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Improving Health with Technology

AT THE forefront of concern during this COVID-19 pandemic are approaches to ensure individuals’ well-being. The good news is our healthcare system is poised to deal much better with care as a result of technological advances that weren’t available during previous full-scale pandemics like those that occurred in the 19th and early 20th centuries. In addition to manufacturing and research efforts to develop equipment and medicines to treat patients, conduct testing and collect data to better understand how people’s bodies react to this new virus, and the quest to develop a preventive vaccine, technology also plays a prominent role right now at the patient and provider levels.

One technological advancement that has been around for some time albeit underutilized is telemedicine, and its usefulness to immune deficient patients who are at increased risk of infection can’t be overstated. We discuss some of the benefits of telemedicine in our article “Can Telemedicine Benefit PI Patients?” (p.16). But, more specifically, we look at how telemedicine has been successfully used in one Southern California health system and how it paved the way for its use when the pandemic struck. Yet, while this case shows patients can adapt to a new way of communicating with providers, whether telemedicine will remain a standard of care in the long term depends upon overcoming several challenges, including licensure requirements and insurance reimbursement.

Telemedicine is primarily initiated at the provider level, but there are many technologies patients can take advantage of on their own. In our second-in-a-series special feature “New Tools and Tech for Patients” (p.24), we identify some state-of-the-art health apps patients can use to improve and monitor their health. From personal comfort and pain relief to sleep quality, in-home health monitoring, emotional and stress management and personal safety, these healthcare tools and apps can also help to communicate with family and friends, as well as provide feedback to caregivers.

Another technology becoming more mainstream is biofeedback, which uses external stimuli to influence how the body responds to stressors that affect everything from emotional health to physical pain. As we explain in our article “Biofeedback for Pain, Stress and Anxiety” (p.28), patients work with biofeedback practitioners to learn how to control their body’s responses to triggers with the help of devices that measure autonomic responses such as temperature, breathing, heart rate and more, and then provide feedback via sounds or indicators on a screen. The goal is for individuals to learn how to control those autonomic responses.

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

Ronale Tucker Rhodes, MS
The Importance of Care During COVID-19

By Abbie Cornett

A YEAR AGO, who would have thought that instead of saying “goodbye,” the new salutation would be “stay healthy,” or that we would be standing in line to enter a store to buy groceries or, worse yet, the shelves would be empty of many of the items we took for granted such as toilet paper, flour and Lysol? Seriously, I doubt the best fiction writers ever dreamed eating in a restaurant would be viewed as an act of faith, or wearing a mask would become a political statement!

But, these are just some of the obvious ways COVID-19 has changed daily life. Unfortunately, the less obvious ways life has changed may be those that impact people most seriously. Since the COVID-19 outbreak, people have stopped seeking both emergency and nonemergency medical care because they are frightened of being exposed to the virus. This means people who need care aren’t getting it. A recent poll conducted by The Hill shows a substantial increase in the number of people skipping dentist appointments, annual exams, medical consultations, elective surgeries, counseling sessions and physical therapy. While this resistance to going to a doctor may be limiting people’s exposure to the virus, it is not limiting their overall risk. For example, forgoing a dentist appointment for an annual cleaning may not be that big of a deal during COVID-19, but not going to the emergency room with chest pain is!

Also since the COVID-19 outbreak, hospital admissions for other critical conditions such as strokes, diabetic reactions and heart attacks have plummeted. Many doctors fear the illness and mortality from unaddressed health problems may rival those of COVID-19. In response to concerns that patients are not seeking needed treatment, the American College of Cardiology launched a campaign titled “Cardiosmart.” The focus of the campaign is to encourage patients to seek immediate medical care if they are showing symptoms of a life-threatening illness, and to keep their medical appointments through telemedicine or in person as needed. Managing overall health during COVID-19 is as important as seeking emergency treatment. Of particular concern are patients with time-sensitive conditions such as chronic disorders, children, the elderly and the injured.

Time-sensitive reasons to seek medical attention are those in which a patient needs treatment or regular monitoring. Chronic illness, as an example, hasn’t taken a vacation from COVID-19. Disease management for patients with a chronic illness means keeping regularly scheduled appointments to monitor progress and to ensure treatments are working. While much of this can be done remotely, some things such as lab work and medical testing need to be done in person. Other time-sensitive needs include preventive medicine and diagnostic treatments such as mammograms, biopsies and colonoscopies. Further, pain management treatments that improve a patient’s quality of life are very important to relieve suffering from conditions such as nerve pain, arthritis, migraines and chronic inflammation. Importantly, children need to get immunized to protect them from preventable diseases such as measles, mumps, rubella, polio and more. These immunizations are given on a schedule that is important to maintain. 3

Healthcare professionals are doing everything they can to ensure patient safety, but part of the responsibility is also the patients’. When going to visit a healthcare professional in person, there are several things people can do to help protect themselves, including practicing proper hand hygiene, not touching their face, eyes or mouth, following social distancing recommendations and wearing a cloth face covering.

When evaluating whether to seek treatment or not, individuals should ask themselves two things: 1) Is this something that will get better on its own, or is help needed? 2) How much is the medical issue impacting quality of life? While COVID-19 has changed the way people seek medical care, it doesn’t mean we should stop getting the care needed.

References

ABBIE CORNETT, is the patient advocate for IG Living magazine. She can be reached at patientadvocate@igliving.com or (800) 843-7477 x1366.
How Do You Control Your Weight with an Immune Disorder?

I have been taking prednisone for about 12 years. The dosage has been as high as 80 mg. I’m now on 18 mg daily. It is very tough as I am hungry most of the day and night. I have been with the same nutritionist for over 15 years. She has taught me the foods to eat that will not put the weight on. Not a diet! We talk once a week to see how I’m doing. She has given me the tools I need to keep the weight off. I eat lots of protein, chicken, fish, meat, vegetables such as a good salad, [and] I love milk, which is high in protein. My weight has only fluctuated by 5 pounds in all the years. — Harriett HW

I have not been able to lose weight in decades over the years. I’ve been on lots of different pain medications that pack on the pounds. I used to exercise three to four times a week when I felt good in the warm water therapy pool, but since COVID [started], pools are closed, and the health center where I go is still closed as well. I have not exercised since March. I was in Arizona at my mom’s house for four-and-a-half months, and I didn’t have access to any exercise. It was too hot to exercise outside with a walk. Now, I’m back home, and I do have dumbbells and a jump rope and a stability ball and exercise DVDs, but to be honest, I just don’t have the energy. — Rachel D

How Have You Adjusted to Life After Diagnosis?

It was a relief to be diagnosed. However, it was a tsunami effect for my husband and me. He has been my unceasing and supportive caregiver. We had so many plans, but with years of extreme sickness, co-pays and deductibles, our lives changed abruptly. “Adjust” is a daily endeavor. When we do make plans, we just roll with the punches and disappointments and have learned to be thankful for the accomplishments. IG therapy is a game-changer and a blessing! — Kay PB

My disease doesn’t make any difference for any of my friends, and I really don’t have to cancel anything because there are not really any plans to make because I don’t have money to go anywhere. For me, there really has not been that much of a difference. I have not been working since 2007, and I was diagnosed in 2009. I didn’t start therapy until 2014, and I’m still not working, so there’s not really any difference other than I do my own subcutaneous infusions every week to keep my levels balanced. I don’t have to use as much premedication as those who have to go to a hospital. I love being able to infuse whenever I want, wherever I want, in any state I want. I take a Benadryl 30 minutes prior to inserting my needles, and I take 400 mg of ibuprofen and use a numbing cream prior to inserting my needles. That’s it. Sometimes, I’m a little bit tired on Tuesday morning, but that’s not from the infusion, it’s from the Benadryl the day before. — Rachel D

[I adjust with] daily adjustments and only tentative plans as they often have to be cancelled. Living like a recluse is necessary, and [I have been] gradually abandoned by friends and family who just don’t understand or don’t want to. It’s not a choice but a necessity, and after 30 progressively worse years, I sometimes wonder if it’s worth it, but continue to hang in there. — Carolyn W

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

I am a chronic lymphocytic leukemia patient who receives monthly immune globulin (IG) infusions to boost my immune system. Those infusions include IgA, IgG and IgM antibodies. Even though I have never had any herpes symptoms, I underwent a test for herpes simplex virus (HSV)-1 and -2, both of which came up positive because of elevated IgG. Is it possible my IG infusions led to a false positive, indicating I have the herpes virus when in fact I might not?

Abbie: I spoke with Roger Kobayashi, MD, an allergy and immunology specialist in Omaha, Neb., regarding your question, and he said yes, intravenous IG (IVIG) and subcutaneous IG infusions are high in anti-HSV antibodies (i.e., passive antibodies). IVIG has very high titers against HSV-1, but less so against HSV-2 because virtually everyone has been infected with HSV-1, but not many have been infected with HSV-2. Therefore, the donor pool for HSV-2 is expected to be lower, meaning a positive test is very likely to be a false positive.

Terry Harville, MD, PhD, medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, says serologic testing while on IVIG is essentially invalid. The IVIG can mask some results that should return positive because of a disease process. Further, due to the potential for autoimmune antibodies to be present in the source plasma used to produce IVIG, tests can become positive in otherwise asymptomatic patients. This means ANA and RF can show up in people receiving IVIG who do not otherwise have symptoms, and they can be absent in patients with antibody deficiencies who have actual disease.

While it’s not ideal, Dr. Harville says you could have serologic testing performed immediately before (at the end of the half-life of the IG antibodies from the infusion) and immediately after an IVIG infusion. If both are identical, this could represent a disease process. And, if the test result prior to treatment is higher than the test result after treatment, this may also represent a disease process. However, if the test result after treatment is higher than the test result before treatment, the results are likely false due to antibodies acquired from the IVIG.

Antibody tests that should be performed include ANA, RF, cyclic citrullinated peptide (CCP), SSA and SSB. In particular, there should be a request for an IgA RF and an IgA CCP. These antibodies are not typically transmitted through IVIG, but if a patient has low IgA levels, these also may be invalid. Antibodies to SSA and SSB are used to diagnose Sjögren’s syndrome. Tests for erythrocyte sedimentation rate, C-reactive protein, complete blood count with differential and platelet count are also useful. Additionally, checking C3 and C4 (proteins that are part of the immune system) can be helpful since these may be elevated in patients with active RA. Peripheral neuropathy may be due to another process so it needs its own specific evaluation.

Is a positive ANA test accurate when receiving IVIG?

I have common variable immunodeficiency, Sjogren’s syndrome and a positive rheumatoid factor (RF) test, so they suspect I have rheumatoid arthritis (RA). My blood work and antinuclear antibody (ANA) tests were normal when tested for Sjogren’s several times through the years. I was started on intravenous immune globulin (IVIG) therapy 10 years ago, and my ANA tests were still normal. However, most recently, I had an ANA test that came back positive at 1:320. Is this test accurate since I am receiving IVIG? Also, are there other blood tests I should consider since I am receiving IVIG and have a positive ANA test? I have been symptomatic with joint pain, and I recently developed peripheral neuropathy.

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» Have a question? Email us at editor@IGLiving.com. Your information will remain confidential unless permission is given.
IN 1963, Gell and Coombs reported on a system of classification of histopathologic reactions observed in tissues through a microscope. What they named type I hypersensitivity is what we now know as true allergic disease mediated through IgE activation of mast cells. The reactions can be annoying such as a stuffy runny nose, sneezing and itchy eyes, but they can also be life-threatening if even a minute amount of an offending allergen is ingested. Common aeroallergens include grass, weed and tree pollen; mold spores; dog and cat danders; and dust mite and cockroach danders. All of these aeroallergens have digestive enzymes the body may perceive as parasitic attacks, which activates the Th2 immunity that provides protection from parasites, but when overactive also leads to allergic disease. As a consequence of the forces of evolution, we needed protection from parasites.

In the recent era, we have become more hygienic, so this strong evolutionary-designed immunity now results in allergic disease, apparently due to the lack of parasite exposure. When an allergen attaches to IgE bound to the surface of mast cells, it triggers the release of several noxious substances, including histamine (the process known as degranulation). Allergens were designed to cause a hostile local environment, thereby resulting in the expulsion of parasites. But in the allergic individual, they result in the features of allergic disease.

IgE-mediated disease is dose-independent, meaning even the tiniest amount of an offending allergen can trigger a response. Therefore, for example, if drinking one glass of orange juice causes no issues, but two glasses of orange juice causes a rash, or if eating six strawberries causes no issues, but after eating 10 strawberries a rash occurs, these are likely not due to a true allergic disease manifestation. Instead, they are dose-dependent reactions that do not occur via an IgE-mediated response. Mast cells can be sensitive to chemical, physical and mechanical forces, which can result in degranulation (release of the noxious substances). This explains why chocolate, opiates, red dyes, salicylates, mint, herbs, spices and some antibiotics have chemical properties that result in direct activation of mast cells. Indeed, one reason spicy foods, chocolate, etc., are liked by so many is they can directly activate specific mast cells in the soft palate that release histamine directly to cells, which then release endorphins into the brain, generating a feel-good high like an addiction. Some foods to which people are allergic can do the same, and these people may crave to ingest foods that can ultimately make them very ill.

In some people, pseudo-allergic reactions can occur. For instance, hot baths or cold baths can result in physical activation of mast cells, as can sitting in front of the full force of a cold air conditioner vent. Vibration from tools and machinery or riding a motorcycle can trigger a pseudo-allergic reaction. These reactions are not caused by an IgE-mediated response, but the ultimate clinical manifestations are the same as an IgE-mediated response since they result from the same noxious substances released from mast cells.

