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About IG Living

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Knowledge Is Power

Two hundred years ago, Thomas Jefferson wrote: “There can be no stronger proof that knowledge is power, and that ignorance is weakness.” That astute phrase could not ring truer today when applied to the strength acquired from better understanding the challenges individuals with chronic illness face. I hope this issue of IG Living will provide our readers with knowledge that will alleviate your concerns about treatment safety, explain treatment coverage to help you plan effectively, empower you to take advantage of accommodations that can make life easier and find purpose in the struggles you face.

It is highly likely that immune globulin (IG) therapy was unknown to our readers prior to their rare disease diagnosis for which there is no alternative drug. And since each batch of this precious resource is manufactured from human plasma collected from between 1,000 and 15,000 donors, it is understandable there is concern about IG safety. In fact, IG Living’s patient advocate Abbie Cornett is asked so frequently about the safety of IG that she authored the article “How Safe Is IG Therapy?” (p.20). Here, she explains how this drug is manufactured and what safety measures are in place to ensure IG therapy is safer today than ever before.

As our readers are well aware, due to huge numbers of donations required and the time-consuming, multistep manufacturing processes to produce IG, it is a very costly drug. It simply would not be possible for most people treated with IG to afford without insurance coverage. And, for those on Medicare, even the 20 percent cost-sharing would be insurmountable. So, in our article “Purchasing a Medigap Policy” (p.24), we provide clarity about Medicare coverage and how to obtain a supplemental policy that will ensure beneficiaries can continue to afford their IG treatment.

Even with IG therapy, those struggling with rare diseases battle a host of concerns that without programs and services created by governments and other organizations, their daily activities would simply be much more of a burden. Many of these well-deserved accommodations related to travel, parking, medical devices, public transportation and more are highlighted in our article “Getting the Most Out of Your Disability” (p.26).

Finally, while living with a chronic illness can seem only burdensome, there are many life lessons to be learned that can provide inspiration and purpose. Author Matthew D. Hansen, a physical therapist, speaks from experience in his article “Life Lessons: Becoming Stronger Through Illness” (p.29). He encourages people to accept and champion their cause and to look for the benefits their illnesses may have for others.

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
Social Distancing Doesn’t Have to Mean Isolation

By Abbie Cornett

THE COVID-19 pandemic has been challenging for people on multiple levels. Besides the threat of the disease itself, people have had to contend with unprecedented economic hardship and the effects of social distancing. But, social distancing, a policy or set of actions taken to stop or limit face-to-face contact to decrease the spread of illness among people in community settings like schools, churches, restaurants and grocery stores, to name a few, comes at a cost, too.

Humans by nature are social creatures.1 This means social distancing can lead to feelings of isolation. And, while being lonely is bad enough, it isn’t the only consequence of feeling isolated. Studies have shown people who socially isolate experience both physical and mental consequences ranging from increased blood pressure, obesity, heart disease, spikes in cortisol levels, sleep disruption, anxiety and depression.2 These issues can lead to significant reduction in quality of life and an earlier death.

People who are elderly or have a chronic illness are at an even higher risk. Research has shown adults with chronic conditions and physical or cognitive limitations are more than two times as likely to report feeling socially isolated (37 percent) than adults who do not have these health issues (15 percent).3 During such stressful times, it’s important to recognize the signs of feeling isolated in yourself and your loved ones and work to alleviate it.

Fortunately, the good news is social distancing doesn’t have to mean isolation. The first step to limit seclusion from others is to make a plan to connect by making a list of family members and friends you miss and setting up a time to talk or, better yet, setting up a meeting for a virtual hangout through apps like Skype, FaceTime, Zoom or Netflix Party. These apps allow people to watch movies, do crafting projects and even eat dinner together electronically!

Consider other ways to reduce feelings of isolation:

• Get outside! Social distancing doesn’t mean you have to remain locked in the house. If you are physically able, take a walk. If you can’t, sit outside where you can see your neighbors and wave. Just seeing other people can help you feel connected. If you can’t go outside, talk to your doctor about what physical activities you can do inside of your home.
• Start a project. Having a sense of purpose will keep you feeling connected. Pick an activity that means something to you. If you sew, make face masks to donate to healthcare organizations.
• Start a garden. Waiting for flowers to bloom or harvesting what you have grown will give you something to look forward to.
• Keep your mind and body active. This is the perfect time to read that book you’ve had on your list, or to take that online course. Many universities are offering free classes in almost every subject. To raise your fitness level, consider the many workouts available online that may suit your needs.
• Make a schedule. A schedule helps keep you on track, and will help you to feel engaged throughout the day. Further, it will prevent you from feeling like time is at a standstill.
• Practice self-care. This is a time to make sure you are eating well and getting enough sleep and exercise.
• Consider adopting a pet. Animals are a great source of companionship. Better yet, having a pet has shown to lower stress and blood pressure.

No one knows how long we are going to have to physically distance ourselves from others, but with a little bit of forethought and planning, isolation and loneliness don’t have to be the new norm. Stay healthy, and stay connected!

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

References

How Loneliness and Isolation Affect Mental Health

• Physical symptoms: Aches and pains, headaches, illness or worsening of medical conditions
• Mental health conditions: Increased risk of depression, anxiety, paranoia or panic attacks
• Low energy: Tiredness or lack of motivation
• Sleep problems: Difficulty getting to sleep, waking frequently or sleeping too much
• Diet problems: Loss of appetite, sudden weight gain or loss
• Substance use: Increased consumption of alcohol, smoking, medications, drugs
• Negative feelings: Feelings of worthlessness, hopelessness or thoughts about suicide

Do you look at the bright side of life?

I do. After my cancer diagnosis, I had a really hard time doing so, though. With the help of a therapist, I was able to get back to a good place again, and even with continuing health issues, I consider myself to be a positive person. I am not going to waste my life moaning over the things I can’t change or have no control over. Life is too precious and short!

— Debbie K

How do you fight loneliness?

I live alone, so I am quite used to being by myself. If I find myself feeling a bit lonely, I usually pick up the phone and call a friend, relative, etc., or I involve myself in a project to distract myself.

