is no easy way to emphasize how awful the outcome will be. It’s best to consult with the pharmacist about any interactions before drinking alcohol and taking any prescription or over-the-counter medications to ensure unwanted reactions are prevented.

> **tip** When in doubt, don’t do it! It is always better to be safe than sorry.

**Adhere to the Package Insert**

Every prescription drug has a package insert that contains all of the important information patients need to know about the drug and its use. The package insert is a thin folded paper provided with the medication. The information on the package insert is also online and can be found by visiting the website for that particular drug, or by searching the medication on the U.S. Food and Drug Administration website.

Patients’ understanding of the interaction between their medication and their bodies can go a long way to maximize the outcome of their treatment with the fewest unanticipated reactions.

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Understanding Combined Immunodeficiencies

While knowledge about this complicated disease has advanced over time, it is still often misdiagnosed and treatment must be individualized for each patient.

By Terry O. Harville, MD, PhD, D(ABMLI), D(ABHI)

AS THE NAME implies, combined immunodeficiencies (CIDs) encompass a category of immunodeficiencies in which T and B lymphocytes fail to function as normally expected. What is not clarified in the name is whether B lymphocytes fail to function due to lack of T lymphocyte assistance or whether the underlying gene mutations affect the functions and numbers of both T and B lymphocytes. Additionally, it is unclear at what point CID is severe enough to become classified as severe combined immunodeficiency (SCID); what the varieties of CID are; whether some have more T lymphocyte dysfunction and less B lymphocyte dysfunction and vice versa; and how CID is diagnosed and treated.

Historical Perspective of CID

In the past, most immunologists understood what the term CID meant. For instance, many years ago (and still today), it was believed SCID was diagnosed in infancy and required bone marrow transplantation (BMT) or hematopoietic stem cell transplantation (HSCT) for infant survival. Furthermore, it was believed CID was diagnosed later in children with less-severe disease that did not require BMT/HSCT. Indeed, lymphocyte deficiencies at that time were basically divided into five categories: 1) pure T lymphocyte deficiency (e.g., DiGeorge syndrome), 2) pure B lymphocyte deficiency (e.g., X-linked agammaglobulinemia [XLA]), 3) SCID, 4) common variable immunodeficiency (CVID), defined as primarily B lymphocyte deficiency with possibly some T lymphocyte deficiency and 5) CID, defined as primarily T lymphocyte deficiency with corresponding B lymphocyte deficiency, but not as severe as SCID (e.g., Wiskott-Aldrich syndrome [WAS]). The latter is now what we refer to as classical CID.

Rebecca Buckley, MD, a pioneer in immunodeficiencies, used the term SCID to describe those with such severe T and B lymphocyte immunodeficiency that they would not survive without BMT/HSCT. Further, infants with SCID could undergo successful BMT/HSCT without the need for chemotherapy. This distinguished them from patients with CID who require chemotherapy to suppress the meager immunity present, enough to prevent engraftment of donor cells if BMT/HSCT were performed. Indeed, this description remains a good differentiator between SCID and CID. Unexpectedly, the category of CID expanded over the years, which complicated its definition.
In Between Past and Present Considerations of CID

Between the definition of CID described above, and the latest definition to be discussed below, CID had become an all-encompassing disease process (Table 1). CID was divided, essentially, based on the severity of T lymphocyte dysfunction into 1) SCID (the most severe), 2) classical CID (less severe than SCID) and 3) CVID (less severe than classical CID with T lymphocyte function ranging from fundamentally normal to some dysfunction).

However, the medical profession began recognizing that some conditions were not as pure as previously thought. For example, there were infants who initially appeared to have SCID, but the evaluation demonstrated their bodies were capable of making some T lymphocytes. Therefore, their disease was worse than classical CID but not as severe as classical SCID. These cases became known as leaky SCID to distinguish them from classical SCID. Leaky SCID could be considered a CID since these patients require BMT/HSCT for treatment and typically require chemotherapy for conditioning for the transplant. Further, in some infants, T lymphocytes were present and functioning to some extent (as expected in CID), but the disease process still required BMT/HSCT for the infant to survive (e.g., Omenn syndrome). Thus, according to Dr. Buckley’s definition, patients with some level of T lymphocyte numbers and function, and who require chemotherapy conditioning, should be diagnosed with CID rather than SCID.

Molecular DNA techniques also led to further clarifications of the definition of CID. As these techniques identified which mutations were responsible for different immunodeficiencies, classifications based on which DNA mutations were present could be performed.

The Recent Past of CID

In 2016, the World Health Organization (WHO) released a reorganization of the International Classification of Diseases (ICD-10) (Table 2) that includes a specific category of CID (D81). Yet, the conditions listed were previously considered mostly in the SCID category, requiring BMT/HSCT for treatment, rather than in classical CID. Interestingly, conditions with milder than SCID T lymphocyte dysfunction, which were included previously as classical CID diagnoses, are not listed under the CID category. These are now primarily listed in separate categories based on additional features associated with the disease process. For example, WAS is listed as “D82.0, immunodeficiency with thrombocytopenia and eczema” rather than as a CID.

The use of ICD-10 is helpful from the standpoint of hospital billing and reimbursement, but it does little to promote understanding the immunologic relationships of the disease processes.

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**Table 1. Former Categorical Hierarchy of Combined Immunodeficiencies (CID)**

<table>
<thead>
<tr>
<th>CID</th>
<th>SCID</th>
<th>Classical CID</th>
<th>CVID</th>
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**Table 2. 2016 Reorganized ICD-10 Classifications of Combined Immunodeficiencies**

- D81 Combined immunodeficiencies
  - Excluding autosomal recessive agammaglobulinaemia (Swiss type) (D80.0)
  - D81.0 Severe combined immunodeficiency (SCID) with reticular dysgenesis
  - D81.1 Severe combined immunodeficiency (SCID) with low T- and B-cell numbers
  - D81.2 Severe combined immunodeficiency (SCID) with low or normal B-cell numbers
  - D81.3 Adenosine deaminase (ADA) deficiency
  - D81.4 Nezelof syndrome
  - D81.5 Purine nucleoside phosphorylase (PNP) deficiency
  - D81.6 Major histocompatibility complex class I deficiency Bare lymphocyte syndrome
  - D81.7 Major histocompatibility complex class II deficiency
  - D81.8 Other combined immunodeficiencies Biotin-dependent carboxylase deficiency
  - D81.9 Combined immunodeficiency, unspecified Severe combined immunodeficiency disorder (SCID) not otherwise specified

The World Health Organization ICD-10 classification of combined immunodeficiencies primarily includes conditions that were mostly previously considered SCID due to the need for BMT/HSCT. It does not include classic CID considerations such as DiGeorge syndrome and Wiskott-Aldrich syndrome, which fall into specific categories based on additional features present. Further, CVID is placed into its own separate category.