Foods are also causes of allergic disease that can be severe. The most common foods resulting in allergic disease include milk, egg, soy, wheat, fish, shellfish, peanuts and tree nuts. These allergies can begin early in life and for most are considered lifelong diseases. Essentially, any food can elicit an allergic response. Unfortunately, food allergies tend to cause the strongest reactions, with the highest risk of causing anaphylaxis, which is the chain reaction of triggering a large number of mast cells to release their mediators. This surge of irritants causes severe lowering of blood pressure and swelling of the airways, resulting in an inability to circulate blood and breathe. This is the worst-case scenario caused by IgE-mediated allergic disease.

In the next issue, we will continue with discussion of type II hypersensitivity. After completion of all four types of hypersensitivities, we will discuss how each can be involved with immune globulin therapies and what the treatments are for the reactions they may cause.
**Tips for Getting Along with Others During the Pandemic**

By Erika Lawrence, PhD

**THIS PANDEMIC** makes it especially frightening for those who are immuno-compromised given the increased health risks, which means being extremely careful about who you are around. So for the last several months, you have probably interacted in person almost exclusively with only one or two people. While this is a great opportunity to spend a lot of quality time with loved ones, it may also be causing more conflict, bickering and getting on each other’s nerves. However, there are some strategies you can use to try to manage arguments when they arise.

1) **Separate discussion of the problem from discussion of the solution.** When there is a disagreement or decision to be made and emotions run high, problems cannot be solved. Many of us want to jump to solutions when we have a conflict or decision to make. Unfortunately, this does not work unless you have already shared all of your thoughts and feelings about the problem with each other and until you both truly feel heard by the other. This means using effective communication skills to share your own thoughts and feelings, including:

   • Using “I” statements instead of “you” statements (e.g., “I feel frustrated” instead of “you are driving me crazy”).
   • Talking about your own thoughts and feelings instead of the other person’s behaviors or characteristics (e.g., “I have less patience these days” instead of “you are acting like a jerk”).
   • Keeping your speaking turns short (i.e., two to three sentences instead of long monologues or diatribes; you want the other person to be able to take in what you are saying).

   If you are listening during the problem discussion phase, it is important to truly understand what the other person is thinking and feeling and to make sure they feel heard. This does not mean you agree or forgive or give up. It just means you acknowledge their point of view. Only then should you move to the problem solution phase.

2) **Consider intent versus impact.** Another strategy to manage conflict and tension is to separate the other person’s intent from the impact of what they said or did. Intent refers to what they meant to communicate with their words or actions. Impact refers to the effect it had on you. There are two ways this distinction becomes helpful:

   • Consider the person’s intent: Often, if someone says or does something that hurts or angers us, we assume they intended to be hurtful or mean. That may be true, but it may not be true. Sometimes people say things that are poorly worded but not meant to be hurtful. If we can think about the intent of the person, that can help us reduce our anger toward them. That doesn’t mean their words or actions did not impact you. It just means that they did not intend to be hurtful.
   • Accept the impact you had on them: It’s easy to get defensive when someone says you hurt them or when someone is angry with you, particularly if you didn’t mean to be hurtful. If you separate intent from impact, you can realize you may not have meant to be hurtful and at the same time understand the person is still hurt.

3) **Separate the person’s “behavior” versus “who they are as a person.”** The main reason we get so defensive when someone tells us we have hurt or angered them is because we take it as a criticism of who we are as a person. Likewise, when we are hurt by or angry with someone, we tend to talk about who they are as a person instead of focusing on the specific words or actions that upset us. Thus, I offer two more strategies:

   • Talk about the behavior: If you attack or blame or criticize someone when you’re upset, they will become defensive. Instead, if someone upsets you, tell them by focusing specifically on their words or actions and on the specific situation.
   • Ask about the specifics: If someone is angry with you, try to focus on or get them to talk about the specific words or actions in a specific situation that upset them. If you can get them to focus on a specific interaction that led to their anger, you can respond to that and reduce the anger.

   This is a challenging, frustrating time for all of us. As a result, we are all more tense, impatient and short with each other. These tips should help you reduce tension and resolve these interactions more quickly and easily.

**ERIKA LAWRENCE, PhD**, is director of translational science at The Family Institute at Northwestern University, Evanston, Ill.
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RECURRING INFECTIONS are the hallmark symptom of people with primary immunodeficiency disease (PI), and frequently, those infections involve the sinuses. Yet, while acute or chronic sinusitis can occur in all people, PI patients often experience sinusitis before being definitively diagnosed. When sinusitis becomes chronic, it’s important for patients to undergo a full workup to determine the exact cause and type. Significantly, while chronic sinusitis can be successfully treated and managed in the presence or absence of an underlying PI, it can potentially be indicative of an undiagnosed underlying immune deficiency.

What Is Sinusitis?
The sinuses are small hollow cavities inside the front of the skull lined with a thin layer of skin, called mucosa. Their exact function is unknown, but when they become irritated and inflamed, it is called sinusitis. Another term for sinusitis is sinus infection because infection is a common cause of inflammation. Sinusitis is usually classified as acute or chronic, depending on how long it has been present, but it can actually be subdivided into four categories: acute, chronic, subacute and recurrent. The type of sinusitis depends on how long it lasts or how often it occurs.

Acute sinusitis is typically an infection in a sinus cavity that lasts no longer than 30 days. Sinusitis symptoms lasting longer than 90 days is chronic sinusitis. However, if sinusitis falls somewhere between acute and chronic, meaning it lasts longer than 30 days but not longer than 90 days, it is subacute sinusitis. Four or more bouts of acute sinusitis in one year is recurrent sinusitis.1

Symptoms of sinusitis are similar regardless of the type, and they can range from minor to severe. Due to an increase in mucus, nasal congestion and drainage can occur. Also pressure and pain in the face can cause a headache. Other symptoms may include loss of sense of smell, numbness in areas of the face and facial swelling. Sometimes there can be gastrointestinal (GI) disturbances if the mucus makes its way into the GI tract.

What Causes Sinusitis?
The cause of sinusitis can be hard to pin down since it isn’t always obvious. Sinus infections have several underlying causes, including polyps, a deviated septum and allergies that result in rhinitis and infection.2

Immune globulin G (IgG) plays a key role in the body’s immune response. IgG antibodies comprise the majority of all immune globulin cells and are vital to fighting foreign invaders, including fungus, virus and bacteria, that cause infection. Consequently, patients whose PI is caused by an antibody deficiency are at a higher risk for chronic fungal sinusitis. Such antibody deficiencies include common variable immune deficiency (CVID), specific antibody deficiency, IgG subclass deficiency and selective IgA deficiency.

Types of Fungal Sinusitis
There are four types of fungal sinusitis:3
• Saprophytic fungus. This occurs when fungus or mold grows on top of mucus or mucus crusts inside the nose. Treatment is simply removal of the crusts with nasal washes or other methods.
• Fungus ball. When fungus gets caught in one of the sinuses and forms clumps of material that often contain bacteria as well, it is called a fungus ball. Often no symptoms appear until the fungus ball grows large enough to block the sinus. Treatment is simple surgery
to open and wash out the sinus.

- **Allergic fungal sinusitis.** With this type, an allergic reaction occurs to one of several common fungi. Symptoms include those of an allergic response, including nasal congestion, runny nose and sneezing. The sinuses can enlarge and change the appearance of the eyes and face. Surgery is required, and without continued medical therapy afterward, recurrence is common. This is the most common type of chronic fungal sinusitis found in people without an underlying immune condition.

- **Invasive fungal sinusitis.** This severe infection of the nasal and sinus lining can lead to the destruction of nasal/sinus tissue. There are three forms: 1) chronic indolent/granulomatous sinusitis, which is very rare and usually not seen in the U.S.; 2) chronic invasive sinusitis, which is seen in people who have a poorly functioning immune system, as well as in diabetics, that very slowly invades the tissue of the sinuses; and 3) acute fulminant invasive fungal sinusitis, seen in people who do not have a functioning immune system (severe diabetics, transplant patients and those with lymphoma or leukemia are at highest risk), where the fungus invades and destroys blood vessels that line the nose, resulting in tissue death. This is a life-threatening disease and often requires emergency surgery and antifungal medications.

### Treating Sinusitis

Sinusitis is treated according to its cause and any underlying conditions. Treatment often involves surgery to remove the fungus and mucus containing the fungus. Antifungal medication may or may not be used.

Patients with certain types of PI are treated with immune globulin (IG) replacement therapy, either intravenously (IVIG) or subcutaneously (SCIG). CVID is managed with IVIG or SCIG. Selective IgA deficiency and IgG subclass deficiency may or may not be treated with IVIG or SCIG, especially at first, because depending on the symptoms, the physician and patient may opt to manage without it and monitor disease progression. The purpose of IG therapy is to create therapeutic levels of IgG to protect the patient from viral, bacterial and fungal infections that may be the cause of sinusitis and other infections. The goal is to decrease the severity and incidence of infection and improve quality of life.

### Ensuring the Best Outcome

Chronic sinusitis, whatever its cause, can be debilitating and frustrating. When chronic fungal sinusitis occurs in the presence of an antibody deficiency, it’s beneficial for the patient to see a specialist in this area who will manage and treat it in collaboration with the immunologist who is managing the immune deficiency. Without proper intervention, depending on the type, permanent damage to the sinus tracts can occur, and the condition can become more difficult to treat. Otolaryngologists (ear, nose and throat [ENT] doctors) specialize in the management of sinus conditions. An ENT physician can work with a PI patient’s immunologist to determine the best course for managing chronic fungal sinusitis to ensure the best outcome and resolution.

### Reference


Editor’s note: To find an ENT in a specific location, go to www.enthealth.org/find-ent, a patient-focused website powered by the American Academy of Otolaryngology-Head and Neck Surgery Foundation.

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MICHELLE GREER, RN, is senior vice president of sales for Nufactor, a Specialty Infusion Company.
CSL Behring Introduces Hizentra Prefilled Syringes for PI Patients

During the first week of July, CSL Behring debuted its new Hizentra (immune globulin subcutaneous [human] 20% liquid) prefilled syringes for primary immunodeficiency (PI) patients. The new ready-to-use prefilled syringes eliminate the need to transfer the drug from the vial to a syringe. The 5 mL and 10 mL syringes are fully assembled, but for the 20 mL syringe, the plunger rod needs to be screwed onto the syringe stopper prior to use. The syringes can be transferred to a pump using a syringe-to-syringe transfer device. A video is available to view the process of infusing with the new syringes at www.hizentra.com/hcp/flexible-infusions/pi.

IDF Creates Collaborative to Protect PI Patients During COVID-19 Pandemic

To ensure there are protections and responsible governmental policies for those with compromised immune systems both during and after the COVID-19 pandemic, the Immune Deficiency Foundation (IDF) has created the Protecting the Immunocompromised Collaborative and has begun working with peers to develop a steering committee. In advance of the collaborative effort, IDF has been advocating to ensure patients have access to home infusions and have joined with other patient organizations to support expanded telehealth services, increased access to health insurance, support for insurance and federal promotion of plasma donations.

To begin the collaborative efforts, IDF, the Lupus Foundation of America and the American Autoimmune Related Diseases Association drafted a letter to Congressional leaders urging them to task the Centers for Disease Control and Prevention to work with the immunocompromised stakeholder community to develop recommendations aimed at addressing the needs of those with compromised immune systems. This letter was signed by a diverse array of 40 stakeholders, including organizations that represent individuals with conditions such as arthritis, lupus, HIV/AIDS, multiple sclerosis, psoriasis, kidney disease and cancer. The letter expressed the importance of retaining flexibilities that enable the immunocompromised to receive home-based medical care because of the risk posed by receiving care outside of the home and the continuation of such flexibilities given the uncertainty that lies ahead. It also noted that guidelines must include more nuanced screening standards, particularly if such screens will determine a person’s ability to travel, work or attend school. In addition, it touched on the importance of educating the public about the safety and efficacy of vaccines since widespread or herd immunity is essential in protecting individuals with compromised immune systems, some who cannot be vaccinated themselves.
A study that assessed the effectiveness, safety and tolerability of transitioning from intravenous immune globulin (IVIG) therapy to subcutaneous immune globulin (SCIG) therapy in the treatment of myasthenia gravis (MG) patients found SCIG enables MG patients to maintain stable disease activity. The multicenter, open label and prospective study contained two parts: an IVIG screening phase (ISP) between 10 weeks and one week prior to therapy followed by the experimental treatment phase (ETP) between weeks 0 and 12. Researchers hypothesized that more than 80 percent of patients who started the ETP would have stable (less than a three-point increase) quantitative MG (QMG) scores at week 12, the study’s primary outcome. Secondary endpoints were differences from week 0 to week 12 in the MG Activities of Daily Living (MG-ADL) profile, the MG Quality of Life-15 (MG-QOL-15), the MG Composite (MGC) and the Treatment Satisfaction Questionnaire for Medication (TSQM). All but one of the 23 patients in the screening stage entered the experimental phase. Among those, 12 (54.5 percent) were women, 18 (78 percent) were Caucasian and the mean age was 51.4 years.