— Donna G

I would normally say by surrounding myself with family and/or friends. This flu would bring that to a screeching halt. That’s when my faith would take over.

— Jenny G

I talk to God. He always listens, and I feel he answers, too. Prayer. Connecting with others. The best way is trying to help someone else. Use your pain for purpose.

— Jerri O

What does it mean to have an underlying condition during a pandemic?

A company has been working very hard to get an oral medication out for a certain primary immunodeficiency disease population whose only current medication consists of weekly intravenous or subcutaneous infusions. It’s really hard to see people say that this medication only serves 0.002 percent of the population, and they would rather see the company work on a COVID-19 medication. I understand the importance, but please don’t push us aside. This medication has already proved itself through the first trials. I have developed what looks like a nipple over my vein because I have to access it every other day to stay out of the intensive care unit. I also see people say they don’t care about the immune-compromised population because we are the minority. These are painful to read. Are we meant to feel like a burden to society?

— Patricia MP

What it means? It means it’s harder to see your personal care physician. It’s harder to obtain a referral for my intravenous immune globulin. Mine is sitting in a pending status. Just great; more anxiety knowing my body needs the immune cells and I won’t be receiving them as scheduled.

— Peggy SG
According to Leslie Vaughan, chief operations officer at Nufactor, a Specialty Infusion Company, no one has the answer to your question yet. If there are enough people exposed to COVID-19 who develop antibodies, it is reasonable to assume IG produced from their plasma may contain antibodies. However, unless the U.S. Food and Drug Administration adds a requirement for IG manufacturers to test for COVID-19 antibodies, the answer may never be known. It is important to remember that all IG manufactured in the U.S. must be made from U.S.-sourced plasma. And, as of this writing, since the peak for exposure to COVID-19 in the U.S. has not yet been reached, current plasma donations may have only a small amount of COVID-19 antibodies. Assuming mass exposure and antibody development in the next several months, IG would likely contain COVID-19 antibodies between nine and 12 months post-peak exposure. So, there likely won’t be COVID-19 antibodies in IG to provide a protective effect for a long time, minimally a year.

As a patient who receives intravenous immune globulin (IVIG) therapy every four weeks, I am curious how long it might take for COVID-19 antibodies in the IVIG therapy to provide a protective effect against the virus. Is there any chance COVID-19 antibodies are already present?

I spoke with Terry O. Harville, MD, PhD, medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, regarding your son’s injury, and he said using inhaled corticosteroids for several years can cause osteoporosis. Further, some patients with immune deficiencies have osteoporosis. Because of this, he recommends your son have a bone density scan and receive the appropriate therapy based on the results.

I spoke with Terry O. Harville, MD, PhD, medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, regarding your son’s injury, and he said using inhaled corticosteroids for several years can cause osteoporosis. Further, some patients with immune deficiencies have osteoporosis. Because of this, he recommends your son have a bone density scan and receive the appropriate therapy based on the results.

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

ABBIE CORNETT is the patient advocate for IG Living magazine.
Type I Hypersensitivity: ‘True’ Allergic Disease, Part 4

By Terry O. Harville, MD, PhD

IN PREVIOUS issues, we have discussed the differences between hypersensitivities and allergic diseases, most recently discussing the issues associated with food allergies. A true food allergy is one in which IgE antibodies have been generated against a food item, which occurs when only a tiny exposure to the food results in an allergic reaction through the release of mediators from mast cells. Allergic reactions can appear as rashes, hives, abdominal cramps, bloating, vomiting, diarrhea, itching and possibly anaphylaxis.

Some food items such as strawberries may or may not result in a true allergy. For example, if a person is able to eat three or four strawberries without any allergic symptoms, but then eats six strawberries and develops a prickly-appearing rash on the chest, this is likely not a true allergy. Instead, it is most likely an intolerance, which some refer to as pseudo-allergic reactions (which occur when the rashes and symptoms appear somewhat indistinguishable from allergic reactions). In fact, a chemical in strawberries can cause this nonallergic rash, which is an example of a dose-dependent reaction (typical of an intolerance). True IgE-mediated allergic reactions are dose-independent, meaning even a tiny exposure could result in a severe allergic reaction. This is a very important distinction because if a child has, for example, an egg allergy, and someone says, “One little bit of a cake won’t hurt,” this is not true. Even a tiny bite of egg-based cake will result in an allergic reaction that will require an epinephrine injector to treat the impending anaphylaxis.

There are also some interesting issues associated with ingesting certain items that cause people to develop specific allergies. For instance, most individuals do not develop an allergy to latex from coming into contact with products that contain nonpolymerized rubber material (latex). Instead, it is likely the latex allergy developed from ingesting bananas, which cross-react with latex. While a person may or may not have issues with eating bananas, he or she may develop IgE antibodies that can bind to latex, so that exposure to some rubber products containing the nonpolymerized rubber material causes allergic manifestations. In some cases, the IgE response can even result in more severe reactions, including anaphylaxis. However, sometimes the reaction to latex, which results in a contact rash, is not due to an IgE-mediated allergy reaction, but due to a type 4 cell-mediated hypersensitivity (which we will discuss in a future column).

Shrimp allergy poses another interesting issue of allergic exposure. Many people with a shrimp allergy have an allergic reaction the first time they eat shrimp. This is contradictory to the established opinion about how an immune response occurs, particularly how allergic immunity develops. According to established opinion, after an initial exposure to an allergen, the immune system perceives there is a parasitic attack, so it is driven into IgE production. The IgE finds its way to the mast cells in the body, and upon re-exposure to the allergen, an allergic reaction occurs. It follows that one should never have an allergic reaction to something to which one has had no prior exposure. Hence, there seemed to be a “shrimp paradox.”

It was eventually discovered that dust mites live off the skin and debris individuals are constantly shedding from their bodies. And, this detritus mainly collects in beds since people spend a third of their lives there. During sleep, individuals are constantly ingesting dust mites (as well as other things), which are arthropods related to shrimp. Indeed, the muscle proteins in dust mites are almost identical to those in shrimp. Therefore, people with shrimp allergy can develop IgE antibodies to dust mite muscle proteins (the sensitizing event), and upon first exposure to shrimp muscle proteins have an allergic reaction or even anaphylaxis. Therefore, there is no true paradox; there was a prior exposure even though it was not to the provocative food.