Source: icd.who.int/browse10/2016/en#/D81
Further Considerations for the Recent Past of CID

It can now be recognized that there is not a single form of CID, but instead a complex category of immunodeficiencies, with the name acting as a catch-all for a myriad of conditions that share both T and B lymphocyte dysfunctions. Thus, the International Union of Immunological Societies Expert Committee for Primary Immunodeficiency published a more comprehensive classification for CID.1 This classification divided CID into 77 types and subtypes. These include the various SCID types due to different mutations, as well as the classical CID diagnoses such as DiGeorge syndrome and WAS, all of which are placed into specific classifications based on clinical, laboratory and genetic features, with many of the types classified based on specific known mutations of genes associated with immunodeficiencies. CVID is no longer in the main CID category, but is placed into a category defined by “primary antibody dysfunction.”

Ultimately, the goal of classification systems of immunodeficiencies is to have all the types and subtypes categorized based on the specific clinical features, expected laboratory test results and underlying DNA gene mutations. UpToDate.com published a review of CID last updated in 2019, in which Luigi D. Notarangelo, MD, states: “Combined immunodeficiency syndromes are somewhat arbitrarily distinguished from severe combined immunodeficiency (SCID) in that they do not characteristically lead to death from overwhelming infection in the first year of life. In addition, combined immunodeficiency syndromes frequently have associated clinical features.” This definition parallels Dr. Buckley’s.

What Is CID?

CID is a broad categorical name for immunodeficiencies with T lymphocyte and B lymphocyte dysfunction that have at least 77 types and subtypes. In most cases, the T and B lymphocyte counts are low, along with abnormal function. For most, mutations are present affecting both T and B lymphocytes, although there may be some in whom the mutation primarily affects T lymphocyte function, which causes a reduction in B lymphocyte function due to the lack of T lymphocyte help.

Patients with CID whose primary dysfunction occurs in the B lymphocytes with less effect on the T lymphocytes may now be in the CVID category. Patients with CID who have the most severe T lymphocyte dysfunction are in the SCID category. Patients with SCID are typically infants who require BMT/HSCT for treatment to survive and usually do not require chemotherapy conditioning for successful transplantation.

Some forms of CID are not recognized until the person is older. In some of these cases, BMT/HSCT may still be helpful, but the risks of transplantation have to be weighed against the risks of not transplanting due to the potential morbidity and mortality associated with the chemotherapeutic conditioning regimen versus those caused by the disease itself.

Some cases of CID are not diagnosed until adulthood. Some of these patients were initially diagnosed with CVID and were subsequently changed to a diagnosis of CID after DNA testing revealed mutations more associated with CID (e.g., milder forms of adenosine deaminase deficiency).

Thus, CID represents a collection of diseases with significant T and B lymphocyte dysfunction, where the T lymphocyte dysfunction is the primary root of the problem. This results in susceptibility to viral, bacterial, fungal and opportunistic infections (infections not expected to occur in persons with normal functioning immunity), in addition to susceptibility to autoimmune disorders.

How Is CID Diagnosed?

Diagnosing CID can be difficult and involved. Those who have symptoms similar to SCID will likely be evaluated in infancy for recurrent, severe and/or opportunistic infections. The lymphocyte count of the white blood count is expected to be low, and assessment of the specific numbers of T and B lymphocytes is expected to be low. Specific markers of T lymphocytes, for example CD45RO expressed on the cell surfaces of CD4 T lymphocytes, may be disproportionally elevated (high percentage, when a lower percentage is expected for age). Additionally, T lymphocyte function is expected to be subnormal, but not to the relative absence observed in SCID. Quantitative antibody levels may be low, and if...
assessed, the ability to make antibodies to specific antigen-
vaccine challenge is expected to be impaired. Thus, for
infants, the difficulty may be determining whether the
diagnosis should be SCID or CID. Currently, DNA analysis for
specific mutations is also helpful for determining the diagnosis.

For older infants, children, teenagers and adults, diagnosis can
be delayed. For younger children and some school-aged children,
their illnesses may be perceived as bad luck and due to viruses
acquired in a daycare or school setting. In this scenario, since
there appears to be a reason for having recurrent illnesses,
there is a delay in laboratory evaluation and a potential
immunodeficiency is not considered. However, when an
evaluation is initiated, it would likely look for an antibody
deficiency due to its relative commonality versus CID.

A less-than-expected improvement with immune globulin
(IG) replacement therapy for a putative antibody deficiency
could prompt a further evaluation for CID. This is because
even when treated with IG, patients with CID may continue
to become ill due to the presence of more pronounced T
lymphocyte deficiency. Unfortunately, more bad luck rather
than treatment failure is often considered, resulting in further
delaying a CID diagnosis. Once again, DNA analysis for
specific gene mutations that cause immunodeficiencies can
help to better delineate the actual condition present.

How Is CID Treated?
There is no one best treatment option for CID. Treatment
should be individualized and optimized for each patient.
Those with more severe disease (more infections) or other
features such as autoimmunity should be considered as candid-
dates as soon as possible for BMT/HSCT, with all the risks
and benefits considered. Earlier transplantation may help prevent
the accumulation of infectious organisms that can make later
transplantation more difficult since these organisms may result
in severe infections after chemotherapy has further compromised
the immune system in preparation for BMT/HSCT.

An example of complications due to accumulation of
infectious organisms occurs in WAS. Even though early-in-life
symptoms could be relatively mild and manageable, commonly
at about 5 years of age, Epstein Barr virus (EBV) infection may
occur. Once infected, EBV remains in the B lymphocytes for
the rest of the life. Patients with WAS are at significant risk
for EBV to cause B lymphocytes to grow abnormally due to
their underlying immunodeficiency. Initially, there may be
lymphoproliferative disease (abnormal growth and expansion
of B lymphocytes), but it can evolve into B lymphocyte
leukemia or lymphoma. If this happens, more drastic
therapy may be required, including BMT/HSCT.
Therefore, early BMT/HSCT should be an important
consideration in CID.