For the primary endpoint, 19 patients had complete QMG data during ETP. One of the remaining three withdrew from ISP due to worsened condition, and the two others quit before week four due to discomfort with the needle. The results of the primary statistical analysis showed stable QMG scores of “treatment success” in 19 of the 22 patients (86.4 percent), while a sensitivity analysis resulted in 17 participants (73.3 percent) with similar findings. A subsequent analysis of 20 patients confirmed treatment efficacy in 17 (85 percent) patients. Secondary measures MG-ALD, MG-QOL-15 and TSQM, comparing week 0 to week 12, showed no significant differences since the levels remained stable. Results from the MG Composite scale, however, showed a positive trend at week 12 for being at least stable if not slightly better. Further, investigators found SCIG was safe and well-tolerated with mostly local skin reactions.

Lopes, JM. *AANAM — Under-the-Skin Immunoglobulin Treatment Maintains Stable Disease Activity in Myasthenia Gravis Patients While Transitioning from IVIG Therapy*  

In a study that tested etanercept, a tumor necrosis factor α receptor antagonist for reducing intravenous immune globulin (IVIG) resistance and coronary artery disease progression in Kawasaki disease (KD) patients, researchers found no significant benefit for IVIG resistance, but they did find amelioration of coronary artery dilation, particularly in patients with baseline abnormalities.

In the double-blind multicenter trial, 201 KD patients received either 0.8 mg/kg of etanercept (100) or placebo (101) subcutaneously starting immediately after IVIG infusion. IVIG resistance was the primary outcome with prespecified subgroup analyses according to age, sex and race. Secondary outcomes included echocardiograph coronary artery measures within subgroups defined by coronary dilation (z score greater than 2.5) at baseline. Results showed IVIG resistance occurred in 22 percent of those receiving the placebo and 13 percent of those receiving etanercept. Etanercept reduced IVIG resistance in patients older than 1 year of age. In the entire population, etanercept reduced the coronary z score in 45 patients (23 percent) both with and without baseline dilation, but no improvement occurred in the placebo group.

Can Telemedicine Benefit PI Patients?

Telehealth has been embraced by most physicians and patients during the pandemic with the use of technology that has long been available, but issues will need to be addressed to ensure its continued use.

By Jim Trageser
ABOUT THE ONLY positive outcome of the novel coronavirus pandemic this year is it is providing a test of telemedicine on a massive scale. With numerous states ordering the cancellation of nonemergency in-person medical care in the early stages of the pandemic, tens of thousands of general practitioners and specialists suddenly found themselves interacting with their patients by email, telephone or live online video chat — or, oftentimes, a combination of all of these. Others were prescribing Internet-enabled remote monitoring devices for their patients to use at home to provide feedback that would normally be gathered during an office visit: weight, temperature, blood pressure, heart rate and blood sugar and oxygen levels.

For those with primary immunodeficiency diseases (PI), telemedicine offers some immediately apparent benefits: decreased exposure to potential infection, less time traveling to doctor visits and more immediate access to specialists.

While data this forced experiment in telehealth is generating is still being gathered, and will likely be analyzed for decades to come, some lessons learned are already making their way into the everyday practice of medicine.

What Is Telemedicine?

Telemedicine or telehealth is any method of medical treatment conducted remotely. This technology in some form has been around for decades. After all, people have been describing their symptoms to their doctors’ nurses for more than a century now to assist their physicians in determining whether they should come in for an office visit. (This has particularly been true for those living in remote areas where a trip to the doctor’s office may take a matter of hours in each direction.)

But some 30 years ago, we saw the first stirrings of what we might recognize as modern telehealth with the introduction of the home uterine monitor. Women with preterm labor could be sent home from the hospital with a modem-equipped personal computer modified to serve as a monitor for contractions, allowing the attending physician to change medication dosage remotely or order the patient back to the hospital.¹

At about the same time, the U.S. Army and U.S. Air Force medical departments combined efforts to develop and deploy a digital (or in the parlance of the time “filmless”) medical imaging solution. By 1998, that effort had been renamed the Telemedicine & Advanced Technology Research Center (TATRC) to help promote research into remote monitoring and provide critical support to the American Telemedicine Association, which helps bring much of that technology to the civilian world. The latest TATRC effort is the National Emergency Telecritical Care Network, which is charged with developing technology to provide specialists remotely in areas where medical infrastructure is overwhelmed by a pandemic or other emergency.²

Today, with the rise of the Internet of Things, there are all kinds of medical devices that can be remotely monitored by medical staff, from mechanical ventilators to blood pressure cuffs, insulin pumps and thermometers. In fact, there are so many new remotely accessible medical devices that it’s garnered its own acronym: IoMT (Internet of Medical Things). An informal survey found nearly every new home-monitoring device currently entering the market has some form of connectivity — either Bluetooth, Wi-Fi or direct Internet capability.

And that’s just for measuring. Once a doctor has that data, the next step is to reach out to the patient either by phone, voice over Internet protocol (VOIP) or the increasingly common online video chat with popular applications like Zoom or GoToMeeting.

A Changed Environment

According to Don Larsen, MD, MBA, MHA, FACHE, a physician and CEO at the University of California Riverside Health System in Southern California, the novel coronavirus
outbreak is providing the kind of financial stimulus that is allowing healthcare providers to bring their practices up to speed with technology that has been available for years but underutilized. “The technology has been advancing faster than the applicability,” he explained. “Probably in the last four, five years, maybe even longer, there have been a plethora of medical device companies that have seen the benefit of taking care of people remotely. It could be people like PI patients who could get sick if they leave the house. It could be people who live in remote areas, or a way to get specialists to places where there are no specialists. What’s been holding it back all this time has been a way to pay for it. Nothing is free. The payers have resisted for a long time in paying for this — especially Medicare.”

But, when the pandemic hit, suddenly private and public insurers were more than willing to reimburse physicians for remote visits via telephone, email or online video conference, Dr. Larsen said, and so far at least, at the same rate as in-person office visits.

Early Adopters

With many PI patients incorporating mental health treatment into their ongoing care plan to help deal with issues of stress and anxiety, the good news is mental health therapists have been among the practitioners most rapidly embracing telemedicine — although not without some trepidation. We worked out some of the bugs. When the pandemic hit, it came fast. [So,] psychiatry said, “We’ve been doing this, and it works, and we can use it in the rest of our department.”

Dr. Larsen pointed out that fear of exposure to the coronavirus led many of his doctors to prefer to work from home, which led to some fast-moving plans to ensure a confidential environment at his psychiatrists’ homes. “Look at it from a privacy [standpoint]. If you’re doing consult in telehealth from your home, who’s controlling your environment? How can we as a healthcare system give confidence to a patient that we’re handling this in a confidential manner?”

UCR Health sent its privacy officers and HIPAA experts to each doctor’s home to make sure the environment was compliant. “What we’ve been doing in psychiatry is that about 90 percent of our visits are remote for both the doctor and the patient. There is still some need to bring patients in face to face for an exam or a test or IV drugs. Even when they’re home, we have our medical assistant facilitating with the technology and the connection.”

Now, said Dr. Larson, “We’ve expanded into primary care, family medicine and women’s health — all of our other clinical specialties.” Still, there was resistance, he said. “Some physicians feared it would destroy our connection to our patients — that they won’t develop this closeness that they’re used to. It’s still a concern; we just have to see how it falls.”

Long-Term Benefits

According to Dr. Larsen, for patients with chronic illnesses who rely on specialists as part of their ongoing regimen, telehealth offers several distinct benefits.
One important advantage is access to a specialist without having to travel. “You can use fewer specialists; you don’t need to have a specialist at every site,” Dr. Larsen explained. He added that in smaller markets, a specialist will often be underutilized with long stretches of down time, while patients in even more far-flung communities will have no access to that same specialist at all.

He offered the example of neurologists treating strokes to demonstrate how telemedicine can extend the reach of a specialist to many more patients: “We have drugs and tools to treat stroke, but time is of the essence. We need to have a neurologist in the emergency room when [the patient] arrives because the clock is ticking. There aren’t enough neurologists to go around. You have to use tPA (tissue plasminogen activator) in the first eight hours of a stroke event. If you’re a small community hospital, you might have the drug but not a neurologist. If they give [the patient] the blood-clotting drug and [they have] a bleed, it will make it worse. Now you can have a neurologist covering 20 hospitals or more.”

Immunologists could be similarly redeployed using telehealth technology, he said. But the primary benefit is in more responsive care, catching problems at home before they progress. For example, he pointed out how his health team is treating patients with congestive heart failure. “[When] fluid builds up in the lungs, [patients] need to take water pills. To monitor that, [they] need to go to the doctor frequently,” he explained. Now, though, the doctors can have the patient acquire a Bluetooth-equipped scale, which can send the readings to the doctor’s office. Software in the office will note any sudden changes in weight and flag the staff who will then reach out to the patient. “If we can monitor them remotely, we can head it off before it gets worse,” Dr. Larsen said. “The ER is the least efficient way to treat disease.”

**Ongoing Challenges**

In addition to resistance from insurers, Dr. Larsen cited government regulation as another barrier to expanded telehealth — even in the current pandemic. For instance, an immunologist who is available via telehealth generally can’t practice medicine across state lines without being licensed in each state, which is a time-consuming and expensive proposition. But, there is now an Interstate Medical Licensure Compact among 27 states, mostly in the Great Plains and Deep South, that offers expedited processes for cross-licensing. However, California and most of the Eastern Seaboard are not yet on board, leaving a patchwork process in the United States. And, Dr. Larsen said, that doesn’t even take into account international licensure requirements.

A recent paper on patient-centric care for PI patients, which pointed out geographic accessibility to medical specialists remains one of the most significant challenges to improving PI care in developing nations, looked to telemedicine to provide a solution. That same paper pointed out a PI patient may, over the course of diagnosis and subsequent treatment, require access to specialists not only in immunology, but also hematology, infectious disease, rheumatology, dermatology, pulmonology, gastrointestinal medicine and neurology.

Other challenges come with using telehealth to treat the elderly or the very young, where unfamiliarity with the technology can be a barrier. And, with special needs patients in whom there may be a cognitive disability, this obviously presents other hurdles. But, Dr. Larsen said with young children and special needs patients, the process is similar to an in-person office visit during which the nurse or doctor communicates via the parent. With older patients, he said, the doctors at UCR Health System are seeing less resistance than anticipated, possibly because the pandemic is forcing people of all ages to become more familiar with technology to stay in touch with family and friends. “Getting familiar with the technology was a barrier in the past,” he explained. “It’s not too different than the technology that seniors use to stay in touch with their grandkids during the pandemic. Or, they know how to use their mobile device better than before.”

Of course, a bigger problem with elderly patients right now during stay-at-home orders is social isolation and depression, but psychologists and psychiatrists are able to use remote video sessions to assist elderly patients in dealing with these stressors.
Hizentra®
Immune Globulin Subcutaneous (Human) 20% Liquid

My Life, My Way
With Hizentra

Hizentra is an Ig* therapy that provides proven PI protection with the convenience of self-administration, so you can focus on everyday living

*Ig=immunoglobulin

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.

Please see Brief Summary of full Prescribing Information on reverse.
Simplify your infusions
with the first and only Ig prefilled syringes
— only from Hizentra

Choose when and where you infuse
Self-administration with Hizentra means you and your doctor can decide when and where you infuse. Convenient dosing options mean you won’t have to adjust or cancel your plans due to IV infusion appointments.

No more IV infusions
IV Infusions can be challenging for people who have hard-to-find or damaged veins. Hizentra allows you to infuse just under the skin, not into a vein, after training from your doctor.

Proven Safety
Hizentra has an established safety profile and demonstrated tolerability. In clinical trials, the most common side effects were 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.

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 full prescribing information for Hizentra, including boxed warning and patient product information, available at 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.

Visit Hizentra.com or ask your doctor about Hizentra prefilled syringes.
HIZENTRA®, Immune Globulin Subcutaneous (Human), 20% Liquid
Initial U.S. Approval: 2010

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

WARNING: THROMBOSIS
See full prescribing information for complete boxed warning.

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

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

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

For subcutaneous infusion only.

DOSAGE FORMS AND STRENGTHS
0.2 g per mL (20%) protein solution for subcutaneous infusion available in a single-use prefilled syringe (5 mL, 10 mL, and 20 mL) or tamper-evident vial (5, 10, 20 and 50 mL).

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 2020 revision

Hizentra is manufactured by CSL Behring AG and distributed by CSL Behring LLC. Hizentra® is a registered trademark of CSL Behring AG. Biotherapies for Life® and IgIQ® are registered trademarks of CSL Behring LLC. Premier Start™ and CSL Behring Assurance™ are service marks of CSL Behring LLC.
Another issue providers need to consider, Dr. Larsen said, is not every patient is equally capable of taking advantage of telehealth. “There are still a lot of people who don’t have Internet access. We have to be careful not to disenfranchise them. With all this enthusiasm about telehealth, some people don’t even have an Internet-enabled phone. This is a major concern. Telehealth is not a panacea.”

Looking Ahead

While most consultations can be handled virtually for now, some treatments will require in-person visits for the foreseeable future. Drawing blood for tests is a regular occurrence for many PI patients and, as of yet, this is beyond the scope of telemedicine. Theoretically, however, it’s not impossible. Learning to do a blood draw is not particularly difficult, but storing and shipping the samples in a climate-controlled environment and ensuring that chain-of-custody and privacy regulations are met mean blood draws will remain a clinical occupation for some time.

Thus far, other technical issues are falling to the side as engineers continue to solve previously vexing issues. Dr. Larsen pointed out that on the horizon is a huge advance in dermatology, where tremendous leaps in digital imaging now mean a skin condition can be photographed at home with a cell phone camera and sent to a dermatologist to review. Beyond that, though, he said he’s seen research into using artificial intelligence to help read images and prioritize them for the physician — a sort of digital triage.