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

TERRY O. HARVILLE, MD, PhD, is medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences and a consultant for immunodeficiencies, autoimmunities and transplantation.
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.
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Simple, convenient, and ready-to-use, so you can get back to everyday living

Choose when and where you infuse
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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.

Visit Hizentra.com or ask your doctor about Hizentra prefilled syringes.

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.
HIZENTRA®, Immune Globulin Subcutaneous (Human), 20% Liquid
Initial U.S. Approval: 2010

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

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

INDICATIONS AND USAGE
HIZENTRA is indicated for:
- Treatment of primary immunodeficiency (PI) in adults and pediatric patients 2 years and older.
- Maintenance therapy in adults with chronic inflammatory demyelinating polyneuropathy (CIDP) to prevent relapse of neuromuscular disability and impairment.
- Limitation of Use: Maintenance therapy in CIDP has been systematically studied for 6 months and for a further 12 months in a follow-up study. Continued maintenance beyond these periods should be individualized based on patient response and need for continued therapy.

For subcutaneous infusion only.

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

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Combating Depression During the Pandemic

By Erika Lawrence, PhD, LCP

The COVID-19 pandemic has made life more challenging for all of us. Yet, while we all know the devastating impact it has had on physical and financial health, less attention has been paid to the widespread influence it has had on our mental health. Rates of depression and anxiety have skyrocketed since we began sheltering in place.

Staying home leads to social isolation and loneliness, which place us at greater risk for depression. But, those with immune disorders are at particular risk for depression during this pandemic since you have to take more precaution than others to avoid exposure to the virus. For instance, you must be more careful when making contact with others, allowing people into your home and where you go when you leave the house. All these extra safeguards generate an increased risk of social isolation and loneliness, which often leads to depression.

Depression can take many forms. It can cause us to feel down all the time, have little energy or motivation to do anything, feel hopeless or no longer find joy in activities that used to be pleasurable. So, since we can’t change reality, what can we do to combat depression? Here are some tips:

1) Engage in activities that gave you pleasure in the past. There is a lot of evidence that doing things we enjoy alleviates depression. Right now, you may not think there is anything you enjoy. What kinds of activities did you like in the past? Think back to your childhood. Jigsaw puzzles? Board games? Card games? Coloring? (There are some great adult coloring books.) Cooking? Baking? Drawing? Yoga? Meditation? Gardening? Walks near the beach or in the woods? Reading or listening to a good book? Sitting somewhere with a beautiful view? Try something new each day until you find an activity you enjoy. Spend just 15 minutes doing this to start.

2) Engage in activities that give you a sense of “mastery.” Similarly, there is a lot of evidence that doing things we are good at also helps alleviate depression. Think back. Are there activities you were good at in the past or are still proficient in? Did you ever play an instrument? Write? Cook or bake? Sew? Perhaps the same things you enjoy (see the list above) are the ones you are good at. Again, just commit to 15 minutes a day. Try different things until you find one.

3) Increase physical activity. Physical activity can work wonders for depression. This doesn’t mean you have to start a major cardio program. Start small by taking a 15-minute to 20-minute walk, trying a beginning yoga class online or riding your bike around the block. It may mean straightening up a room or gardening. Anything that gets you moving a little more than you did yesterday is added physical activity.

4) Increase social engagement. We all miss seeing people in person, and many of us are yearning for simple physical contact with others. However, there are other ways to connect. FaceTime and Zoom video calls make seeing others easy. You can ask friends to come by your house and speak to you through the door. You can invite friends over to sit in your backyard, keeping at least 6 feet distance from each other while wearing masks. These face-to-face interactions (including FaceTime/Zoom interactions) are critical to combating loneliness and depression. Sometimes you may not be able to participate in a conversation due to illness (coughing) or concern about your appearance. It’s OK to just be on the call and let the other person talk. Or, do an activity together. Again, face-to-face contact, even 5 minutes a day, makes a big difference.

5) Utilize teletherapy. Most insurance companies are covering the entire cost of teletherapy right now, so take advantage of it. There are also a lot of free on-call services for teletherapy. You need not commit to long-term therapy. Perhaps one or two meetings will help you start getting involved in the strategies above. These resources are readily available and affordable, so try them to see if they help.

Ample evidence shows these strategies help combat new, low-level symptoms of depression. If your depression was a concern before COVID-19, if your depression appears more severe than this, or if these strategies do not work, contact a therapist for a more involved approach to treating depression and/or a psychiatrist to discuss the possibility of trying an antidepressant.

The bottom line: With depression more prevalent than ever right now, taking care of our mental health is extremely important.

Erika Lawrence, PhD, LCP, is director of translational science at The Family Institute at Northwestern University, Evanston, Ill.
The CVID-Autoimmune Connection

By Michelle Greer, RN, and Marc Goldstein, MD

**COMMON VARIABLE** immuno-deficiency (CVID), named because it is the most common primary immune deficiency disease, results from a defect in the immune system that causes frequent infections. And, in many CVID patients, autoimmune disorders are a frequent comorbidity. Although incidence rates vary, it is estimated approximately 20 percent to 25 percent of people with CVID also have an autoimmune condition. In some, the autoimmune disorders are diagnosed first, whereas in others, CVID manifests first. And, in some cases, CVID is diagnosed at the time of the autoimmune diagnosis before symptoms of CVID are present. For example, the development of an autoimmune disorder at an unusually early age or due to multi-organ involvement should lead to an evaluation of underlying CVID even before recurrent infections are apparent.

CVID is estimated to affect between one in 25,000 people and one in 50,000 people worldwide, although the prevalence can vary across different populations. The cause is generally unknown, although in a small population, a genetic mutation and/or environmental factors are likely responsible. B cells responsible for producing antibodies are affected, resulting in low antibody (immunoglobulin [IgG]) levels. People with CVID usually have at least two specific antibody deficiencies. They also lack the ability to mount a response to vaccinations, which results in the inability to effectively fight off infections. However, the type, frequency and severity vary from person to person.