As noted, some patients have an illness more akin to
CVID that causes milder T lymphocyte dysfunction than the more severe cases of CID. These individuals may respond
reasonably well to IG replacement therapy. Daily antibiotics
may be required by some, whereas others may need only
courses of antibiotics, particularly during the winter months.
Ongoing evaluations are required to denote if there is
progression in T lymphocyte dysfunction, especially onset
or worsening of autoimmune disease features. Again, DNA
analysis for specific gene mutations can help toward a decision
of undergoing earlier BMT/HSCT for definitive treatment.

It is recognized that autoimmune processes (which can
occur in patients with CVID) are potentially the most signif-
icant issue in some patients with CID. Their infections may
be controlled with IG replacement and antibiotic therapies,
but patients may remain quite ill from autoimmune disease.
Determining which genes are responsible can be helpful. For
example, if the PI3Kδ protein has a mutation causing
increased activity, immunodeficiency and autoimmune
may be present.3 In these persons, the use of rapamycin, a
so-called mTOR inhibitor, can result in dramatic improvement
in symptoms. This form of therapy may help re-establish a bal-
ance in immune system function, resulting in good outcomes.

While patients may be treated for a long time with these
treatments, it remains unknown whether these patients should proceed to BMT/HSCT for a more definitive treatment of the underlying disease. The mTOR inhibitor can only establish a certain level of homeostasis, and disease breakthroughs may occur. By determining the underlying genetic process resulting in CID, it may be possible to individualize treatment with specific medications as described above. Overall, though, BMT/HSCT should be considered as early as possible as a potential cure for the underlying disease.

A Complicated Immunodeficiency

CID is complicated. There are multiple individually named disorders, in addition to unnamed ones, which have both T and B lymphocyte dysfunction and are thus CIDs. In patients with CID, T lymphocyte dysfunction can range from being less severe like CVID to extremely severe such as with SCID. Some patients may respond to treatment with IG replacement and antibiotics. But, those with additional conditions such as autoimmunity will likely require further individualized therapies and therapeutic options. Some of these may actually work well, but it is unknown if they can provide the needed long-term benefit. DNA studies for mutations responsible for immunodeficiencies can help with diagnosis and, potentially, with individualized treatment options. BMT/HSCT should be considered as early as possible for definitive treatment in most patients with CID.

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.

Bibliography

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Lifestyle changes that include proper food choices, exercise and other healthy habits can help individuals gain or lose weight healthfully as needed.

By Emily Cooper, RDN

WEIGHT CAN OFTEN be a challenge for those with an immune disorder. For those carrying around excess weight, losing extra pounds can help improve health outcomes, lower the risk for certain diseases and increase energy levels. And, while getting rid of the stubborn weight can be more of a difficult task for those with immune and/or autoimmune diseases than the average dieter, it can ease symptoms and reduce inflammation in the body. “Losing just 5 percent to 7 percent of weight may reduce joint pain in patients with rheumatoid arthritis by reducing overall inflammation,” says Lisa Andrews, MEd, RD, LD, owner of Sound Bites Nutrition, LLC, in Cincinnati, Ohio.

On the other side of the coin, certain immune disorders can make it difficult to gain or maintain weight, or they can cause unintentional weight loss. Being underweight can endanger health and keep the body from getting the nutrients it needs to look and feel its best. Some of the problems of being underweight include bone loss, decreased immune function and iron deficiency, among others.

Following are some tips for achieving a healthy weight loss or gain.
Healthy Weight Loss

Slow and steady reigns true when it comes to weight loss. Aiming for a 1-pound to 2-pound weight loss each week is a good place to start, and it reduces chances of regaining the weight. Cutting back on 250 to 500 calories each day by making good food choices, exercising or both can help individuals reach weight-loss goals in a more sustainable way. These methods can help to shed pounds:

Eat plant-powered foods. Putting plant-based foods like fruits, vegetables, beans and whole grains at the forefront of a diet can aid in weight management and immune support. Since immune disorders can impact the body’s immune system and the ability to fight off infections, including foods that support immunity such as plant-powered foods is especially important and beneficial to health and weight maintenance. Research has shown that those who follow a vegetarian diet have a significant benefit on weight reduction compared to nonvegetarian diets. This doesn’t mean everyone should adopt a vegetarian diet, but it helps to show that plant-powered diets can play an important role in weight maintenance.

One of the key benefits of plant-based foods when it comes to weight maintenance is fiber. A higher intake of fiber has been associated with lower body weights, as well as a reduced risk of heart disease and type 2 diabetes. Naturally occurring dietary fiber can be found only in plant-derived foods. Therefore, increasing the amounts of these foods in the diet is one of the easiest ways to increase fiber intake. Snacking on cut-up vegetables and fresh fruits and including at least one serving of either at each meal or snack are easy ways to put fiber at the forefront of a diet.

Unfortunately, certain immune disorders can make choosing the right fiber-rich foods a little more involved. This includes avoiding gluten-containing grains with celiac disease, limiting raw or undercooked vegetables, and avoiding nuts and seeds or specific foods that exacerbate symptoms, especially with Crohn’s disease or ulcerative colitis. Yet, while consuming lots of fiber-rich foods in the diet may not be suitable with all immune disorders, including as many as tolerable can help support weight maintenance and immunity.

Hydrate. Hydrating is not only important for daily bodily functions like waste removal, joint lubrication and nutrient absorption, it can also help when it comes to weight loss and maintenance. In those actively working toward weight loss, research has shown increased water consumption can help improve weight-loss efforts. Drinking more water can help decrease the amount of calories consumed, especially prior to eating a meal. A 2010 study in the journal *Obesity* found that those who drank about two cups of water prior to each meal lost an average of 44 percent more weight than those who did not drink water before meals.

While individual water needs can vary based on age, sex, exercise level and environment, The National Academies of Sciences, Engineering and Medicine recommends an average of about 12 cups to 16 cups of fluid for healthy adults each day. Individuals who struggle to consume enough water can try some of these tips:

- Drink a large glass of water first thing in the morning.
- Flavor water with frozen fruit, fresh herbs or unsweetened seltzer waters.
- Sip on herbal teas for an afternoon pick-me-up or after dinner.
- Keep a water bottle within arm’s reach during the day as a gentle reminder to stay hydrated.
- Snack on fruits and vegetables with a high water content such as celery, bell peppers, watermelon, berries, melon and cucumbers.