And while insurance companies and Medicare have been willing to pay full freight for telehealth during the pandemic, there is no guarantee that will continue, Dr. Larsen said. “The payers are concerned about how you measure overhead. I think they’ve always feared telehealth was a shortcut to make money — that we’ll charge the same amount, but not deliver the same service. What helped was the insurance payers covered it and are still covering it at the same rate as a face-to-face [appointment]. They’re paying the doctors the same amount. [But,] that doesn’t mean they’ll pay us forever, or pay us the same amount. They may argue our overhead is lower, and they may want to pay less.”

For Dr. Larsen, though, what has stood out above all in his role supervising a large, sprawling public health district during a pandemic is the speed and openness patients have shown in adapting to telehealth. “What was surprising to us is that patients really like it! Our no-show rate went really far down, and our patients who showed up late went down. What it told us is that access is a big issue. More people kept their appointments when they could do it from home. You don’t have to take time off from work or get a babysitter for a remote appointment. I think this was an eye-opener for patients, too. It forced them to try it. I think people are surprised at how intimate these Zoom calls can be. In some respects, you’re pretty funneled and focused.”

A recent paper on patient-centric care for PI patients, which pointed out geographic accessibility to medical specialists remains one of the most significant challenges to improving PI care in developing nations, looked to telemedicine to provide a solution.

For any patients who are still reluctant or apprehensive about trying telemedicine, Larsen offered this advice: “Give it a try! There’s very little risk involved in it. For us, if it’s not working, you can always come in.”

References

JIM TRAGESER is a freelance journalist in the San Diego area.
New Tools and Tech for Patients

This second-in-a-series of product highlights can help patients deal with many of the discomforts that often accompany chronic illness.

By Meredith Whitmore

LET’S FACE IT. So far, 2020 has not met our previously hopeful expectations. Not even close. There has been a lot of discomfort for many of us, whether the distress has been physical, emotional or both. But take heart, there are definitely ways to feel better!

We have found some of the coolest, most helpful, state-of-the-art personal health technologies. Whether you want to feel more comfortable, improve your mindset or monitor your health, there’s definitely a way to do it in the comfort and privacy of your own home.

Personal Comfort and Pain Relief

- **TechCare Massager Touch 24 Modes, $44.95**
  (techcaremassager.com)

  If you have arthritis, sciatica or virtually anything else that causes pain, this transcutaneous electrical nerve stimulator (TENS) unit could become your new best friend — no matter where you are since it’s easily portable. Delivering low-voltage electronic pulses through the skin, the device stimulates muscles and nerves, blocking the pain signals to your brain while increasing endorphins, a natural pain reliever. Sporting a rechargeable lithium battery and a touch LCD display, the unit also includes 24 preprogrammed massage modes, including acupuncture, deep tissue, cupping and combo. And an A-B out means you can have two different massages at the same time!
• **Fancii Cool Mist Personal Travel Humidifier**, $24.99 (amazon.com)

On the road in a dry climate or just wanting an affordable, toteable boost for your dry skin or nose? This gadget allows you to (almost) magically transform a water bottle into a personal humidifier, providing relief from dryness wherever you are. Breathe easier using this quiet, compact humidifier with wireless and USB features. Requires three AA batteries.

• **ClearUP Sinus Pain Relief**, $149 (tivichealth.com)

Do you remember the last sinus headache you had? Not a pleasant memory, certainly. Now there’s an easy bioelectronic way to relieve the pressure, pain and overall awfulness in five minutes without chemicals and in a noninvasive manner. Use ClearUP on your cheek, nose and brow bone, and its gentle vibrations guide you. A five-minute treatment can last as long as six hours! For those who suffer regular sinus pain, the recommended usage is at least twice a day for two weeks to promote happier breathing and less discomfort.

• **The Wave Bracelet**, $299 with payment plans available (www.embrlabs.com)

This personal temperature regulation device allows you to cool down or warm up at the touch of a button — and without changing your core temperature. Developed by a team of MIT grads, the device uses a physiological “hack” to activate a body/mind response that changes how you perceive the temperature of your environment. Now you can find comfort with this watch-like tool even when everyone else in the room is too cold or too hot. Choose between several different modes for comfort in any environment. Connect to the Wave Mobile app to personalize your experience and receive continuous improvements.

• **Sleep Quality**
  
  • **Withings Sleep Mat**, $99.95 (withings.com)

Tossing and turning this year? Join the club. Maybe understanding what makes a good night’s sleep would help you to get one. The Withings Sleep Mat offers a sleep score and indicates how breathing disturbances, heart rate, REM and snoring can affect your slumber or even indicate health problems such as sleep apnea. The pad goes under a mattress to ensure it allows comfortable rest without distractions. Wake to view the night’s data in the accompanying wireless app. Compatible with iOS and Android, and there’s even personalized coaching available.

• **Noisli**, a free version and several different price plans, $1.99 or $10 for personal and $24 for team memberships (www.noisli.com)

Having trouble winding down when you go to bed? There’s an app for that, and it’s simple! Just choose your favorite sounds — thunder, white noise, coffee shop chatter, wind and more — to line up a sleep playlist that will calm you and soothe your cares. Noisli’s hypnotic, relaxing and gentle tones are guaranteed to bring greater peace.

• **In-Home Health Monitoring**
  
  • TytoHome, $300 (www.tytocare.com)

This remote exam kit, the first of its kind, allows you to conduct a basic medical exam at home (and quarantine has certainly proven the importance of having this ability). Diagnosis, treatment planning and prescriptions are all possible with this groundbreaking gadget, which comes equipped with a digital camera and thermometer. It also
includes accessories such as a tongue depressor, an otoscope for ears and a stethoscope for heart, lungs and abdomen. The device then pairs with a teleconferencing app that connects with a healthcare provider for consultation. The possible catch? Healthcare providers must have the ability to teleconference using a TytoCare device designed for professionals. You will need to download the TytoCare app (Android or iOS), log in and create a patient profile to get started. The device charges via a 5-volt 2A port rather than micro USB.

- **KardiaMobile EKG Monitor, $89; or KardiaMobile 6L Portable 6-Lead, $149 (alivecor.com)**

  The monitor is a U.S. Food and Drug Administration-approved mobile electrocardiogram (EKG) that allows you to track your heart health at your convenience anywhere. A medical-grade EKG is sent to your smart device in just 30 seconds. It won’t break the bank or your heart, and it’s compatible with iPhone, iPad and Android devices. The KardiaMobile 6L is a step up that allows you to glean more health information to give your doctor an even better view of your heart, with the same power of a six-lead EKG. Both devices are HSA-eligible.

- **Withings Move ECG ScanWatch, $129.95 (www.withings.com)**

  For those who want to wear their EKG equipment without the obvious look of a medical device, there is this clinically validated watch. At the first sign of atrial fibrillation or other heartbeat irregularity, just press a button and place your finger on the bezel. Within 30 seconds, a medical-grade EKG is ready to view and share with your health professional. The accompanying Health Mate app offers an instant analysis of your heart rhythm and tells you whether it looks normal or shows signs of atrial fibrillation. If signs are detected, the app will recommend contacting a doctor. Data can be stored in the watch until the next sync.

- **Omron HeartGuide Watch, $499 (www.omronhealthcare.com)**

  The Omron HeartGuide is a smartwatch that never misses a beat. It not only tracks blood pressure, it monitors sleep quality and activity, allowing you to track health trends over time and compare your readings to current American Heart Association/American College of Cardiology guidelines. Finally, you can understand how your lifestyle impacts your heart, and with the HeartAdvisor app, you can have a well-informed dialogue with your doctor. You can also access real-time heart health coaching, as well as your health history to gain insight into your daily readings. The watch uses the same type of technology as an inflatable oscillometric blood pressure cuff. This ensures the device’s readings are accurate enough to provide data that your doctor can use to adjust your medications and make more precise recommendation regarding your health. Users can also receive smart notifications on the device such as texts, calls and emails. The watch reminds you to take your reading, allowing you to sit beforehand for even greater accuracy.

### Emotional and Stress Management

- **Light Therapy Lamp 19, 10000 Lux LED Light Source, $36.99 (www.taotronics.com)**

  Winter is coming, and those gray doldrums can be hard on the emotions even during a brighter year. But you can have a happier disposition any time of day with this sleek, tablet-sized, easy-to-use lamp that mimics sunlight (without the UV damage). It’s clinically effective to fight seasonal affective disorder, jet lag and shift-work gogginess, among other fatigue-inducing situations. It also has a timer, dimmer and rotatable stand, so it’s sure to please even the most ardent sun lovers who need to stay indoors.

- **Headspace, $69.99 per year subscription (headspace.com, Google Play and App Store)**

  Whether you are stressed and seeking mental relief or you would like to begin meditating just out
of curiosity, Headspace is a guided tour for beginners. There’s also a lot available for more seasoned meditators! Tutorials and classes are progressive and include training in such things as how to sit quietly, calm an active mind and accept reality without judgment. There’s even a free trial period so you can warm up to the app and decide if it’s for you.

- **EMOTIV Insight 5 Channel Mobile Brainwear**, $299 (www.emotiv.com/insight)

  This former Kickstarter project grew up into a sleek, futuristic-looking headset that monitors brain activity and translates EEG into meaningful data that you can use to manage stress. It can also increase productivity by spurring motivation, improve your mental focus and help you to relax. If you’d like to learn how to maximize your cognitive abilities, this rechargeable Android- and iOS-compatible device is a great way to start.

- **Bond Touch Bracelets**, $98 per pair (www.bond-touch.com)

  Separated from a friend or loved one and missing their presence during these stressful times? There’s an immediate way to remind yourselves that you both still care. The Bond Touch Bracelet allows friends, family members and significant others to physically interact even from the other side of the world. When one person touches the bracelet, the other feels the gesture. Available in three colors with a USB charger and user manual.

- **Muse 2 and Muse S**, $249.99 to $349.99 (chosemuse.com)

  If you’d like to dive in even deeper into contemplation than with phone apps such as Headspace, wearable tech can help you meditate in ways you’ve never imagined. These multi-sensor devices provide real-time feedback on brain activity, breathing, heart rate and movement, helping you to build and maintain a consistent meditative state. Not only can the devices help you focus like a Zen master, these comfy Muse products can improve your meditative posture, helping you to relax and breathe optimally with award-winning biofeedback technology. Micro USB port/cable and LED indicator are included.

**Personal Safety**

- **Verilux CleanWave Portable Sanitizing Wand**, $100 (verilux.com)

  Easy to use and able to kill up to 99.9 percent of all viruses, bacteria and mold found on hard surfaces, the Verilux CleanWave Wand uses a UV-C to keep you safe from MRSA, H1N1 and E-coli, among other pathogens. All you have to do is turn it on and hold it for 30 seconds to 60 seconds over the surface area you’d like to clean. The 10-inch wand is easily carried in a purse or briefcase and its chemical-free approach means less toxicity. Great for travel, parents, caregivers and anyone else who cares about their and others’ health. Quick and effective for peace of mind in our germ-laden world.

- **WELT Smart Belt Pro**, price starts at $395 (www.weltcorp.com)

  Winner of the 2020 CES Innovation Award, the WELT Smart Belt Pro works to protect its wearer from falling. Though such accidents aren’t dangerous for the average person, for the more vulnerable, a fall can mean devastating complications. The WELT Smart Belt Pro protects the wearer (and eases the fears of any caregivers) by monitoring walking patterns and detecting the likelihood of falls before they occur. To accomplish this, it uses the signals provided by a belt sensor positioned at the center of the body to detect patterns in small steps. These steps predict fall risk by providing gait analysis. If risk is detected, a warning is sent to the user’s smartphone. This device provides not only security but greater confidence.

**MEREDITH WHITMORE** is an English professor and freelance journalist in the Northwest.
BIOFEEDBACK for Pain, Stress and Anxiety

This technique uses external stimuli to help influence how the body responds to stressors.

By Amy Scanlin, MS
“I WAS WORRIED about my health, what would happen to me and what if the doctor found this or that,” said Angie Snow. “I was worried about things that weren’t even things yet, and it was taking a big toll on my health, my sleep and my ability to concentrate.” So Angie tried biofeedback to lessen her anxiety, and she found the biggest impediment to her success was believing in her ability to succeed and in consistently practicing the technique.

Biofeedback is a mind-body, noninvasive and low-risk technique to manage the body’s autonomic (automatic) responses to internal or external triggers. Autonomic responses, which include heart rate, breathing rate, respiration, temperature and even brain waves, can be controlled with practice to lead to a healthier response and better management of chronic health challenges. An example of an autonomic response is the fight-or-flight response (also called hyperarousal or the acute stress response), which causes panic, a racing heart, a spike in blood pressure, an adrenaline rush and tense muscles. While the fight-or-flight response is automatic, over time it can also become a learned response. And unless a new response is learned, people are likely to react in the same way again. In cases in which autonomic reactions are harmful, individuals can begin to develop health concerns.

By becoming aware of one’s reactions and utilizing methods to control them, individuals can unlearn habits and replace them with breath control and healthier physical and emotional responses. Whether termed mindfulness, awareness, positivity or biofeedback, the ability to recognize stressors and influence the reaction to them can lead to more productive responses and better control of health.