**Figure 1. Autoimmunity Causes**

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**Types of Autoimmunity Associated with CVID and Their Treatments**

Immune dysregulation caused by CVID can lead to a variety of autoimmune disorders not necessarily influenced by age or gender. The leading autoimmune disorders include hematologic cytopenias (low white blood cells, low red blood cells and/or low platelets), endocrine disorders, inflammatory bowel disease (IBD), systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), lymphocytic pneumonitis and lymphoproliferative disorders, including lymphoma and granuloma formation.

**Cytopenias.** Autoimmune thrombocytopenia purpura, one of the most common cytopenias, occurs when platelets are destroyed by the person’s immune system. Platelets play an important role in blood clotting, and when the platelet count becomes dangerously low, bleeding results. Another frequently associated cytopenia is autoimmune hemolytic anemia in which red blood cells are affected. An example of this is Evans syndrome, a rare disease characterized by the simultaneous or sequential development of autoimmune hemolytic anemia and immune thrombocytopenia and/or immune neutropenia.

Cytopenias are typically treated with steroids. However, if blood levels become significantly low, high-dose intravenous immune globulin (IVIG) might also be prescribed intermittently. While CVID patients may already be receiving either IVIG or subcutaneous IG therapy, dosing for autoimmune conditions is generally much higher than for CVID, especially at first.
For instance, dosing for autoimmune conditions can range from 1 gram to 2 grams per kilogram of body weight, whereas dosing for immune deficiencies is typically 0.4 grams per kilogram. In some cases, Rituxan (rituximab) and other immunosuppressants such as azathioprine (Imuran, Azasan) and 6-mercaptopurine (6-MP) (Purixan, Purinethol) or mycophenolate (Cellcept) and splenectomy have been used in refractory (treatment-resistant) cases.

Joint manifestations. Joint conditions resembling RA and SLE are other autoimmune conditions associated with CVID. With these, damage and inflammation occurs in the joints, specifically the synovium (a membrane that provides protection and lubrication of the joints), especially during movement. A wide range of oral, subcutaneous and intravenous treatments are available for RA and SLE. However, for many who have RA and SLE prior to developing CVID, it may be unclear whether immunosuppressant treatment for the primary disease produces low IgG levels or if the autoimmune disease and the hypogammaglobulinemia arise from the same immune defect.

Immune dysregulation caused by CVID can lead to a variety of autoimmune disorders not necessarily influenced by age or gender.

Other autoimmune conditions. Other conditions associated with CVID include IBD, juvenile idiopathic arthritis, Sjogren’s syndrome, vitiligo, reactive arthritis and autoimmune hepatitis. IBD (chronic inflammation of the digestive tract) is the most common, occurring in 6 percent to 10 percent of CVID patients. Intestinal inflammation can be difficult to treat, and prolonged oral steroid treatment is not ideal given the risk of immunosuppression. Other treatments for IBD such as oral budesonide (a second-generation steroid that allows local, selective treatment of the gastrointestinal tract and the liver, minimizing systemic exposure), immunosuppressants azathioprine and 6-MP, and immunomodulatory monoclonal antibodies such as infliximab, vedolizumab and ustekinumab can be used with various results.

Awareness Is Key

Autoimmune disorders occur in approximately one-quarter of CVID patients. Therefore, awareness of those associated with CVID is helpful for monitoring patients, particularly if there are new signs and symptoms consistent with autoimmunity.

MICHELLE GREER, RN, is senior vice president of sales for Nufactor, a Specialty Infusion Company. MARC GOLDSTEIN, MD, is chief of allergy and immunology at Pennsylvania Hospital at the University of Pennsylvania and associate professor of clinical medicine at Drexel University College of Medicine.

References
IN THE NEWS

Medicines
FDA Grants Orphan-Drug Designation to Privigen for Systemic Sclerosis

The U.S. Food and Drug Administration (FDA) has granted Privigen (immune globulin intravenous [human], 10% liquid) orphan-drug designation as an investigational therapy to treat systemic sclerosis (SSc), a chronic and potentially life-threatening autoimmune disorder characterized by a buildup of scar tissue (fibrosis) in the skin and other organs affecting approximately 100,000 people in the U.S. There are currently no FDA-approved, disease-modifying treatments to stop or reverse the overall course of the disease.

CSL Behring recently initiated a Phase II clinical trial to evaluate the safety and efficacy of Privigen to treat adults with SSc, and in October 2019 received fast track designation for the clinical development of Privigen to treat serious pulmonary, skin and musculoskeletal manifestations resulting from this condition. FDA confers fast track designation to aid and expedite the development and review of drugs that show promise in treating a serious or life-threatening disease and address an unmet medical need.

“CSL Behring is driven by our promise to develop and deliver innovative therapies for patients with the highest unmet need,” said Mittie Doyle, vice president, research and development, immunology and neurology therapeutic area at CSL Behring. “Receiving orphan-drug designation for Privigen as an investigational SSc therapy is an important milestone in our quest to address the devastating impact of systemic sclerosis.”


Research
PI Patients Treated at Home with SCIG Versus IVIG Have Similar Health-Related Quality of Life Scores

A recent study shows patients with primary immunodeficiency disease (PI) on home intravenous immune globulin (IVIG) versus home subcutaneous IG (SCIG) have similar composite health-related quality of life scores as measured by short form 36 (SF-36), an indicator of overall health status.

In the study, SF-36 surveys were administered by a specialty pharmacy to 630 PI patients receiving home SCIG and home IVIG at baseline and then every three months between 2014 and 2016. Researchers found patients receiving SCIG reported statistically significant higher energy fatigue scores but lower perceived role limitations due to physical health scores. However, these differences were observed only in patients older than 36 years. And, there were no differences in the composite SF-36 score for patients receiving SCIG versus IVIG. In addition, all IG-naive patients improved their health-related quality of life scores, but a larger improvement was seen in those initiating SCIG versus IVIG. According to the researchers, while initiating IG replacement with SCIG may result in more health-related quality of life improvement compared with IVIG, personal preferences should also be considered.