Lower the impact. Staying active with both daily movement and formal exercise is another important aspect of weight maintenance. This can sometimes be more difficult when faced with joint pain and mobility limitations or the threat of exacerbating symptoms that can come with certain immune disorders. But, opting for more gentle or low-impact exercise can be one way to make staying active more approachable and beneficial.

While staying physically active can be daunting, especially when pain, fatigue and stiffness are a part of living with an immune disorder, it has been shown to actually help improve symptoms overall. In fact, the incidences of multiple immune disorders such as rheumatoid arthritis (RA), multiple sclerosis (MS) and irritable bowel disease have been found to be lower in those who are engaged in physical activity.
Regular physical activity can also help to increase joint mobility for those with RA, improve mood and mobility in those with MS and help lower the risk of cardiovascular disease that can accompany many immune disorders.³

Low-impact exercise, including yoga, tai chi and walking, can be suitable options for many as a way to stay active, as well as to help alleviate symptoms. Individuals should find an activity they look forward to doing each day and that helps them feel their best. The key is for people to listen to their bodies and do what they have energy for. It is best to work with a healthcare provider to determine the best type, amount and frequency of low-impact exercise to include in weight-maintenance efforts.

Stress less. Eating right and staying active may be the first things that come to mind for weight maintenance, but one strategy that often slips under the radar is reducing stress. Not the short-term stress people may feel when giving a speech or running late for work. It’s the day-in and day-out stress (chronic stress) that can wreak the most havoc. Stress has been shown to have a connection to the increased risk of developing or exacerbating symptoms of immune disorders,⁶ and it can also interfere with weight-loss efforts.

Chronic stress can contribute to higher levels of the hormone cortisol in the body, which can also increase appetite. This can help to explain why many individuals “stress eat” or eat as a way to deal with stress. Stress is not a good feeling, and it can increase the desire for higher-calorie, heavy comfort foods like macaroni and cheese, ice cream or chocolate chip cookies.

Using stress-relieving or stress-management techniques can help control eating habits and how stress affects overall health. Some common practices include meditation, yoga, exercise, journaling and social support. Finding the method that works best is different for everyone, and it can take some trial and error before landing on the one or combination of many that works best for keeping stress under control.

Journal. Self-monitoring is an important aspect of weight loss, which can be accomplished by journaling or keeping a food diary. Those who track what they are eating tend to lose more weight than those who do not track or self-monitor. Being honest about what is consumed by writing it down can help to show that a small bite here, a couple handfuls there or an extra teaspoon or two truly add up.

Whether someone is more of a pen-and-paper journaler or a tech-savvy type, either method can be helpful for sticking to weight-loss goals. It comes down to being honest about everything that is consumed, both food and drink, during the day that makes the biggest difference between an effective and ineffective journaling experience.

Avoid triggers. Eating a well-balanced diet that includes an array of fresh fruits, vegetables, lean proteins and whole grains is beneficial for any weight-maintenance program. But, certain considerations should be included for immune disorders that often come with limitations or challenges when including some or many of these foods.

For those with celiac disease, avoiding gluten-containing whole grains is paramount, but they can be swapped for gluten-free options like quinoa, popcorn, brown rice, buckwheat and oats. For those with Crohn’s or ulcerative colitis, avoiding foods that tend to trigger flares should be avoided, and finding alternative choices to regularly include in the diet can help to support weight-loss efforts while controlling symptoms. Keeping track of when symptoms occur and what foods were eaten can also help rule out what foods to include or avoid, and help prevent symptoms from getting worse or continuing in the future.

Healthy Weight Gain

Preventing too much weight loss with immune disorders is just as important and impactful to overall health as losing excess weight. For those struggling to improve their daily diets, here are some ways to help support healthy weight gain:

Focus on fats. Per gram, fats have more calories than both carbohydrates and proteins combined, making them a concentrated source of energy for the body. For people who may not be able to tolerate or stomach a higher volume of
food or simply want an easy way to increase calories, opting for more sources of healthy fats in the diet can be effective.

Not all fats are created equal though. Choosing unsaturated and omega-3-rich fats helps to support immune function, heart health and general well-being, in addition to a hefty dose of calories. These types of fats include olives and olive oil, fatty fish like salmon and mackerel, nuts and seeds, avocados and nut butters.

Some ways to incorporate more fats in the diet are:
• Drizzling salads or cooked vegetables with olive oil;
• Snacking on apple slices or celery sticks with peanut butter;
• Opting for fatty fish like salmon at dinner meals;
• Adding avocado slices to salads, sandwiches or omelettes; and
• Blending in avocado or peanut butter into a morning smoothie.

Concentrated calories. Methods for gaining weight are almost the complete opposite of trying to lose it. While proper nutrition is still a focus, opting for more calorie-dense options instead of the lighter choices of many dieters can help keep the weight on.

When it comes to vegetables, including more starch-heavy options like potatoes, corn and peas, especially when paired with a serving of healthy fats like a drizzle of olive oil or cubed avocado, can boost calories in a much smaller portion than the same amount of nonstarchy vegetables like spinach, peppers or broccoli. Fruits like bananas and mangos or dried fruits like dates, apricots and figs are all more calorie-heavy than melons, berries or citrus fruits.

Blending multiple higher-calorie foods into a morning smoothie is an easy way to pack a lot of nutrition and support healthy weight gain. This can be a convenient option during busy mornings, when traveling or if someone doesn’t have a big appetite for a full meal. Some smoothie boosting mix-ins include:
• Banana with peanut butter or oats
• Mango and Greek yogurt
• Avocado and cocoa powder
• Dates and walnuts

Beverage choices. While downing a glass of water before a meal may help when it comes to losing weight, beverage choices at mealtime can make gaining weight more difficult. Consuming beverages toward the end of the meal or even spaced out from meals can make consuming more calories easier. Carbonated beverages such as seltzer or mineral waters can also interfere with weight-gain efforts since they can increase the feeling of fullness or cause bloating. These types of beverages should be avoided during mealtimes.

Lifestyle Change is the Key

Maintaining a healthy weight is key to keeping the immune system functioning at its best. While those with an immune system disorder may struggle with either losing or gaining weight, the key is to make gradual lifestyle changes that include healthy eating, regular physical activity and other healthy habits that become lifelong behaviors.