Though not yet studied on a large scale, there is scientific and anecdotal evidence of biofeedback’s effectiveness. Conditions that can benefit from biofeedback include Raynaud’s syndrome, tension headaches, fibromyalgia, chronic pain, hypertension, cardiac arrhythmias and sleep disorders. Through practice, biofeedback can help individuals feel ownership of and empowerment over their conditions, as well as change their mindset from powerless to empowered by understanding their own biology and their ability to influence their symptoms. In short, biofeedback improves self-efficacy.¹

What Is Biofeedback?

Biofeedback can mean many things, can be practiced anywhere and refers to the use of instruments to measure and provide real-time feedback on the body’s responses to triggers. Clinically, biofeedback practitioners use a variety of tools such as an electrocardiogram to check heart rhythm, electrodermograph to measure sweat gland activity, electromyography to assess muscle tension and electroencephalograph to measure brain waves, as well as assessments of breathing rates and temperature (increased stress can lead to lower body temperatures).

Feedback from these devices is provided to participants through beeps, flashing lights or readings on a monitor screen, which indicate arousal states and prompt a purposeful effort to change emotions or reactions. The idea behind biofeedback is while individuals may not readily notice their physiological reactions to stress, they can learn to recognize them and develop techniques to actively control their sympathetic arousal states. Through biofeedback, individuals have the ability to improve their emotional and physical health.

But learning biofeedback requires sessions with a practitioner and practice in between sessions to master a lifelong practice of voluntary control over what were previously thought to be involuntary reactions. Biofeedback is a training, not a treatment, with much emphasis placed on education.² It can be used to complement traditional medicine or it can be used on its own.

How Does Biofeedback Work?

Autonomic disorders can affect any part of the body and its processes, including blood vessels, intestines, heart, digestion, bladder, sweat glands and more. These disorders cause dizziness, reduced sweating, constipation and a variety of other ailments. Common causes of autonomic disorders
are diabetes, peripheral nerve disorders, disorders of the muscle-nerve connections and aging (to name a few). And while autonomic disorders can be progressive, they can also be reversible.

Autonomic disorders can affect any part of the body and its processes, including blood vessels, intestines, heart, digestion, bladder, sweat glands and more.

During the first biofeedback session, individuals and their practitioners will discuss concerns, health history, current medications and expectations. They’ll then be connected to the biofeedback equipment so sensors can detect responses. This equipment may include a temperature sensor on the fingertip and a band around the waist to measure breathing. Prompted by the feedback via sounds or indicators on a screen, individuals can learn to begin to quiet their body’s autonomic response to stressors by slowing their breath, relaxing their muscles and attaining a feeling of ownership over their ability to cope.

Biofeedback alerts individuals to their bodies’ signals so they can learn to recognize how stress and anxiety feels, enabling them to adjust their responses and lessen the negative effects. The cues provided by biofeedback are similar to the visual cues one can see when exercising in front of a mirror such as body position and correction. And although it takes practice, the ultimate goal is to be able to successfully use these relaxation techniques on one’s own.

For some, medical conditions such as abnormal heart rhythms or temperature fluctuations may lessen biofeedback’s effectiveness or render it ineffective. In those cases, a doctor can make an assessment.

For many, the addition of wearables offers good motivation to assess and make positive changes. There are a plethora of wearable devices that feed information from the body through a device or computer application. From cognitive feedback, sleep monitoring, muscle stimulation and more, if it can be measured, research is underway to capture and utilize that data. By some estimates, the wearable market is expected to generate $48.2 billion in revenue by 2023. However, it is important to note that self-diagnosis based on self-gathered data should be verified with a medical professional. Wearables should never replace medical care.

How is Biofeedback Different from Mindfulness?

Mindfulness is a precursor to understanding how emotions impact the body, facilitating a response similar to biofeedback. By focusing on the moment, staying present and accepting one’s circumstances, individuals can begin to understand how stressors affect them. Mindfulness helps individuals learn to refocus attention when it is recognized the body’s emotions are not productive. It allows them to recognize internal sensations produced by their reactions and become aware of their breath and begin to slow their nervous system. The greater one’s ability to be mindful, the greater one’s ability to handle stress.

While it is hard to separate the effort of mindfulness from the hope that its practice will produce an improved physical response, the very definition of mindfulness is acceptance of the way things are. Instead of attaching a hoped-for future outcome to mindfulness, a better approach is being mindful as those improvements happen. That being said, mindfulness has been shown in multiple studies to be effective in improving immune response and psychological coping.

Mindfulness is very much aligned with biofeedback. Awareness is the first step in biofeedback, which is used to learn to bring
voluntary control over involuntary reactions. The difference, however, is with biofeedback, external stimuli prompts individuals to make changes, whereas with mindfulness, it is internal awareness.4

Finding a Practitioner

Referrals from individuals’ healthcare teams are a good source of finding an appropriate biofeedback practitioner. Additionally, the Association for Applied Psychophysiology and Biofeedback and the Biofeedback Certification International Alliance offer a certification and listing of approved practitioners. And, while a certification or license to practice is generally not a state requirement for practitioners, many may be licensed in another area of care such as psychology or physical therapy.

Individuals considering biofeedback should choose a reputable practitioner by checking credentials, experience and references. In addition, the practitioner should have experience treating the conditions a person has, and it should be clear how many sessions may be needed to master the technique. Individuals may also want to ask about costs and insurance coverage (biofeedback is often not covered by insurance).

And, as with any complementary therapy, individuals should follow the advice of their healthcare providers and keep communication lines open, including discussions about biofeedback efforts with the entire healthcare team.

A Healthy Outlet Is Needed

Whether individuals choose to try biofeedback or to intentionally become more mindful, finding a healthy outlet for stress reduction and management can lead to improved health outcomes.4

References


AMY SCANLIN, MS, is a freelance writer and editor specializing in medical and fitness topics.
Adaptive Mobility Solutions

Designed to help stability during movement, the devices come in many forms, and there is often assistance for obtaining them.

By Matthew D. Hansen, DPT, MPT, BSPTS

According to the U.S. Census Bureau, more than 20 million people in the U.S. (approximately 7 percent) have difficulty with ambulation, with twice as many struggling with some more comprehensive forms of physical functioning and activities of daily living. Fortunately, a number of adaptive mobility solutions are available to those who need them. However, many of those who need them don’t use them — or at least they don’t use them properly, which can result in decreased mobility and oftentimes unnecessary falls and injury. If an adaptive mobility device has been recommended by a healthcare professional, or if individuals have to ask themselves if one is necessary, then a device is likely needed (at least for partial use or specific circumstances).

I’m a firm believer in maintaining function and doing all one can do without help, as long as it can be done safely. Yet, the first fall for most people is often preceded by at least one conversation with a healthcare provider and/or loved one about adaptive mobility. And, if they’re honest with themselves, the thought had at least crossed their mind. Regrettably, equipment acquisition can sometimes be delayed due to the potentially difficult process of submitting an insurance claim and gaining approval; oftentimes, the process hasn’t even been initiated prior to a first fall due to concerns about cost and/or the fear of being seen as disabled.

Many people who know me, have read enough of my articles or have viewed my presentations recognize one of my personal mantras is: “Keep on moving!” The phrase doesn’t come with a qualifier.
It doesn’t say: “Keep on moving if it’s easy” or “if you don’t have to depend on some help” or “if you can do it while looking like everyone else.” The first step to successfully using adaptive mobility options is to ignore the stigma. People who are potential candidates for adaptive mobility devices are so because they have underlying conditions requiring them. They’re already different. We’re all different. Adaptive mobility is not a forfeiture of independence unless we let it be. Rather, it’s a way to preserve independence!

Adaptive Mobility Options

In a broad sense, adaptive mobility options are assistive devices used to help improve stability during movement. Assistive devices include canes with varying points of stability (e.g., single-point, tripod and quad), walkers with and without wheels and crutches. People will likely have had experience with at least one of these assistive devices before needing a wheelchair or power scooter. Still, because most of these devices can be relatively easy and inexpensive to obtain, even from the local drug store, the term “adaptive mobility” is more traditionally associated with manual and power wheelchairs and power scooters.

For those living with a primary immunodeficiency, access to a manual wheelchair for home use while recovering from an illness or following an infusion may be sufficient to help preserve strength and functional mobility while preventing injury. However, a power wheelchair or scooter may be required for greater mobility in the community (for instance, if someone becomes easily exhausted while using a manual wheelchair or if a progressive autoimmune disease affects the neuromuscular system).

While there are many benefits to wheelchairs and power scooters, there are also some obvious challenges. For example: Is there enough room in the home or apartment to allow safe and effective mobility? Are doorways and hallways wide enough? Does furniture get in the way of turning in the bedroom or living spaces? Are there steps leading into the home or inside the home that need to be negotiated (split level homes can be a nightmare)? Is there a vehicle that can transport the wheelchair/scooter? If the answer to any of these questions is “no,” there are solutions, but they may require some sacrifice in the form of home modifications and time spent conducting research, and they almost always require funding.

Attaining Adaptive Mobility

Manual and power wheelchairs are classified into five different types, with varying degrees of support and features. If a manual wheelchair is needed solely for use around the home, or if there is someone who can push the wheelchair, a standard foldable wheelchair or a “transport wheelchair” from a local thrift shop or the classified ads is all that is really needed. These are the stereotypical wheelchairs seen in hospitals and medical clinics used to transport patients to and from locations inside the facility or to their car to return home. Some insurance plans, including Medicare, cover these chairs, while other insurance carriers don’t. If one must be purchased out of pocket, the cost for one online is approximately $200 to $300.

The downside of standard wheelchairs is they can be clunky, and they aren’t built to withstand the wear and tear caused by outdoor use. They’re simply used to get someone from point A to point B. If more time in a wheelchair is required, better support will be needed than the sling-style seating provided by standard wheelchairs.

Private insurance benefits for adaptive mobility devices vary...
widely, as do state-by-state Medicaid benefits. Individuals 65 years and older or those who qualify with a disability and are approved for Social Security Disability Insurance can receive Medicare benefits. Medicare Part B covers both manual and power wheelchairs and scooters under the durable medical equipment (DME) benefit as long as several requirements can be met:

- First, a physician must conduct a face-to-face assessment of the person and submit a written order stating he or she demonstrates a medical necessity for the scooter or wheelchair. A medical necessity includes a health condition that causes significant difficulty moving around in the home and an impaired ability to perform activities of daily living (e.g., bathing, dressing, getting in or out of a chair or bed, or using the bathroom) even with the aid of an assistive device.
- Additionally, the person requesting a wheelchair or scooter must be able to independently and safely operate the equipment within the home and transfer into and out of it, or have someone who is always available to help him or her use the device.
- If these conditions are met and a physician’s order has been attained, a request to obtain a chair or scooter can be made to a Medicare-contracted DME supplier. The DME supplier will often have to request prior authorization for the chair or scooter from Medicare before it can be secured.

As part of the pre-authorization process, it’s not uncommon for Medicare to require a documented wheelchair evaluation. Some private insurance plans may ask for a letter of justification (letter of medical necessity) instead. Even if a wheelchair evaluation is not required, I strongly recommend one. In collaboration with the patient, DME provider and physician, a physical or occupational therapist can perform an evaluation and help recommend what equipment and accessories are needed, and then present supportive documentation in the proper format. Just as important as the chair or scooter is the seating configuration; the wrong chair seat or back can soon lead to all kinds of other problems, even in a new wheelchair. It’s a process, but the professionals who perform evaluations are very familiar with the requirements, and they attempt to make everything as easy as possible for recipients.

Again, private insurance companies set their own rules, but if a wheelchair or scooter is approved under Medicare B coverage, the recipient must pay 20 percent of the Medicare-approved amount after paying the deductible for the year. Medicare pays the remaining 80 percent. Those with a Medicare Advantage plan instead of traditional Medicare should contact the plan provider to ask about costs and which DME suppliers can be used. However, Medicare Advantage plans must cover the same services as traditional Medicare benefits.

Medicare and other government insurance plans typically only pay for manual or power mobility equipment once every five years. This is what they consider the minimal reasonable useful lifetime, so it’s important to select a chair or scooter that meets a person’s current and anticipated needs. New mobility equipment may be acquired sooner than every five years if there is a change in medical condition that requires a different configuration or if the chair falls into disrepair under extenuating circumstances. It’s also typical for insurance companies to cover an annual wheelchair evaluation and to pay for replacement parts that break or become worn out before they will consider paying for another new chair.

Other Associated Needs

Acquiring a wheelchair or power scooter may be just the beginning of what is needed to maintain mobility at home and in the community. Based on individual circumstances, additional requirements may include wheelchair ramps, vertical platform lifts, stair lifts, patient transfer lifts and/or adaptive vans.

If it can be demonstrated to be medically necessary, Medicare will cover the cost of a patient transfer lift or wheelchair ramp. However, traditional Medicare does not pay for residential wheelchair lifts or adaptive vans, although some Medicare Advantage or private insurance plans may help to cover the cost of a home wheelchair lift or stair lift. These items, which can range in cost from several thousand dollars to tens of thousands of dollars, can be prohibitively costly as an out-of-pocket expense. Fortunately, there may be assistance available from state and private grants or other sources, including nonprofit foundations.
An Internet search will produce a broad list of grants and programs for adaptive mobility equipment. However, these may be difficult to qualify for, and potential candidates must be careful not to provide personal information or money to potentially dubious sources, regardless of what promises might be made. It’s certainly still worth conducting the research and taking a look at what is available. But, the safest first step may be to contact one of the reputable foundations specializing in help for those with an immunodeficiency or autoimmune disease to ask what programs they recommend.