**Campaign**

**FDA Campaign Designed to Help Consumers Use New Food Label**

The U.S. Food and Drug Administration (FDA) has launched a campaign to help consumers use the new Nutrition Facts label that appears on packaged foods to maintain healthy dietary practices. The label was finalized in May 2016, but most manufacturers with $10 million or more in annual food sales had until January 2020 to begin displaying it on their products. Manufacturers with less than $10 million in annual food sales have until Jan. 1, 2021, to start displaying the new label, although many already have.

The new campaign is part of FDA’s comprehensive, multi-year Nutrition Innovation Strategy, which is designed to empower consumers with information about healthy food choices and to facilitate industry innovation toward healthier foods. The campaign’s tagline “What’s In It For You?” is designed to reach the general public and also focuses on consumers at increased risk of nutrition-related chronic diseases, including obesity. Included in the campaign are videos and educational materials of food products modeling their new looks, including on a fashion runway, after receiving a makeover.

The new label is the first redesign of the Nutrition Facts in more than 20 years, and its design is based on updated scientific information, including the link between diet and chronic diseases such as obesity and heart disease. It is most distinguishable by its bold listings for serving sizes and calorie counts. Additional changes include new required listings for added sugars, vitamin D and potassium, and a dual column version of the label for food packages that contain two to three servings that can be reasonably consumed at one time. On the dual label, one column lists the nutritional facts related to a single serving, and the other column lists nutritional facts for the contents of the entire package.

Serving sizes have also been updated to reflect that the amount of food and beverages people eat and drink has changed.

The campaign is intended to educate consumers, as well as healthcare professionals, teachers, dietitians and community leaders. Information about the campaign can be accessed at [www.fda.gov/food/nutrition-education-resources-materials/new-nutrition-facts-label](http://www.fda.gov/food/nutrition-education-resources-materials/new-nutrition-facts-label).

**Correction**

**Plasma Donations in the U.S. Can Be Made Only Two Times Per Week, Not Three**

In the February-March 2020 issue of *IG Living*, it was incorrectly stated in the article “Managing Care During Immune Globulin Product Shortages” (pp.20-23) that plasma may be donated up to three times a week, with a 24-hour break between donations. This is not correct. In the U.S., plasma can be donated up to two times a week (a maximum of 104 times per year), with at least 48 hours between donations.

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**Nutrition Facts**

8 servings per container

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<tr>
<td><strong>% Daily Value</strong>*</td>
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<tr>
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<tr>
<td>Potassium</td>
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* The % Daily Value (DV) tells you how much a nutrient in a serving of food contributes to a daily diet, 2,000 calories a day is used for general nutrition advice.
IVIG Combined with IVMP Raises Platelet Counts Faster Than IVIG Alone in ITP Pediatric Patients

A new study shows a rapid response to intravenous immune globulin (IVIG) with and without steroids, as well as provides evidence to support the use of IV methylprednisolone (IVMP) with IVIG in pediatric immune thrombocytopenia (ITP) patients.

In the randomized, double-blind, placebo-controlled study, 32 children ages 1 year to 17 years old with ITP and platelet counts (PC) less than 20 x 10^9/L were given either an IV placebo (18) or IVMP 30 mg/kg (14), both given over one hour, followed in both cases by IVIG (Gamunex 10%) 1 g/kg over two hours to three hours. Eight hours after initiating therapy, 55 percent of all children had a PC less than or equal to 20 x 10^9/L (no group difference). But, at 24 hours, mean PCs were 76.9 x 10^9/L for the IVIG group versus 55 x 10^9/L for the placebo group. No patient experienced severe bleeding or unexpected severe adverse events. And, there were statistically fewer IVIG-related headaches in the group receiving combination therapy.


Potential Biomarker Test Identifies CFS in Patients

Researchers at Stanford University School of Medicine have come a step closer to developing a possible diagnostic test for chronic fatigue syndrome (CFS); however, further research is needed before an actual test can be developed. In the pilot study, researchers analyzed blood samples from 20 people who were healthy and 20 people diagnosed with CFS using a nanoelectronic assay, a test that measures changes in tiny amounts of energy as a proxy for the health of immune cells and blood plasma. They then stressed the blood samples using salt and compared the responses. Results showed all CFS patients’ blood samples created a clear spike, while the healthy controls’ blood samples remained relatively stable. According to the researchers, this potential biomarker test correctly identified those who were ill. “We don’t know exactly why the cells and plasma are acting this way, or even what they’re doing,” said Ron Davis, a professor of biochemistry and genetics who co-led the study. But, “we clearly see a difference in the way healthy and chronic fatigue syndrome immune cells process stress.”

Other experts caution, however, there is still a long way to go before a biomarker is found that can establish a CFS diagnosis and distinguish it from other conditions with similar symptoms. In addition, this research was unable to solve two key issues, according to Simon Wessely, chair of psychiatry at King’s College London’s Institute of Psychiatry, Psychology and Neuroscience, who has worked with CFS patients for many years. “The [first] issue is, can any biomarker distinguish CFS patients from those with other fatiguing illnesses? And, second, is it measuring the cause and not the consequence of illness?” said Wessely. “This study does not provide any evidence that either has finally been achieved.”

Research

Etanercept Has No Benefit for Reducing IVIG Resistance in KD Patients

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.


Research

Study Finds SCIG Maintains Stable Disease Activity in Myasthenia Gravis Patients While Transitioning from IVIG Therapy

In 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 OMG 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.

The Safety of IG Therapy

While IG therapy is now safer than ever due to manufacturing processes and improvements in IG production, many considerations must be taken into account when deciding product choice and infusion setting.

By Abbie Cornett
**IS IMMUNE GLOBULIN (IG) therapy safe?** As the patient advocate for *IG Living* magazine, I am asked this question frequently by both new and existing patients. My first response is always, “Yes!” But, after saying that, I feel it is important to elaborate. That means explaining what IG is, how it is produced, who is treated with it, what types of products are available and what the side effects are.

**What Is IG?**

IG is prescribed to treat or prevent serious diseases or medical conditions for which no acceptable drug alternative is available. It is a sterile solution made from antibodies in human blood plasma. Unlike traditional pharmaceutical products that rely on a chemical process, plasma-based therapies are derived exclusively from proteins found in human blood plasma. Plasma is the light yellow portion of blood that is composed of water, salts and proteins that performs a variety of functions in the body, including clotting and fighting disease. It makes up 55 percent of a person’s total blood volume.