EMILY COOPER, RDN, is a nationally recognized registered dietitian, health writer and recipe developer based in New Jersey. She is the author of The Easy Superfoods Cookbook and the website sinfulnutrition.com.

Reference
Profile: Michelle Searle

By Trudie Mitschang

As a young adult living with common variable immune deficiency (CVID), Michelle Searle has been fearlessly pursuing her dreams and refusing to let her primary immunodeficiency disease (PI) hold her back. After spending several years as a teen advocate for the Immune Deficiency Foundation (IDF), Michelle is now living abroad in Italy where she is fulfilling her goal of obtaining dual citizenship and embarking on a teaching career. With tenacity and grit, Michelle is an inspiration for anyone who strives to live life without limitations, ignoring the naysayers and maintaining a positive outlook no matter what.

Michelle Searle has overcome the challenges of living with CVID and is pursuing her dream of living and gaining dual citizenship in Italy and working as a teacher.

Eventually, we were sent to see a specialist who diagnosed me with CVID.

Michelle: CVID has, of course, impacted my health in a negative way, but in the other aspects of my life, CVID has impacted my life in a very positive way. I don’t think I would be as empathetic and compassionate if I wasn’t living with a chronic illness. I also don’t think I would be able to truly appreciate my life and all the moments when I feel healthy. CVID has given me more strength, more opportunities, more knowledge and so much more. Growing up with this disease has instilled and strengthened many of the positive traits I have today.

Trudie: What motivated you to become involved with IDF?

Michelle: Being involved with IDF reminds me I’m not alone, and it shows me the positive parts of this disease. I’ve met incredible people I would not have met otherwise, and I have traveled to many different states for IDF events I’ve participated in. When I talk with someone I’ve just met and we’re able to relate with each other about our PI, it’s a connection like no other. It keeps my attitude and spirits high when I meet older people with a PI who are thriving with it. It inspires me and motivates me when I meet younger people with PI to keep going because if they can do it, so can I. I’m always learning something new at IDF events, as well as from people who attend them.

Trudie: Tell us about your experiences as an IDF Teen Council Member.

Michelle: Although I’m a pretty independent person, being a Teen Council Member allowed me to become...
even more independent. I started flying and staying in hotels by myself and transitioning into leadership roles. Listening to other teens talk about their experiences with their illnesses allowed me to see what I went through as a child/teen with CVID from someone else’s perspective. I started to understand how difficult it must have been for my parents and how strong I really was growing up with CVID. I was in the position to help and inspire others with a PI, which in turn helps me deal with my own issues.

**Trudie:** What was it like attending Advocacy Day on Capitol Hill?

**Michelle:** Attending Advocacy Day on Capitol Hill was something I initially didn’t believe I could do. I was so nervous just thinking about talking to legislators! I then realized we all have a story to tell, and it’s essential for people in positions of influence to hear our voices. Over the years of attending, I could see the difference it was making, not only in the rare disease community but also within myself. It’s a very empowering experience, and I’m so grateful for it. This past year, I attended with my sister Mandy, and watching her advocate for me and everyone else with a PI was an amazing reminder of the support I have around me.

**Trudie:** What inspired you to live abroad?

**Michelle:** I graduated college in 2018 with a degree in elementary education. My mom’s side of the family is from Italy, and I’ve always been interested in learning more about the culture and language. I told myself many years ago I would live in Italy so I could learn the language by being fully immersed in it. I also wanted to obtain my dual citizenship. I started working a second job in 2017 to save for Italy, and I worked two jobs until I moved to Italy in September 2019. I first lived in Florence and went to school to become certified in teaching English as a foreign language. I then moved to Pescara and applied for my dual citizenship. I’m in the process of achieving that now, and I just recently got a job teaching English at a private language school. Italy has been the most incredible experience. Although it’s hard to be away from loved ones, and it can be scary when I become sick because I’m not near my doctors or family, I’m so thankful to live in an environment that teaches and challenges me every single day. I’m grateful my parents raised me without ever putting limitations on me because of my CVID. I can do anything someone without a PI can do; it just takes some extra planning.

**Trudie:** Has the outbreak of COVID-19 in Italy impacted you personally?

**Michelle:** Currently, it has not impacted me other than having to cancel trips to northern Italy. I’m just being more conscious of how often I wash my hands, touch my face, wipe down surfaces on buses I take and living my life as normally as possible.

**Trudie:** What have you found most challenging about living with a PI?

**Michelle:** I’m someone who gets nervous going to the doctor, so having to see a doctor as often as I do has not been easy. Being diagnosed at a young age, the most challenging parts were more external: things like needles, side effects and medication. Now that I’m older, the most challenging parts are internal: the fear and worries about what my future health will look like. For instance, I wonder: Will I be able to work as a teacher without getting sick all the time? Will I pass this onto my children? Will I develop other complications or illnesses the older I get? It can be challenging sometimes to calm the doubts, fears and worries in my head. I’ve also had to learn to not let other people’s fears and doubts affect me. I’ve had multiple people tell me I won’t be able to work as a teacher with CVID, and I shouldn’t even try. Instead of projecting our fears onto others, we should try to motivate and encourage others, so I do my best to tune out that negativity.

**Trudie:** What are your goals for the future?

**Michelle:** For the longest time, my main goal had been to move to Italy, and now that I’m living here, I’m assessing what new goals I have for myself. I know I’d like to become fluent in Italian, eat healthier, adopt some cats and become more involved as a patient advocate.

**Trudie:** What advice do you have for other young people living with CVID?

**Michelle:** Get involved, advocate and spread awareness for PI, whether that’s with other people in the PI community or just in your community in general. Go out and meet other people with PI, thank plasma donors, have honest conversations with your doctors and make sure to focus on your mental health, as well as your physical health. Your mental health is just as important!

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

Laugh It Off!
By Whitney L. Ward

I STAND at a towering height of 4 feet 9 inches. At times, it’s been frustrating, but I’ve learned to laugh at the blunders my height causes such as being asked if I want a children’s menu at restaurants or being mistaken as the child of my sister who is only two years older than me. I’ve learned to own my uniqueness. Laughing at questions I get from people because of my Polly Pocket status is an art I’m glad I mastered because it got me through a blunder to end all blunders.

I was a junior at Asbury University gaining confidence I never had before. I was making new friends. New possibilities were on the horizon. I was gaining a swagger I never knew I had. So, at dinner one night, I decided to venture out and eat at the main buffet for dinner — the side I never dared crossing because that’s where all the jocks and legacy students sit. But, on this night, I was bound and determined to feed my newfound confidence.