For those without a personal/family vehicle that can accommodate a wheelchair or the means to acquire one, public transportation can be a great alternative — if not the preferred alternative for mobility outside of the home. Those with impaired mobility should familiarize themselves with the options accessible to them by contacting their state department of transportation.

Now Is the Time to Take Action

It might not be time for a wheelchair or scooter yet, and perhaps that time will never come. Notwithstanding, it’s important that individuals become familiar with their diagnosis and whether it is known to impact mobility. They should consider what the future could look like, and monitor their symptoms and disease state progression so their first clue that they might need to look at adaptive options isn’t after they’ve taken a nasty spill and are dealing with the consequences of a debilitating, albeit possibly temporary, injury.

To maintain mobility and independence, now is the time to take action. Now is the time to plan!

Reference


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- Christina, SCIg Patient

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COMMON VARIABLE IMMUNE deficiency (CVID) is the most common serious primary immunodeficiency disorder and the one most often treated with immune globulin (IG) therapy. The disorder was first identified in a 39-year-old agammaglobulinemic woman in 1956 by Jay P. Sanford, MD, Cutting B. Favour, MD, and Melvin S. Tribeman, MD,¹ four years after agammaglobulinemia was described in a 9-year-old boy in 1952 by Ogden Bruton, MD.²

The incidence of CVID is approximately one in 25,000.³,⁴ Onset is usually between 20 years and 45 years old, so most patients are adults. Also, there is no single genetic cause; all cases are sporadic. Females have a slightly higher incidence of CVID except for those with disease onset before age 20 years. Caucasians have a higher incidence of CVID than Asians or Africans.

The definition of CVID includes 1) a marked reduction of serum immunoglobulin levels (IgG usually less than 400 mg/dl, low levels of IgA in 90 percent and low levels of IgM in 50 percent), 2) near normal numbers of B and T cells, 3) poor or absent antibody responses to immunizations, 4) undue susceptibility to infection and 5) no other immunodeficiency. Abnormal B and T cell function leads to antibody immunodeficiency and other manifestations of CVID.⁵,⁶

CVID Complications

As its name implies, CVID is highly variable with multiple complications in addition to infections (Table 1).⁵,⁶ Further, patients who have one or more complications have a 40 percent survival rate after 40 years compared to patients with infections only who have an 80 percent survival rate after 40 years.⁷ Multiple complications are common and can include pulmonary complications with granulomas, autoimmune cytopenias with splenomegaly and lymphoid hypertrophy. Most complications are lifelong and require specialty care, diagnostic tests and additional treatments.

The most common CVID complication is autoimmunity.⁸,⁹

By E. Richard Stiehm, MD
Autoimmune Disorders in CVID

Autoimmune disorders occur when some of a person’s defense mechanisms are directed at her/his own cells or tissues. These defense mechanisms include both autoantibodies and self-directed cellular immune reactions, which occur in 35 percent of all CVID patients at some time in the course of their illness.

Autoimmune disorders affecting CVID patients are listed in Table 2 in approximate order of frequency. The percentage of these disorders exceeds 100 percent, which emphasizes the coexistence of many of these conditions.

Causes of Autoimmunity

Why do CVID patients develop autoimmunity? Unlike patients with agammaglobulinemia, CVID patients produce some immunoglobulins and antibodies and have near normal and sometimes overactive T cell (cellular) immunity. The onset of autoimmunity usually occurs several years after the onset of CVID, suggesting that some event triggers it. The most likely event is an infection such as Epstein-Barr virus or cytomegalovirus infection. Both of these initiate an autoimmune response in normal subjects (e.g., multiple sclerosis after an Epstein-Barr virus infection and autoimmune cytopenias after a cytomegalovirus infection).

An infectious agent in a CVID patient may elicit an immune response to an infected or altered cell or organ that persists for a lifetime. These may persist because some CVID patients lack T regulatory cells directed against self-reactive clones of B or T cells.5 In addition, these aberrant responses may be enhanced by the presence of several autosomal genes that predispose to autoimmunity.10 Three of these genes are TACI (transmembrane activator and CAML interactor), ICOS (inducible T-cell costimulatory precursor), BAFF (B-cell activating factor), as well as several others.10 Up to 10 percent of CVID patients have one or more of these three autoimmune genes, and while some centers test CVID patients for these for research purposes, their presence or absence does not alter treatment.

Specific Autoimmune Illnesses

Autoimmunity can affect many different cells and organs. Severity is also variable, ranging from annoying (e.g., vitiligo) to debilitating (e.g., inflammatory arthritis) and life-threatening (e.g., granulomatous pneumonitis).

Three autoimmune disorders (splenomegaly, lymphoid hyperplasia and granuloma formation) are not discrete diseases, but they accompany many other illnesses and contribute to their pathogenesis and treatment. These three often coexist with each other.

Following is a brief description of autoimmune disorders that may accompany a CVID diagnosis:

Autoimmune cytopenias (30 percent). Autoimmune cytopenias are the most common autoimmune diseases in CVID,8,9 caused when autoimmune antibodies attack platelets (causing thrombocytopenia), red blood cells (causing hemolytic anemia) or white blood cells (causing low white blood cell count [neutropenia]). When this occurs, these antibody-coated cells are quickly removed from the circulation in the spleen and liver. Sometimes cytopenias are the first manifestation of CVID, particularly in patients under age 21 years. Fortunately, effective therapy is available, and these treatments sometimes result in complete remission.

Immune thrombocytopenia, sometimes called immune or idiopathic thrombocytopenic purpura (ITP), usually presents with bruising, bleeding and a very low platelet count (less than 20,000 cells/ul; normal is greater than 200,000 cells/ul). Brain hemorrhage can occur with very low platelet counts.

<table>
<thead>
<tr>
<th>Table 1. Clinical Phenotypes of CVID</th>
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<tr>
<td>• Infections (&lt;95 percent)</td>
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<tr>
<td>• Infections only (40 percent)</td>
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<tr>
<td>• Autoimmunity (35 percent)</td>
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<td>• Bronchiectasis (30 percent)</td>
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<td>• Splenomegaly (30 percent)</td>
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<td>• Lymphoid hyperplasia (25 percent)</td>
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<td>• Granuloma (25 percent)</td>
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<td>• Pulmonary disease (25 percent)</td>
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<td>• Gastrointestinal disease (15 percent)</td>
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<td>• Hepatic disease (10 percent)</td>
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<td>• Malignancy (5 percent)</td>
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<table>
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<tr>
<th>Table 2. Autoimmune Disorders in CVID</th>
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<tr>
<td>• Immune cytopenias (30 percent)</td>
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<td>• Splenomegaly (30 percent)</td>
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<td>• Pulmonary autoimmunity (25 percent)</td>
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<td>• Gastrointestinal autoimmunity (20 percent)</td>
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<td>• Granulomatous disease (20 percent)</td>
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<td>• Lymphoid hyperplasia (15 percent)</td>
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<td>• Rheumatic diseases (5 percent)</td>
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<tr>
<td>• Hepatic autoimmunity (5 percent)</td>
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<tr>
<td>• Others: Endocrine, Dermatologic, Nervous system (3 percent)</td>
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These antibody-coated platelets are removed quickly, principally in the spleen. Treatment includes high-dose corticosteroids and/or high-dose intravenous IG (IVIG). Refractory cases are sometimes treated with rituximab (a monoclonal antibody against B cells) or even splenectomy (removal of the spleen).

Autoimmune hemolytic anemia presents with pallor, fatigue and anemia due to antibodies against erythrocytes (red blood cells). These antibody-coated red blood cells are destroyed in the spleen to cause anemia (hemoglobin less than 10 g/dl; normal is 13 gm/dl). The released hemoglobin is converted to bilirubin in the liver to cause jaundice (yellow skin). Treatment is similar to that for immune thrombocytopenias.

Autoimmune neutropenia is the least common cytopenia and potentially the most serious since a low white blood cell count can lead to life-threatening infection. It is treated like other immune cytopenias in addition to the administration of the cytokine G-CSF to stimulate white blood cell production.

Evans syndrome is a distinct disorder in young children, which is the combination of immune thrombocytopenia and autoimmune hemolytic anemia and sometimes immune neutropenia.

Splenomegaly (30 percent). Splenomegaly is a common feature in many autoimmune disorders, particularly immune cytopenias and liver diseases with portal vein hypertension. The latter obstructs the flow of blood from the spleen, causing its enlargement. Unless the spleen is massively enlarged or causing severe cytopenias, it does not warrant splenectomy. The presence of a very large spleen carries the risk of splenic rupture, so contact sports should be avoided.

Granulomas (20 percent). Granulomas are a collection of inflammatory white blood cells, particularly macrophages, whose goal is to block a persistent foreign substance (e.g., a resistant bacterium, an inhaled particle or an altered self-antigen). Fibrosis and lymphoid hyperplasia are often present. Granulomas can be localized into nodules or present diffusely through an entire organ such as the lung or liver where they displace normal tissue to compromise organ function. Treatment usually requires a combination of steroids, anti-inflammatory drugs and immunosuppressive drugs.

Lymphoid hyperplasia (15 percent). Lymphoid hyperplasia is an expansion of lymphocytes into the lymph nodes, lungs, liver and gastrointestinal (GI) tract. Concurrent chronic Epstein Barr infection may be responsible in some cases. Like granulomata, lymphoid hyperplasia can be localized (e.g., in lymph nodes or the spleen) or diffused throughout the organ such as the bowel wall or lung. When severe, biopsy of a lymph node is sometimes needed to exclude a lymphoid malignancy. Treatment with steroids or immunosuppressive drugs are effective but rarely indicated.

Autoimmune pulmonary disease (25 percent). Pulmonary autoimmunity along with respiratory infections (e.g., bronchitis, pneumonia, bronchiectasis) combine to make the lung the most common target for CVID complications. Clinical features include chronic cough, dyspnea, fatigue and exercise intolerance. Autoimmune pulmonary diseases are often grouped under the term interstitial lung disease (ILD), which is characterized by diffuse lymphoid infiltration, granuloma formation, fibrosis and progressive loss of lung function.

ILD also includes two distinct syndromes: granulomatous-lymphocytic interstitial lung disease (GLILD) and nodular regenerative hyperplasia (NLH). GLILD patients have a distinct ground glass appearance on lung biopsy, and NLH patients have coexistent chronic liver disease with a characteristic pathologic picture. Both can be treated but not cured with corticosteroids and immunomodulatory drugs. Stem cell and lung transplants have been employed in a few patients.

Collectively, severe pulmonary disorders in CVID are the leading cause of a shortened life span; their mortality is about 40 percent 20 years after their diagnosis.

GI autoimmunity (20 percent). Autoimmune GI complications of CVID are manifested by abdominal pain, chronic diarrhea, malabsorption, anorexia and weight loss. They must be distinguished from the multiple infections affecting every portion of the GI tract, including candidial esophagitis with Candida albicans, gastric ulcers with Helicobacter pylori, small intestine enteritis with Giardia lambia and Cryptosporidium, and colitis with cytomegalovirus or norovirus.

Autoimmune gastritis results in reflux, atrophic gastritis and pernicious anemia. Autoimmune enteritis leads to a flat-
tended mucosa, lymphoid hyperplasia, malabsorption, diarrhea and weight loss. Inflammatory bowel disease involving the small intestine (Crohn’s disease) or large intestine (ulcerative colitis) are also present in some CVID patients.

**Rheumatic syndromes (5 percent).** The more common rheumatic syndromes in CVID are rheumatoid arthritis or juvenile idiopathic arthritis. Other rheumatic syndromes include lupus erythematosus, Sjögren’s syndrome, mixed connective tissue disease, dermatomyositis and Raynaud’s syndrome.

**Hepatic autoimmunity (5 percent).** Ten percent of CVID patients have mild elevation of liver enzymes. The most common serious liver disease associated with CVID is nodular regenerative hyperplasia characterized by a markedly elevated serum alkaline phosphatase, chronic hepatitis, portal vein hypertension with splenomegaly, ascites and jaundice. A liver biopsy shows multiple small nodules of regenerating liver cells and lymphocytes that impinge on normal liver cells. Treatment includes steroids, immunomodulatory drugs and sometimes liver transplantation.

**Other autoimmune disorders (less than 3 percent).** Less common manifestations of autoimmunity in CVID include skin disorders (scleroderma, vitiligo, psoriasis, alopecia), endocrine disorders, (thyroiditis, type 1 diabetes) and nervous system disorders (multiple sclerosis, peripheral neuropathy, uveitis).

**Treatment and Prognosis**

Most autoimmune diseases are lifelong but responsive to many modes of treatment. Most are treated with corticosteroids or high-dose IVIG. Other treatments include immunomodulating drugs such as azothioprine (Imuran), cyclophosphamide (Cytoxan) or rituximab (Rituxan), or anti-inflammatory drugs such as etanercept (Enbrel) and adalimumab (Humira). These drugs also inhibit the residual immune system making patients even more susceptible to certain infections. These patients must be informed about measures to prevent infection, including use of antibiotics, avoiding risky exposure and choice of vaccines for the patient and family. Prognosis is largely dependent on the type and severity of the complications and their response to treatment.

**Outlook**

CVID is the most common serious primary immunodeficiency of adults and the one for which IG is most frequently prescribed. The 45 percent of CVID patients whose only problem is an increased susceptibility to infection do extremely well on IG therapy even after 40 years. In contrast, those with one or more additional complications have a more guarded outlook. These complications include chronic lung disease, autoimmune cytopenias, inflammatory bowel disease and autoimmune liver disease. Effective management of these complications, in addition to IG therapy, provides patients with many years of long-term well-being.