IG was first used almost 70 years ago after a medical discovery by Ogden Bruton, MD, chief of pediatrics at Walter Reed Army Hospital. Dr. Bruton successfully treated an 8-year-old boy diagnosed with agammaglobulinemia who suffered repeated bouts of pneumonia with regular intramuscular (IM) injections of human plasma-derived IG.

Although IM injections were successful at treating immune disorders, they were very painful and could only be given in a limited dose. It wasn’t until the U.S. Food and Drug Administration (FDA) approved the first intravenous IG (IVIG) product that IG was regularly used for treatment. Since then, the demand for plasma protein therapies used to treat rare and chronic conditions has grown at a tremendous rate.

**Safeguarding Plasma Collection**

Patients are often surprised to learn it takes between six months and 12 months from the time plasma is collected until it is manufactured into an available IG product. Production takes this long due to the number of steps in place ensuring the safety of both the donor and the recipient.

In the United States, the licensing of plasma collection facilities is regulated by FDA and the voluntary standards of the Plasma Protein Therapeutics Association. U.S. blood and plasma are collected, processed and distributed by private industry that is regulated by FDA under two national laws: The Public Health Service Act and the Federal Food, Drug and Cosmetic Act.

Any facility that collects source plasma must demonstrate in its license application submitted to FDA that it can produce a product that is both safe and effective. The application states the collection center must have the appropriate laboratory tests and equipment, donor safety measures, appropriate manufacturing methods, data establishing stability of the product through the dating period, specimens of the labels, and the address of each location involved in the manufacture of the product. After FDA reviews the license application, it inspects the facility to observe manufacturing, and decides whether the facility is ready for licensure. FDA routinely inspects licensed facilities every two years or “for cause” to ensure they meet applicable regulations and standards.1

**Screening of Donors and Units of Donated Plasma**

The first step in producing IG takes place at a donation center. To ensure the safety of the plasma collected, donors must go through a detailed screening process to determine they are healthy enough to donate. Once donors are screened, their plasma is collected via a specialized process called plasmapheresis, which separates the plasma from the red blood cells and other components. After separation, the red blood cells are returned to the donor.2

Patients are often surprised to learn it takes between six months and 12 months from the time plasma is collected until it is manufactured into an available IG product.

Once plasma is collected, it is stored while it undergoes a thorough screening process to ensure it is free of viruses. The plasma is then pooled with donations from thousands of other donors,3 and the pooled product is put through a process called fractionation. The fractionation process isolates and purifies the therapeutic proteins found in the plasma, including those used in the production of IG.
FDA mandates that plasma pools are derived from a minimum of 1,000 donors; however, a batch of IG can include plasma from approximately 15,000 donors. As a further safety precaution, FDA requires all IG products sold in the United States to be derived solely from U.S. donor plasma, although the final IG product may be manufactured in FDA-approved facilities outside of the United States.4

Who Is Treated with IG?

IG was originally prescribed as antibody replacement therapy for patients with primary immunodeficiency diseases (PI). Since then, its use has grown at a tremendous rate, and it is now prescribed to treat and prevent multiple other conditions. Currently, IG is approved by FDA to treat:5

- Chronic inflammatory demyelinating polyneuropathy
- Chronic lymphocytic leukemia
- Immune thrombocytopenic purpura
- Infections following bone marrow transplants
- Kawasaki disease
- Multifocal motor neuropathy
- PI

IG is also prescribed to treat several autoimmune and neurological diseases; however, these diseases are not approved by FDA for IG treatment. These conditions include but are not limited to:1

- Guillain-Barré syndrome
- Lupus
- Polymyositis and dermatomyositis
- Multiple sclerosis
- Myasthenia gravis

Site Safety for IG Administration

As the number of approved IG products continues to grow, so have the delivery methods and infusion site options. In the past, IG was only infused intravenously, and it was administered only in a hospital setting because of the risk of an adverse reaction. Today, patients can infuse IG intravenously or subcutaneously.

Several brands of IG are available in the United States, all of which are essentially therapeutically equivalent to one another. However, this does not mean they are pharmaceutically equivalent. When determining which IG product is best for a patient, it is important to understand the differences between them. Factors that can affect how well a patient tolerates a product include its stabilizers, osmolarity (see What Is Osmolarity?), sodium, IgA content and concentration. Selecting a product suited to a patient’s health history is a critical step in both optimizing care and ensuring patient safety. This means the prescribing physician and infusion nurses must be well-trained and fully knowledgeable about the different dosing requirements. They must also understand that no two IG products are the same, and how each product differs in preparation.

As mentioned previously, IG can be administered in two ways: intravenously (IVIG) and subcutaneously (SCIG). Unlike IVIG, which is infused into a vein, SCIG is infused by slowly injecting the medicine into fatty tissue just underneath the skin. When deciding which route of administration is best, it is important to understand how the route of administration may also influence the risk of adverse reactions and increase the probability of exacerbating comorbid conditions. Patients’ medical histories must also be taken into consideration, as there are specific comorbid conditions that can be exacerbated by components of some products.6

Site-of-Care Considerations

IG patients have safe site-of-care options: clinical settings (doctor’s office, hospital, infusion clinic) or at home. When choosing the site of care, it is important to remember the administration of an IG product is a complex process that involves the consideration of multiple factors.

Most importantly, providers must determine which environment is safest for each patient. Patients who need a higher level of care due to comorbidities such as diabetes and heart disease or who have experienced adverse side effects such as anaphylactic reactions, hypotension, seizures, pulmonary

What Is Osmolarity?4

Osmolarity is the concentration of osmotically active particles in solution, which may be quantitatively expressed in osmoles of solute per liter of solution. Most of the osmolality of IVIG, IV/SCIG and SCIG products are within the range of physiologic osmolality of approximately 290 mOsm/kg. Products that deviate substantially from physiologic osmolality levels may put the patient at risk for various infusion-related adverse effects such as thrombotic events and aseptic meningitis, particularly in elderly or neonatal patients, patients with cardiometabolic impairment, and patients with renal dysfunction.
edema or aseptic meningitis should be infused in a clinical setting. Beyond the immediate safety concerns, a clinical setting allows patients to have more one-on-one contact with doctors and nurses, which can be very important for those who have been chronically ill for a long time or who run the risk of declining health or infection. On the other hand, if patients don’t require a higher level of care, home-based treatments are not only convenient, but they can reduce patients’ exposure to pathogens in a clinical setting — a factor particularly important for PI patients.