I joyfully walked with a spring in my step to the back of the line and grabbed a plate with a big smile on my face. The girl in front of me kept looking over her shoulder. You know those kids who stare at you like you’re growing a horn in the middle of your forehead? Yeah, it was like that. So, I offered a smile, which caused her to face me. When I saw this girl’s face, I immediately thought she was a freshman or possibly even a prospective student checking out the campus.

“Can I ask you a question?”

“Sure!” I said, fighting the urge to clap enthusiastically, thinking she was going to ask me where the biology building is or for directions to the library. I was fully ready to give Asbury University an amazing endorsement and reasons why she should attend my college, when she hit me with: “Are you a dwarf?” Lips pursed, eyebrows scrunched, she leveled me with a solemn look every doctor gives when they are about to give a shocking diagnosis. Which, little did I know, she was.

When someone makes an ignorant comment at your expense, laugh it off.

I stared at her for a few seconds, baffled at the words my unsuspecting ears had just heard. “Um, no, I’m not a dwarf. I’m just really short.”

Pity filled her eyes as her mission became clear: to educate me into acceptance. “I actually really think you are. You see, I did a paper on dwarfism, and I’m pretty sure you’re a dwarf.”

My mouth dropped as I fought the urge to look around for a camera crew filming an episode of “Punk’D.”

“Uhhh, no, I promise you I’m not a dwarf. I’m just extremely short.”

There, that would be the end of it. Wrong, again.

“But, I think you are, because I wrote a paper on dwarfism.”

Torn between knocking the girl over her head with my dinner plate or laughing in her face because of her complete lack of a filter, I raised my hand to put an end to the episode of the “Twilight Zone” I had accidentally been zapped into.

“I promise I’m not a dwarf. But, you have a great night.”

I giggled all the way to my seat, thinking, “Is this really my life?”

The takeaway of the hilarity is this: Yes, there will always be someone who thinks Google research gives them more expertise than what you have lived. Laugh it off! You know your authenticity, and the lack of someone’s validation or understanding doesn’t make your experience less true. And, yes, your disease may give you something that makes you unique, but I encourage you to own it! When someone makes an ignorant comment at your expense, laugh it off.

Dr. Seuss said it best: “Why fit in when you were born to stand out?” God made you knowing how much this world needs the colorful and inspiring “You.”

After all, learning to laugh when someone makes you feel “small” will only cause you to grow — pun intended.

WHITNEY L. WARD was not only the first person in the world diagnosed with MAGIS syndrome, she had the honor of naming the new primary immune deficiency. MAGIS means “more” in Latin, and Whitney hopes to instill in her readers the message they are more than their disease. Find out more about Whitney’s journey at www.whitneylaneward.com.
Blunt Advice for Thriving with Chronic Illness

By Ilana Jacqueline

AFTER BEING diagnosed with a chronic illness, I was learning for the first time that a lifetime of symptoms weren’t a result of anxiety or boredom or hysteria. Now that I have a diagnosis, here’s what I want you to know and what I wish I would have known at the start of this very long and strange journey:

We have barely scratched the surface of diagnostics. Doctors aren’t gods, and the ones who claim they know all and have seen all are not the doctors you want to put your life in the hands of. Fight for genetic sequencing, demand every test and exam and follow every hunch until you find the reason why you’re as sick as you are.

DIY your own recovery as you go. In all my years of multitude abdominal surgeries I underwent that destroyed my abdominal muscles, not one single surgeon or doctor ever prescribed physical therapy for me. No one even suggested it. It blows my mind how no one thought to introduce me to the world of pelvic floor therapy or physical therapy. Diagnosis or not: Don’t let your body fall further into disrepair when you have a chronic illness. Get your PT on now!

Take charge of your mental health. You may be hearing “it’s all in your head” when it comes to the physical manifestation of your disease, but what you fail to hear is “it’s a lot of fear and anxiety and stress to manage a chronic illness, and you deserve support.” Find a good therapist, someone who pushes you to keep going when the going gets tough. Look for someone who won’t let you wallow in depression and who will intervene when the stress gets to a boiling point. #TherapyIsCool

Don’t let your disease define you. In the beginning, it may seem enticing to join a thousand support groups and lament in the misery that is the reality of your disease. But you’re not just a spoonie or a warrior, you’re a whole you made of a million different pieces. Don’t box yourself in on the worst part of your existence. Get support and be supportive, but don’t let it be your identity.

Stop falling for scams. If they found a cure for cancer, it would be front-page news. If they knew of a diet that reversed a disease, no one would keep it a secret. And, the same goes for cure-all clinics that don’t take insurance and all the Drs. House who will give you all the time and sympathy in the world (for enough money) but won’t give you a straight answer on what’s wrong and how to fix it. Stop joining multi-level marketing schemes whose 20-supplement-a-day regimen claims to cure everything from rheumatoid arthritis to pancreatitis. Think. Research. Investigate.

Keep living every part of your life, even when it seems impossible. Because you won’t always be this sick. Your illness won’t always be the focus of your existence. You need friends and family and an education. You need a career that gives you purpose. You need love. You need to explore your talents and dreams. You might have to do it from your bedroom or your bathroom floor, but keep your head up and live as much as you can. Impossibly, unimaginably, life goes on. Go with it. And don’t be afraid that enjoying small moments will somehow hinder your road to a diagnosis.

After all, what’s the point of fighting so hard to live if you aren’t creating a beautiful life? Pursue your own journey; take on your own investigation.

Author’s note: This article was adapted from my blog at www.letsfeelbetter.com published Oct. 4, 2019.
Swimming Health Concerns for PI Children

By Jessica Leigh Johnson

As residents of the “Land of 10,000 Lakes,” my family and I spend a lot of our summers in the water. Whenever the temperatures rise above 80 degrees, we head to the cabin for a day of swimming, paddleboarding and lounging on our new favorite toy: the aqua mat. I’m sure most parents share my opinion that any time kids spend outside enjoying the sun is far better than time spent indoors staring at a screen. But when stories about bacteria outbreaks at public pools or flesh-eating parasites found in lake water make headlines, I wonder if my children might be safer indoors. Parents of primary immunodeficiency disease (PI) children are used to taking extra precautions in certain situations. But, is swimming in pools, lakes or the ocean simply another area that requires parents of PI kids to be extra vigilant, or is it something our kids should avoid altogether?