**References**


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**Profile:**

**Lauren Dunlap**

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By Trudie Mitschang

**Trudie:** You were diagnosed with PI in 2010. What symptoms did you have that led to your diagnosis?

**Lauren:** Prior to my PI diagnosis, I kept getting severe, unusual infections. These infections also eventually led to a diagnosis of noncystic fibrosis bronchiectasis due to continuous pneumonia and upper and lower respiratory tract infections. I was prescribed antibiotics every week for something new, but no one seemed concerned about why this was happening, and no one asked why a 26-year-old woman would have such a rare lung disease.

**Trudie:** How did you meet Dr. O’Connor?

**Lauren:** I met Dr. O’Connor on a cold, sunny February day in 2010. After reviewing my immunoglobulin panel, my ENT, Chad Kessler, sent me to her and said I needed to get in as soon as possible. I remember I was looking out the ceiling-to-floor window waiting for her to enter the room and thinking to myself, “This is going to be just another doctor who doesn’t believe me. This appointment is going to be a waste of time.” I turned around when she came in and was greeted with a smile from a physician who forever changed my life.

**Trudie:** Why do you describe your relationship with Dr. O’Connor as “hitting the doctor lottery”?

**Lauren:** I had gone through many physicians prior to being diagnosed with PI (and even afterward), and none of them believed me or took seriously the symptoms I was having, even though I was severely ill and continued to deteriorate. The physical pain was unbearable at times. When patients have a physician who believes them, partners with them in their care and makes decisions with them together as a team, this is worth more than any amount of money a person could receive. This has always been my relationship with Dr. O’Connor in a clinical setting. She is true to the Hippocratic oath she took as a physician.

**Trudie:** How do you encourage patients to keep searching for their own Dr. O’Connor?

**Lauren:** It’s such a rare thing to be able to find these special doctors, but I tell patients they do exist. Persistence is key! I cannot believe after all I’ve been through, I have not only hit the “doctor lottery” once but multiple times. This includes meeting my National Institutes of Health immunologist Ivan Fuss, my primary care physician Melissa James and several other specialists who have been with me on this complex and often difficult journey. A lifetime of thanks would never be enough to show my gratitude toward each of them.

**Trudie:** Tell us about your early days of volunteering for the Immune Deficiency Foundation (IDF).

**Lauren:** I have been a volunteer with IDF for many years. When IDF held their first three national fundraiser walks in 2013, I wanted to get involved and do more. In 2014, I flew to Chicago to participate in the IDF Walk there. I phoned Dr. O’Connor from the airport afterward and said, “We need to have one in Charlotte. Let’s do this!” Over the course of a year, we planned the walk in Charlotte as volunteers. When October
2015 rolled around, we had 120 participants on a beautiful fall morning and raised almost $15,000 for IDF. We went on to host another walk in 2016. I have also been an IDF Get Connected Leader, a guest speaker for various educational days and Teen Escape weekends, and just this year I participated in IDF Advocacy Day on Capitol Hill.

Trudie: How has PI changed your life for better?

Lauren: PI has changed my life for the better because it helped me to appreciate the little things in life, and it has also helped me to appreciate my own health. Many times, people take their health for granted, and I often say, “If you don’t have your health, you don’t have anything.” I have lived this time and again, and can state without hesitation it is a very true and real statement.

Trudie: Tell us about the origin of AAIDA.

Lauren: The calling to start a nonprofit has always been in my heart. After Dr. O’Connor and I planned and executed two successful walks for IDF and began volunteering for other organizations, we started to discuss the idea of getting our own nonprofit off the ground. This was a lifelong dream for both of us. Our nonprofit would not be focused on just PI or be a disease-specific organization. We wanted to incorporate all immune dysregulation conditions, which included PI, secondary immunodeficiency and autoimmune diseases, to help more patients across the U.S and abroad. I dove in headfirst and started the 501(c)(3) process after we agreed on a name for our organization. We launched AAIDA in February 2017.

Trudie: How has AAIDA grown and evolved, and what are your goals for the future?

Lauren: It’s amazing to see how much AAIDA has grown and accomplished in such a short time. AAIDA is an all-volunteer nonprofit that is governed by a board of directors. Although we have many goals for the future, one is to eventually have a full-time staff to better equip and educate patients and physicians about immune conditions they live with or treat. Being an available resource to patients has always been our No. 1 priority. AAIDA has also been an active voice on Capitol Hill over the last three years in the area of patient advocacy, and we hope to one day have a person dedicated to being the patient’s voice on a full-time basis.

Trudie: What are you thankful for?

Lauren: I am thankful for so many things it’s hard to narrow it down to a single paragraph or sentence. I would say I am thankful for my family and friends who have been with me through my diagnoses of PI and all the illnesses along the way. I am also thankful for the many opportunities I have been given on this ever-changing journey called life. Most important, I am thankful to be alive and to be able to share my journey with other patients to give them hope and reassurance that they are not alone, even if it feels like they are.

Trudie: What is your favorite inspirational quote or message?

Lauren: “In the midst of hate, I found there was, within me, an invincible love. In the midst of tears, I found there was, within me, an invincible smile. In the midst of chaos, I found there was, within me, an invincible calm. I realized, through it all, that… In the midst of winter, I found there was, within me, an invincible summer. And that makes me happy. For it says that no matter how hard the world pushes against me, within me, there’s something stronger — something better, pushing right back.” — Albert Camus

Trudie: What’s the best advice you’ve ever received?

Lauren: This isn’t advice I received directly (I wish I could have met him in person though!), but a quote I live my life by and also say to myself every single day: “Be who you are and say what you feel because those who mind don’t matter and those who matter don’t mind.” — Dr. Seuss

Trudie: Finish this sentence: If I knew then what I know now, I would...

Lauren: …I would not have wasted time on what others might think. I would have started to appreciate the little moments in life more, long before 2010. I would have gotten up to watch more sunrises. There’s going to be a lot of heartache and pain along the way, but life is so precious and a gift — vis ta vie (live your life).
MY LIFE resembles a bop bag toy. You know, the one you can punch, slap or body-slam to the ground, yet it pops back up no matter what? Well, I have been punched, slapped and body-slammed to the ground by my disease and other people, but no matter what, I always bop back up. And I’m so glad I’ve mastered this powerful defense because I might not have bopped back up when I came face to face with the unexpected, but familiar past.

In my previous column, I shared my experience that happened when I was a junior at Asbury University, when I thought I had blossomed as a person. Actually, it was my senior year when I fully bloomed. It was then I was recognized as a leader on campus, and I became a part of a group known among the student body as special and unique. The soundtrack of my life was “I’m Walking on Sunshine,” and it felt good!

One beautiful sunny day, I headed to the new state-of-the-art communications building for my journalism class. I headed up the elevator, walked off with a smile on my face, turned the corner and stopped. There she was. The student-turned-doctor who had diagnosed me with dwarfism. I knew God had a sense of humor, but this was ridiculous. If I turned to walk the long way to class, I’d be late. But if I passed by her, she might recognize me. Decision made, I said, “Stop, Whitney! You are a confident woman now, so just march right past her to your class and be done with it.”

“You look familiar,” she said. Does nothing get past this girl? Nothing?

I put on my “be the bigger person” smile, while mentally kicking myself for not taking the long way to class. A tardy looked pretty good about then.

“You look kind of familiar too,” I replied. I was not lying, just not revealing too much.

Don’t say it, don’t say it, don’t say it, I thought.

“You’re the girl I thought was a dwarf!” she exclaimed. Punch! She knocked me down, but I bounced right back up.

“That’s me,” I said. I took note of the small victory that she used the word “thought.” She must have done more research. But, I had never been more wrong.

“But aren’t you? I really think you’re a dwarf,” she persisted.

Slap! She still believed I had life-altering truths about myself I had never accepted. But, I floated right back up from the ground because I had the perfect comeback, a response I knew she could not dispute.

“No, I’m not a dwarf. Look at my arms and legs, they are…,” I replied, but she didn’t let me finish.

“They aren’t proportional, are they?” she blurted. Sadness and regret etched over her face.

Body slam! But just like Rocky Balboa, I popped back up just when it counted.

“They are proportional,” I replied. “But, you know what, I have to get to class. You have a great day.”

I left this girl once again chuckling, except this time, I was secretly hoping she would never see me with my mom who is only 4 feet 8 inches tall. She would think we stepped right out of a Disney movie.

Here’s what I’d like you to take away from this story: You’re going to have days when you forget you have an immune disease, you feel great and you’re juggling friendships, work and family life at just the perfect balance. And then, someone knocks you off your feet. No matter who causes the impact, it won’t lessen the hurt, confusion or urge to educate that person so they may become more compassionate and understanding about immunocompromised people.

It’s been my experience that despite all of the things your disease has taken from you, it has also given you so many qualities no one can ever take away: determination, perseverance and strength. And because of that, my friends, whenever you are punched, slapped or body slammed to the ground by life, you won’t remain down for long.

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.
We Deserve to Find a Supportive Partner

By Michelle Searle

I REMEMBER it like it was yesterday. I was sitting in a circle with other teens and a therapist at an Immune Deficiency Foundation (IDF) Teen Escape. One girl was telling us how supportive her boyfriend was about her primary immunodeficiency. She said having a partner who is supportive is something we all deserve and not to settle for anything less. Those words have always stuck with me. I’ve heard many people express discomfort when talking to their partners about their health issues, about the lack of support they receive, and about people no longer wanting to continue dating after learning about their disease. Finding the right partner is difficult enough; add the fact that you have a chronic illness, and it becomes much harder.

That teenage experience taught me it was important to find someone who would show interest in my disease, be there for me on my bad days, have a shoulder for me to cry on, but also encourage me to pursue my dreams. I also wanted someone who would see me for me, and not just see my disease.

About six years after that Teen Escape, I was on a dating app looking for an Italian guy in my area to help me practice speaking the language before I moved to Italy. I met Marco who was on Tinder looking to make new friends. On the first night we met, he told me he read an article about me and my life with common variable immunodeficiency (CVID) in our college newspaper. He said he did some research about CVID and realized how difficult my life must be, and he wished he could have met me sooner. I’m not someone who usually talks about my immune deficiency on a first date, but I’m also not going to hide it. I told him the brief history of my disease, and much to my surprise, he asked me many thoughtful questions.

Marco and I liked each other from the moment we met, and within a week of knowing each other, he said he wanted to be there for my next infusion. Although I was a little nervous, I was excited. Since moving away from home, I was used to doing my infusions alone, so it was nice to have someone to share that time with again. We spent our first infusion together watching “The Wolf of Wall Street” and baking cookies that he surprised me with. During my infusion, he was nervous if I moved, and kept telling me he would help with whatever I needed. After the infusion, I suggested getting pizza, but he was concerned about my ability to walk around, thinking it might mess up how the medicine absorbs into my body. His concerns were so sweet and cute that I couldn’t help but laugh.

Since that infusion, I have not done one infusion by myself. Though we often are living on separate continents, he is always on the other end of the phone for every infusion. It has become a special biweekly date for us.

For me, sometimes the hardest part of living with a chronic illness is feeling unheard, not believed and alone. With this relationship, I’ve learned what it’s like to be with someone who hears me and reminds me I’m not alone. He shows his interest in my disease and his support by always learning more about CVID, going to doctor appointments with me, making us dinner while I infuse, attending support groups and events, and more.

My intent is not to tell you my love story nor to say I’ve met the perfect guy. Instead, I want to share that I’ve learned not only how I would like to be treated in regards to my disease, but also how I deserve to be treated. While we are more than our chronic illnesses, we still need people in our lives to remember and acknowledge we have them. We should never feel ashamed about our illnesses or our struggles because they are what makes us strong and unique. They help make us who we are.
AS A MOTHER of three children with primary immunodeficiency (PI), I’ve handled most of my kids’ medical care throughout their lives. Because I was a stay-at-home mom when they were young, I was home during the day to administer their weekly subcutaneous immune globulin infusions, and I drove them to all of their doctor appointments — even the ones that were three hours away. I’ve ordered prescription refills, scheduled deliveries and handled the beginning-of-a-new-year insurance hassles. I’ve done it all happily, knowing my efforts are keeping my kids healthy. I also want them to have a normal childhood, without having to worry about their healthcare. And why should they? I’m Super Mom. I’ve got this.

But now, I have a 17-year-old. He has two years at home before he leaves the nest, and it’s beyond time for him to learn to take responsibility for his own medical care before he’s on his own trying to figure things out without his mother’s ever-present guidance. And while I’m transitioning him to self-care at the eleventh hour, it would probably be wise to start handing over responsibilities to his younger brothers earlier, teaching them a little at a time so their transition period doesn’t feel like a last-minute cram session.

What Is Self-Care?

Self-care or self-management is defined as “the set of behaviors that people engage in as part of living with a chronic health condition.” Self-care can include many tasks that children and teens are more than capable of doing on their own such as taking medication on time, monitoring symptoms, eating a well-balanced diet (and avoiding non-nutritious food), keeping physically active with a fitness routine or sports, learning new skills, setting goals, getting a good night’s sleep and practicing stress-reduction techniques (such as meditation, yoga or deep breathing). Some self-management tasks include responsibilities that directly relate to and affect a person’s healthcare (such as getting IgG levels checked or receiving infusions). Social and lifestyle behaviors such as attending school or trying out for a school play or sports team can impact wellness in a more indirect way. All of these are tasks and decisions older children and teens can manage on their own or with the help and supervision of a parent.