**Potential Side Effects**

Like most medications, both IVIG and SCIG can have side effects. Most occur during or immediately following the infusion. The good news is most side effects can either be treated or eliminated. With IVIG, up to half of all patients experience at least one adverse side effect such as headache, low-grade fever, aching muscles or joints, and rashes. These are especially likely to occur if patients are not receiving IVIG on a regular basis and/or if they are receiving higher doses of IVIG. When these side effects occur during the infusion, the infusion may be slowed down or stopped.⁷ IVIG patients also have a greater risk of thrombosis because infusions are administered through the veins. Most side effects are usually minor, while more serious side effects are rare and include anaphylactic shock, aseptic meningitis or blood clots. Should patients develop an allergic reaction, healthcare providers are trained to handle it.

Patients can reduce the side effects associated with IVIG with premedication such as acetaminophen or antihistamines, and corticosteroids are another option doctors might consider. Proper hydration prior to and during an infusion is also important to reduce side effects.

With SCIG, the severity of side effects is mostly reduced or eliminated since the medicine is absorbed by the body more slowly through fatty tissue, rather than in large doses entered directly into the circulatory system. The main side effects that occur with SCIG include headaches and local irritation (redness, swelling, itching, blanching) at the needle sites. Some reactions, especially for patients new to SCIG therapy, are expected, and most decrease with time once the body becomes accustomed to therapy. For patients bothered by reactions, applying ice or heat to the needle sites can help decrease some of the symptoms. Using a topical anesthetic cream 30 minutes to 60 minutes prior to starting the infusion also can be helpful.⁸

**Is IG Therapy Safe? Yes**

IG therapy has come a long way since its first use. Because of the multistep manufacturing process and improvements in production, IG products are safer today than ever before. Yet, it is important to remember every IG product has different properties, and there can be variations from batch to batch of the same product that can affect patient tolerability. The key to patients’ success and safety with an IG infusion is finding which product and setting is best suited to their individual needs.

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

**References**

Purchasing a **Medigap Policy**

Supplemental Medicare policies, known as Medigap, can help beneficiaries pay for what is not covered under Original Medicare; however, individuals need to research the many available plans to determine which is best for them.

By Leslie J. Vaughan, RPh

**THE U.S. HEALTHCARE** system has many complexities and moving parts. But, perhaps the most complex part is Medicare, which has multiple coverage components. Understanding how these components work together and how individuals can structure their own Medicare benefit can feel like a research project. Nevertheless, it’s important to know Medicare does not pay for all services and may only pay a portion of charges, which means a supplemental policy, known as a Medigap policy, is often necessary. Following is a summarization of Medigap coverage; however, it does not replace the need for individuals to research the best solution for them as they reach Medicare age.

**Medicare Coverage Choices**

To best understand what a Medigap policy does and doesn’t cover, it is important to have a basic knowledge of the components of Medicare and Medicare coverage choices. Generally, there are two coverage choices available: Original Medicare and a Medicare Advantage (MA) Plan (Part C).

Original Medicare consists of Medicare Part A (hospital insurance), Medicare Part B (medical insurance) and Medicare Part D (prescription insurance). Part A pays for inpatient hospital care, skilled nursing facilities, lab tests, surgery, hospice and home healthcare. Part B pays for many outpatient services, including doctor visits, outpatient care provided by other healthcare providers (i.e., physical therapy, home healthcare and medical equipment), some prescription drugs and some preventive services. Part D pays for most prescriptions. There are rules for and limits on how each of these components of Original Medicare will reimburse for care.

MA plans (Part C) are similar to HMO and PPO plans. MA plans are offered by traditional insurance providers and are required to cover all services offered under Parts A and B. Prescription drug coverage may be included or may be offered for an extra charge. MA plans may cover additional services not routinely covered by Original Medicare such as vision, dental, hearing aids, wellness plans (including gym memberships), transportation to doctor visits, over-the-counter drugs and adult day care services.

There are also other Medicare health plans such as Medicare cost plans, demonstration/pilot programs, Programs of All-Inclusive Care for the Elderly (PACE) and medication therapy management programs for complex health needs, which are described on the Medicare website (www.medicare.gov).

With either choice (Original or MA), a beneficiary will need to sign up for each of the components and will have different costs for each component of the plan.

**Medigap Policies**

Medigap policies are offered by private insurance companies and fill “gaps” in Original Medicare. Medigap plans only supplement Original Medicare, not MA plans. In fact, it is illegal for private insurance companies to sell a supplemental policy to someone with a MA plan, unless they are switching to Original Medicare. Some Medigap plans also have prescription drug coverage. In those instances, individuals would not need to also have a Medicare Part D prescription drug plan, but they would need to ensure the Medigap plan qualifies as having “credible drug coverage” to avoid paying a late enrollment penalty if it will replace a Medigap plan with a Part D plan in the future. A beneficiary must have both Parts A and B to purchase a Medigap plan.
Original Medicare does not pay for all healthcare services costs. Instead, Medigap plans pay for some of the remaining costs such as co-payments, coinsurance and some deductibles. For example, Medicare Part B may pay for 80 percent of the charges for a physician office visit. And, as long as Medicare pays the 80 percent, a Medigap plan will typically pay the remaining 20 percent. But, there are exceptions to what Medigap plans cover. Generally, they don’t cover long-term care, vision or dental care, hearing aids, eyeglasses or private duty nursing. In addition, if Medicare Parts A or B do not cover a service, Medigap plans generally won’t either. This is a key difference between Medigap plans and true secondary insurance. Some secondary insurance plans may cover services not covered by Medicare. However, prior authorization is generally required.

Three states (Massachusetts, Minnesota and Wisconsin) have different ways of standardizing benefits than the other states. The following information is specific to the remaining states.