Pools

Last summer, a bacteria called Cryptosporidium made headlines as dozens of people started coming down with cryptosporidiosis, which causes watery diarrhea and can last for up to three weeks after swimming in public pools. The bacteria from the feces of an infected swimmer enters the pool, and is transmitted to other swimmers when they swallow the contaminated pool water. Unfortunately, this bacteria is very resistant to chlorine and can survive in a chlorinated pool for up to seven days.\(^1\)

Between 2009 and 2017, there have been 444 outbreaks of cryptosporidiosis in the U.S. and Puerto Rico. The number of outbreaks has been on the rise, increasing 12.8 percent each year since 2009. While public pools, including kiddie pools and water parks, are responsible for 156 (or one-third) of those outbreaks, untreated water found in lakes, well water and other sources contributed to 22 cases. Unfortunately, other than teaching kids not to swallow water when swimming, there isn’t much parents can do to prevent their children from acquiring this type of infection. To help stop the spread of the illness, the Centers for Disease Control and Prevention (CDC) recommends anyone who has suffered from diarrhea avoid swimming in pools for two weeks afterward.\(^1\)

Fresh Water

Before the threat of gastrointestinal infection causes parents to shred their local pool membership cards, they should be aware that there are plenty of recreational water illnesses that can be found in fresh water as well. According to the CDC, “Oceans, lakes and rivers can be contaminated with germs from sewage spills, animal waste, water runoff following rainfall, fecal incidents and germs rinsed off the bottoms of swimmers.” Besides diarrhea, contaminated water can also cause infections of the skin and ear.

“Swimmer’s itch,” officially known as cercarial dermatitis, is an itchy rash that can occur after swimming and is most common in freshwater lakes and ponds. Most often, swimmer’s itch is a reaction to parasites that burrow into a
A beach website at www.epa.gov/beaches has been tested, or they can go to the Environmental Protection Agency’s (EPA) website since many U.S. beaches are contaminated. Once they’re home, eating in the sand, which can also be contaminated. And, although that is enough to scare any parent right out of the water, swimming is a fun activity that kids with PI shouldn’t have to miss out on. Taking wise precautions and being vigilant about hygiene can ensure that kids stay as safe as possible.

Saltwater

Since saltwater is known for its antiseptic properties, swimming in the ocean must not pose any health threats to PI children, right? Unfortunately, a type of bacteria known as enterococcus, which causes infection in the gastrointestinal tract, can be found in the ocean’s salty water. This bacteria is commonly found in fecal matter. During times of increased rain, water levels rise and can carry waste upstream from septic tanks and treatment centers and into the ocean. Another scary but rare bacteria found in saltwater is the potentially deadly “flesh-eating” bacteria called Vibrio vulnificus. This rare infection is spread through warm seawater and raw shellfish, and it can affect the skin or the gastrointestinal tract. While most people aren’t at risk, people with compromised immune systems such as those with PI or who have open wounds could experience ulcers, amputation, infection, and potentially death.

When deciding whether or not to swim at an ocean beach, it’s best to use common sense. If there aren’t many people in the water, the beach could be closed — and for good reason. It might be wise to check the EPA website since many U.S. beaches are regularly checked for bacteria, although it is not a federal requirement to post signs when the water’s bacteria levels go beyond the federally established limit. Parents should encourage their kids to wash their hands before they eat if they’ve been swimming or digging in the sand, which can also be contaminated. Once they’re home, kids should shower their entire body. Again, teaching kids never to swallow any swimming water is a must. So is scouting out the area surrounding a swimming beach to be sure there are no discharge/run-off pipes nearby, which can be a sign that environmental pollutants are dumped into the water. It’s also a good idea to have kids avoid swimming if they have open wounds or sores, as these allow bacteria to enter the skin.

Be Vigilant

When it comes to waterborne pathogens, children, pregnant women and people with weakened immune systems are at increased risk of severe illness, which means swimming — with or without a lifeguard present — should be done at one’s own risk. Parents of children with weakened immune systems must keep in mind that any recreational water, whether it’s a lake, pool or ocean, might be contaminated. And, although that is enough to scare any parent right out of the water, swimming is a fun activity that kids with PI shouldn’t have to miss out on. Taking wise precautions and being vigilant about hygiene can ensure that kids stay as safe as possible.

References


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.
Safely Soaking Up the Sun

By Heather Bremner Claverie

THE SUN IS not only essential to survival, it can boost mood and well-being. Yet, it can also be deadly. Most skin cancers are caused by an excessive amount of exposure to ultraviolet (UV) rays, the radiation emitted from sunlight. Fortunately, there are many ways to enjoy fun-filled outdoor activities while protecting the skin and eyes from harmful rays.

The Source

Deep-seated wrinkles are caused by UVA rays that have the least amount of energy from UV rays, which cause skin cells to age and are mainly linked to long-term skin damage. And, painful, red sunburns are caused by UVB rays that have more energy than UVA rays and that damage the skin cells directly, triggering sunburns and causing most skin cancers.

While sunlight is the main source of this type of electromagnetic radiation, UV rays can also result from man-made products such as tanning beds and sunlamps.

Who Is at Risk?

Although any exposure to sunlight between 10 a.m. and 4 p.m., particularly during the spring and summer months, can cause skin cancer, it’s really long-term exposure that is most harmful. Children who suffer from multiple sunburns throughout childhood or individuals who work in the sun all day every day are at a higher risk of developing skin cancer.

In addition, certain risk factors can cause some individuals to be more susceptible to developing skin cancer. Fair-skinned individuals and those with freckles, blue and green eyes, and blond, red or light brown hair should be particularly vigilant when in the sun.

The Best Protection

Individuals can still enjoy the great outdoors under a veil of protection. The best advice is to stay in the shade by bringing an umbrella to the pool or finding a nice leafy tree to shield the sun.

Protective clothing can also help. If light filters through fabric, UV rays can seep through, too. Long-sleeved shirts and pants with tightly woven fabrics are the best bet to combat the sun. A wide-brimmed hat with at least a 2- to 3-inch brim that protects the face, neck and ears is essential. And, there is UV-protective clothing available, as well as products that give clothing SPF protection. Also, people should not forget sunglasses.