Benefits of Teen Self-Care

The adolescent years are a time of self-discovery. Teens are looking for ways to separate themselves from their parents, create their identity as an individual and make their own way in the world. When it comes to managing chronic illness, the teen years are a very natural time to gradually hand over responsibility for health management. According to the American Academy of Pediatrics, teens are actually more compliant when they’re involved in their own healthcare decisions. They follow the treatment plan more closely (because they had a say in it), while also learning to solve problems and overcome challenges.

Work on a Schedule

One of the first things parents can do to get their children involved in their own healthcare is to get their input on infusion or therapy schedules. When our children were young, we administered
infusions on Sundays. As our children got older, weekends became busy and we tried to fit infusions in whenever we could. As kids mature, having them sit down with parents weekly or monthly to go over schedules and plan infusions is a good way to get them involved in their healthcare. This will also avoid disappointment or surprise when they find out they can’t go to that much-anticipated party or sporting event because it’s “infusion day.” Planning ahead makes life much easier for everyone.

Some other examples of self-care responsibilities that kids and teens can take on are taking medications without reminders, drinking plenty of fluids (especially before and during infusions), keeping up with schoolwork and scheduling clinic appointments. Not only does self-management of healthcare improve a teen’s overall health by increasing adherence to their treatment plan, but it also helps to improve the teen’s ability to solve problems and overcome challenges.¹

Teaching Teens to Self-Infuse

One of the biggest components of healthcare for teens with PI is the administration of infusions. It is a necessary part of life that occurs weekly or monthly, depending on the method of administration. For teens who receive subcutaneous infusions, acquiring the skill of self-infusion is a must if they wish to take over their own healthcare and relieve their parents of the duty someday.

Most parents receive instructions on how to perform infusions on their child by a nurse at a hospital or infusion center. Sometimes a nurse comes to the home to carry out this instruction, demonstrating the many steps involved in a successful infusion. Parents must learn about proper hygiene, choosing the right needle insertion site, numbing the skin, priming the pump, infusing the medicine at the right speed and disposing of needles in a safe way.² Parents are instructed to keep track of the infusion schedule so they take place at roughly the same day each week, going no more than 10 days between subcutaneous infusions in order to keep IgG levels high.

Because infusing takes precision, a steady hand, cleanliness and a calm environment, turning over this responsibility to their kids can be difficult for parents. Many parents might feel more comfortable with continuing to manage the infusions for their children because they know they’ll do it right. Letting go of control and learning when to back off and let the child take over can also be a challenge. Another challenge is knowing the right age to start this transition process. According to Debra Moffitt at CSL Behring, a manufacturer of several immune globulin medications, “There’s no one ‘right’ age when a child should take over this important task. But kids often learn this skill between the ages of 8 and 14.”² Because each child or teen is an individual, every child will approach learning to self-infuse differently, and parents, who know their children best, should be able to tell when their child is ready to begin the process.

Even after learning how to perform their first few self-infusions, kids will still need occasional help. As parents know from experience, things can always go wrong. Sites can leak, needles can go in cockeyed, and intense burning or itching can occur during or after infusions. This is why it’s important for parents to provide gentle guidance and instruction, sometimes watching from a distance and sometimes being a more hands-on assistant. “An independent and not-independent back and forth is often part of the learning process,” says Moffitt.²

Teaching Independence Is Necessary

As much as parents want kids to be independent, it can be a hard step to pass their healthcare responsibilities on to them. As with all of life’s milestones, there is a lot of emotion and anticipation when a child begins to take on his or her own self-care. But children eventually become adults, and adults with chronic conditions such as PI need to be independent and manage their own health if they’re going to thrive. Parents can be their children’s best supporters by keeping things positive, knowing when to step in or back off, and by giving practical and encouraging tips.²

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.
TAKE THESE meds four times a day and this one twice. Monitor your blood pressure. Check your glucose levels after every meal. Sounds like quite the to-do list, right? For the more than 133 million Americans living with a chronic condition, this is their reality. Yet, for many of these individuals, adding these complicated daily regimens to an already bursting schedule is a recipe for failure. And when patients fail to take medications on time, fail to stick to a specific diet or slip back into old habits, they often end up back in the emergency room.

Captive Audience
Thanks to the growing number of disease-specific mobile health apps, this picture is starting to change. More than 194 billion apps were downloaded in 2019, up from 178 billion in 2018, and there are currently at least 165,000 health-related apps available, according to App Annie, a global provider of mobile market data.

Chronic disease management is a huge player in this arena with apps that deal with everything from heart and lung disease to arthritis and diabetes. And these days — particularly with the onset of the novel coronavirus — many patients are turning to their phones instead of heading to the doctor’s office.

The No. 1 Killer
Nothing causes more deaths in this country than heart disease and stroke, according to the Centers for Disease Control and Prevention (CDC). Every year, more than 859,000 Americans die of these conditions that deliver a serious blow to our healthcare system, costing a whopping $199 billion per year and causing $131 billion in lost productivity on the job, says the CDC.

So, what can a little app on a smartphone do to help curb this massive health issue? Maintaining blood pressure is key when it comes to combating heart disease, and it turns out apps are a huge help in this department.

For instance, the app Blood Pressure Companion Pro tracks heart rate, measures blood pressure and then analyzes the data. In addition, patients are free to share the results with their healthcare team. Another success story is the remotely monitored electronic pillbox that’s included on some apps. This technology helps alert heart failure patients to take their medications.

Dealing with Diabetes
A variety of apps are available to manage both type 1 and 2 diabetes. The free app Glucose Buddy has received rave reviews for its user-friendly interface and ability to allow patients to log in and check blood glucose levels, carbohydrate intake, medication doses, test results, exercise and more. The downside to this free app? It doesn’t sync with meters, continuous glucose monitors or pumps.

Patients at risk of developing type 2 diabetes can look to the Omada Health app to curb the onset of this disease. The 12-month program kicks off with a 16-week online course teaching better lifestyle habits. Each participant is assigned a personal health coach and is expected to log their dietary intake and exercise.

Patient Control
Disease-specific health apps give patients the steering wheel, allowing them to become more confident advocates of their own health. And, a bonus is the apps give doctors the opportunity to gather information about patient behavior and intervene when protocols aren’t followed.

But not all apps are stellar. Only 10 percent can connect to a device or sensor while 2 percent sync into providers’ systems, according to the IMS Institute for Healthcare Informatics. Patients should check with their physician to find out which app will work best for their specific ailment and, hopefully, help them avoid landing in that not-so-enticing emergency room.

HEATHER BREMNER CLAVERIE is a contributing writer for IG Living magazine.
**An Apple a Day**  
The Apple Watch Series 5 not only includes all the core fitness, step-tracking, heart monitoring and connectivity features that make these devices so popular, but Apple has tacked on even more health-inducing additions to its newest watch. Download the ECG app and the 5 generates a single-lead electrocardiogram (ECG). This ECG addition can help patients deliver critical heart data to their doctors and give them a little peace of mind. The 5 also tracks menstrual cycles and buzzes if noise levels rise to a point that it could impact hearing. Also, leaving it on while sleeping gives users a full sleep report when the sun rises. *Starting at $399; www.apple.com/watch*

**Relieve Some Pressure**  
There's no need to head to a drugstore to check your blood pressure when you have the OMRON Evolv blood pressure cuff. Individuals can fasten the cuff to their arm in the comfort of their own home to track blood pressure. And, the numbers can be easily shared with doctors to manage heart health with the Kardia app. *$89.99; store. alivecor.com/products/omron-evolv*

**Join the Club**  
At One Drop, the motto is “we work relentlessly to make life with diabetes better.” What’s one way the New York-based company transforms those words into actions? With diabetes supplies and support — all available through its website. One of those is the Diabetes Health Package, which includes a Bluetooth-enabled glucose meter and a membership to SugarRx, One Drop’s digital coaching and education program and a new meter annually. The company also provides a free mobile app that tracks and analyzes blood sugar, medications, food and activity. *Packages start at $22.95/month; onedrop.today/products/diabetes-package*

**Hand-to-Heart**  
Placing one’s fingers on the little silver pads on AliveCor’s compact EKG provides stats about heart health within 30 seconds. The U.S. Food and Drug Administration-cleared portable device detects atrial fibrillation, bradycardia, tachycardia or normal heart rhythm. The device is available with one or six leads, with the six-lead providing doctors more detailed heart information. Connecting the KardiaMobile device with the Kardia app keeps track of information. *$89 for Kardia Mobile and $149 for KardiaMobile 6L; www.alivecor.com/kardiamobile*

**Dear Diary**  
Individuals can start recording how they’re feeling in My Pain Diary & Symptom Tracker: Gold Edition to get more answers about what is causing their chronic pain. The app tracks arthritis, depression, fibromyalgia, reflex sympathetic dystrophy/complex regional pain syndrome and undiagnosed conditions. With smart features like automatic weather tracking and reminders, it’s easy to customize the app to specific needs. *$4.99; mypaindiary.com*

**Track Your Day**  
Adding what foods have been consumed to a MyFitnessPal food diary can help people get back on track with their dietary goals. The free app breaks down ingredients, adds up daily caloric intake and tallies up exercise. Goal weight can also be added to the app, which will break down how many calories a person can consume per day to hit that target. *Free; www.myfitnesspal.com*
The Pain Survival Guide: How to Become Resilient and Reclaim Your Life, Revised Edition
Authors: Dennis C. Turk and Frits Winter
Publisher: APA LifeTools

The authors of this bestselling book are highly respected experts on pain management who have successfully treated thousands of patients. Their 10-lesson self-management program offers clinically-proven strategies for making simple, gradual adjustments to daily patterns so individuals can cope with chronic pain and take charge of their life. Readers will learn how to identify and reject harmful myths about pain; balance activity and rest; fix sleep problems; leverage family and friends for support; manage emotional response to pain; regain self-confidence; and avoid relapsing into unhealthy behaviors. This edition offers important information on the risks of opioid use for chronic pain, and discusses new treatment options, including acceptance and commitment therapy, mind-body interventions such as tai chi, and cannabinoids.

Celiac Disease (Updated 4th Edition): A Hidden Epidemic
Authors: Peter H.R. Green, MD, and Rory Jones
Publisher: William Morrow Paperbacks

In this user-friendly guide, celiac disease specialist Dr. Peter Green and writer Rory Jones help readers identify the symptoms, understand their diagnosis and cope with the challenges of living with celiac disease. The book covers the disease from a medical, dietary and psychological standpoint, covering all aspects of celiac disease from the varied symptoms and proper diagnostic process to the many related conditions and specific diet. The book also includes appendices on related articles and books of interest, medical contact information and a list of national celiac disease support groups.

The Things We Don’t Say: An Anthology of Chronic Illness Truths
Author: Julie Morgenlender
Publisher: Three Barrel Bluff

Spanning different ages, ethnicities, genders, sexual orientations and diagnoses, 42 authors from around the world open up in 50 true stories about their chronic illnesses and their search for answers, poor treatment by doctors, strained relationships with loved ones, self-doubt and more. They share the warmth of support from family and friends, the triumph of learning coping mechanisms and finding ways to live their dreams. These stories are honest, raw and real, and people with chronic illness will find comfort and companionship in these pages. It is also for those who have ever wanted to know more about their loved one’s experience with chronic illness but didn’t want to ask the wrong questions.

Chronic: The Hidden Cause of the Autoimmune Pandemic and How to Get Healthy Again
Authors: Stephen Phillips, MD, and Dana Parish
Publisher: Houghton Mifflin Harcourt

Dr. Steven Phillips and his former patient, singer-songwriter Dana Parish, reveal evidence that a broad range of microbes cause a variety of recurrent conditions and autoimmune diseases. Dr. Phillips was an internationally renowned physician specializing in complex chronic diseases when he became a patient himself. After nearly dying from his own mystery illness, he experienced firsthand the medical community’s ignorance about the pathogens that underlie a deep spectrum of serious conditions, including Lyme disease, fibromyalgia, lupus, multiple sclerosis, chronic fatigue syndrome, rheumatoid arthritis, depression, anxiety and neurodegenerative disorders. Parish also watched her health spiral after 12 top doctors missed an underlying infection that caused heart failure and other sudden debilitating physical and psychiatric symptoms. In this book, they come together with a mission: to change the current model of simply treating symptoms and shift the focus to finding and curing root causes of chronic diseases that affect millions around the world.
“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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<td>- GBS/CIDP Foundation International: <a href="http://www.gbs-cidp.org">www.gbs-cidp.org</a></td>
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<td><strong>Idiopathic Thrombocytopenic Purpura (ITP)</strong></td>
<td>- ITP Support Association – UK: <a href="http://www.itpsupport.org.uk">www.itpsupport.org.uk</a></td>
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<td><strong>Kawasaki Disease</strong></td>
<td>- American Heart Association: <a href="http://www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsofChildhood/Kawasaki-Disease_UCM_308777_Article.jsp#T1T2boePWE0">www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsofChildhood/Kawasaki-Disease_UCM_308777_Article.jsp#T1T2boePWE0</a></td>
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<td>- All About Multiple Sclerosis: <a href="http://www.mult-sclerosis.org/index.html">www.mult-sclerosis.org/index.html</a></td>
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<td><strong>Secondary Immune Deficiency</strong></td>
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<td><strong>Stiff Person Syndrome (SPS)</strong></td>
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<td>- Living with Stiff Person Syndrome (personal account): <a href="http://www.livingwithspss.com">www.livingwithspss.com</a></td>
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<td><strong>Scleroderma</strong></td>
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