The Skinny on Sunscreen

Sunscreen is crucial sun defense, but only when reapplied amply and often. A shot glass full of broad-spectrum cream or spray of SPF 30 or higher should be applied and reapplied at least every two hours. When swimming or exercising, it may be necessary to apply it more often.

The higher the number on the sunscreen bottle, the higher the level of protection, but there is a point when a higher number is no longer more protective. For example, SPF 50 already filters out approximately 98 percent of UVB rays.

Enjoy That Fire-Filled Star Safely

It’s not surprising that people often feel energized and optimistic after a day in the sun. Vitamin D, the so-called “sunshine vitamin,” is derived from that golden orb. In addition to reducing depression, vitamin D boosts the immune system and fights disease. While certain foods and supplements can help satiate a vitamin D deficiency, many people want to enjoy those sun-kissed days. The good news is they can, as long as they slather on the sunscreen, throw on a straw hat and don sunglasses.

HEATHER BREMNER CLAVERIE is a contributing writer for IG Living magazine.
Coolibar sells an array of sun-protective apparel and accessories and all its fabrics are UPF 50+. The Minnesota-based company says it refers to all its products as “UPF 50+” rather than “SPF” because SPF is specific to the amount of time sun-exposed skin is protected when wearing sunscreen, while UPF indicates what fraction of the sun’s UV rays can penetrate fabric. Coolibar’s lightweight bathing suits, shirts, pants and hats are designed with breathable, fast-drying fabric. Prices vary; [www.coolibar.com/featured-collection.html](http://www.coolibar.com/featured-collection.html)

Cover Up

Go Undercover

Made with Solarteck sun-blocking material, UV Blocker’s UV Protection Travel Umbrella measures 44 inches and will block out 99 percent of UVA and UVB rays. Also, the company’s patented Vented Mesh System allows wind to pass between the upper and lower canopies, keeping individuals 15 degrees cooler. Even better, the umbrella is light and small enough to fit into a carry-on bag or suitcase. $44.95; [uv-blocker.com](http://uv-blocker.com)

Shopping Guide to Sun Safety

Slather It On

Badger’s Tangerine and Vanilla Broad Spectrum SPF 30 sunscreen with its Creamsicle-like smell is designed for children. And, since active, young children need an ample amount of sun protection, this sunscreen offers a protective coat that lasts up to 40 minutes when swimming and sweating. $12.95; [www.amazon.com/Badger’s+Tangerine+and+Vanilla+Broad+Spectrum+SPF+30+Sunscreen](http://www.amazon.com/Badger’s+Tangerine+and+Vanilla+Broad+Spectrum+SPF+30+Sunscreen)

Stay Sensitive

Babyganics Sunscreen Lotion 50 SPF is a broad-spectrum lotion with UVA/UVB protection for babies. Non-allergenic and tear-free, its plant-derived ingredients provide babies with a fragrance- and paraben-free lotion. $13.97 for 6oz 2 pack; [www.amazon.com/Babyganics-Baby-Sunscreen-Lotion-Tube/dp/B00HYV2F7E](http://www.amazon.com/Babyganics-Baby-Sunscreen-Lotion-Tube/dp/B00HYV2F7E)

Pucker Up

The lips are often neglected when it comes to sun protection, but Sun Bum’s fun line of colorful tinted balms places them in the spotlight. The company’s Original SPF 30 Sunscreen Lip Balms are filled with aloe and vitamin E to protect and moisturize. All of Sun Bum’s products are made for sensitive skin and are vegan- and paraben-free. $3.99; [www.sunbum.com/collections/sunscreen-plp/products/spf-30-sunscreen-lip-balm-pomegranate](http://www.sunbum.com/collections/sunscreen-plp/products/spf-30-sunscreen-lip-balm-pomegranate)

Just Wash It

One laundry cycle with the Rit Sun Guard Laundry Treatment UV Protectant will give clothes 20 washes worth of sun protection. The versatile product is safe to use with any washable fabrics, including cotton, linen, rayon and silk. While it won’t alter the look or feel of the clothing, it will provide a layer of UPF protection of 30. $24.95; [www.amazon.com/Rit+Sun+Guard+Laundry+Treatment+UV+Protection-Protectant&qid=1584466998&s=8-3](http://www.amazon.com/Rit+Sun+Guard+Laundry+Treatment+UV+Protection-Protectant&qid=1584466998&s=8-3)
**New and Useful Reading**

Authors: Frances Cole, MD, Helen Macdonald and Catherine Carus
Publisher: Robinson

This self-help book is based on highly effective self-help methods developed by specialists and used in community and hospital pain management programs. The authors describe how the experience of pain can be greatly reduced by pacing daily activities, reducing stress and learning relaxation techniques and effective ways to cope with depression, anxiety, worry, anger and frustration. Topics covered include why pain can persist when there’s no injury or disease present; how to become fitter and pace activities; practical ways to improve sleep and relaxation; and tips for returning to work, study and gaining a life.

**How to Be Sick: Your Pocket Companion**
Author: Toni Bernhard
Publisher: Wisdom Publications

In this easy-to-use, easy-to-carry book, Toni Bernhard shares practices from her bestselling classic *How to Be Sick* and also offers new suggestions and strategies for coping with a life impacted by chronic pain and illness. Because the book is organized by specific challenges, readers can immediately find practices that can help when they’re needed most.

**I Need to Manage My Invisible Chronic Illness: Practical Strategies to Manage the Struggles and Live a Productive and Enjoyable Life**
Author: Brenda Koranda, RN, MSN
Publisher: Difference Press

Brenda Koranda, a registered nurse for more than 28 years who currently assists those with chronic illness to make informed decisions about their health and well-being, has been managing invisible chronic illness for over 16 years. In this book, she shows readers how to manage invisible chronic illness to live a happy and meaningful life with strategies to accept a new “normal”; the importance of becoming an expert on each person’s invisible chronic health condition; how to build a healthcare and support team; strategies to manage the common struggles invisible chronic illness presents; and more.

**Crazy, Chronic Life: A Handbook for Living with Chronic Illness**
Authors: Joe Early and Tiffany Early
Publisher: Independently published

This guidebook discusses everything from faith to fears to bodily functions — basically, the scope of the frustrations that come with living the sick life. Readers will receive practical tips for surviving chronic illness and still managing to live a full life. Topics include marriage and romantic relationships, caregiver advice, traveling, research and advocacy, and many other aspects of life with chronic illness.
“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
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