Insurance companies may offer up to 10 different types of Medigap policies, but all must follow state and federal laws designed to protect beneficiaries. Each policy is labeled with a letter (A, B, C, D, F, G, K, L, M and N). A policy labeled with the same letter must offer the same benefits regardless of which company offers it. However, prices may vary by company. And, all policies offer the same basic benefits, but they can offer additional benefits beyond the basic ones.

Similar to Medicare plans, there are certain time frames for enrollment. If a beneficiary is choosing Original Medicare, the best time to enroll in a Medigap policy is during the open enrollment period. Under federal law, beneficiaries have a six-month enrollment window that starts the month they are 65 years or older and enrolled in Medicare Part B. During this period, if a beneficiary chooses to purchase a policy, a company must sell the policy at the best available rate without consideration of health status. The company can consider age, gender, marital status, location of residence and whether the beneficiary smokes to determine the rate, but it can’t consider preexisting conditions if purchased during this time frame. Purchasing outside of this time frame may result in higher premiums and exclusion of coverage for preexisting conditions for six months.

Prior to 2020, some Medigap plans included coverage for the Part B deductible. However, as of Jan. 1, 2020, new Medicare beneficiaries are not eligible to purchase these plans. But, Medigap plans may have coverage for the following benefits after the deductible has been paid. In some cases, the Medigap plan will cover only a percentage of the remaining charges for services. All plans must offer the following basic benefits:

- Part A coinsurance and hospital costs up to an additional 365 days after Medicare benefits are depleted
- Part B coinsurance or co-payment (plans may have a reduced percentage of coverage)
- Coverage for three pints of blood annually (plans may have a reduced percentage of coverage)
- Hospice coinsurance

Plans may offer enhanced benefits and typically charge a higher premium for those. Enhanced benefits may include:

- Skilled nursing facility care coinsurance
- Part A deductible
- Part B deductible (plans C and F pay only for those eligible prior to Jan. 1, 2020)
- Part B excess charges (nonparticipating doctors can charge fees above the Medicare-approved amounts for services)
- Emergency care outside of the U.S.
- Out-of-pocket limit
- At-home recovery
- Preventive care not covered by Medicare

Understanding the Options

Medicare has many resources to help beneficiaries understand the options available to them as they become Medicare-eligible and as coverage and other policies related to Medicare change each year. The Medicare website (www.medicare.gov) provides detailed information on all components of Medicare, including Medigap policies. Medicare also publishes a helpful booklet titled Medicare and You, which is updated annually and can be downloaded from Medicare’s website or delivered by mail. There are also options to speak with someone at Medicare by dialing (800) MEDICARE (800-633-4227).

In summary, careful evaluation of all the components Medicare offers is necessary to determine which options are best for each beneficiary. If choosing Original Medicare, a Medigap policy will help offset many of the charges not covered.

LESLIE J. VAUGHAN, RPh, is chief operations officer of Nufactor, a Specialty Infusion Company.
Getting The Most Out Of Your Disability

Those who need assistance to accommodate their disabilities should not be reluctant to take advantage of all the many programs and services available.

By Surayyah Morris, PharmD

HAVE YOU EVER looked at (seemingly) perfectly healthy people and wished just for a moment you could enjoy one day in their bodies? No handful of pills in the morning, no weekly infusions and no physical limitations? They can get up and get ready in a fraction of the time it takes you. Their lives aren’t planned around doctor appointments. And, they don’t think twice about accommodations they’ll need when going to a store, park, school or on vacation.

Although your life requires a little extra work, there are resources for those with chronic illnesses who have disabilities and/or are receiving disability benefits as a result. So if the resources are there, why not take advantage of them! But, before we discuss how to get the most accommodation for your disability, I need you to understand one very simple rule: Always ask. The worst someone can tell you is “no,” as long as they are not violating any Americans with Disabilities Act laws. And, even if one person tells you “no,” others may be kind enough to go out of their way to accommodate you anyway. There may be modifications available that you would never have even imagined.

The Fun Stuff

When it’s time to play, make sure it’s fully enjoyable! If you want to visit the zoo or a state/national park, check to see about their guests with disabilities rates, service animal policies, sighted guides or sign language interpreters for sight and hearing limitations, rentable mobility devices for physical limitations, and accessibility maps to help navigate with ease. Contact the facility in advance to request specific material in a format accessible to you and also for special assistance requests.

If you’re more of an indoor person (hello, air conditioning) and the movies are your thing, they have something for you, too. Assistive listening devices, accessible seating, audio description and closed captioning devices can be provided to you. You may have to trade in a cup holder for use of one these devices, but the experience will be worth it. It also never hurts to ask about discounts for care companions who are joining you.

When staying at hotels or resorts, request a first floor room with accessibility and handicapped parking spaces nearby. These types of accommodations typically require at least one week to two weeks advance notice, so some planning is definitely necessary.

Traveling

Most know this already, but when traveling by air, you can book your flight with accommodations for assistance at the airport with a wheelchair, service animal, etc. Request to have your checked baggage fee waived and get assistance with your luggage if you are unable to manage it yourself. This is
especially helpful when traveling alone. Some airlines will even call you prior to your departure date to assist you with this and honor your request for a window/middle/aisle seat and/or extra leg room. Please remember not to be placed in emergency exit seating since you will need to be physically able to assist others in the event of an emergency. In addition, you get to board ahead of passengers who do not require accommodation.

You can choose to be specific with the type of disability and accommodation required. This information is printed on your ticket and must be honored. So, no more standing in long lines or having people question your service or support animal. Service animal relief areas are available, too.

If you decide not to use your requested accommodations, there is no penalty. If you forget to add these requests online, you can call and have a representative add them to your travel documents. Oh, and did I mention these services are free?

**Handicap Parking**

There are two types of handicap parking placards: temporary and permanent. The temporary permit (red) is valid for six months and must be renewed upon expiration if more time is needed. Typically, this is for people with short-term injuries or a temporary disability. There is also a permanent placard (blue) that lasts for at least three years or more. These are especially useful for people who have mobility issues and trouble walking long distances. Your provider has to check off one or more of your limitations supporting why you need a handicap placard and return the form to you to take to your local department of motor vehicles (DMV). The DMV takes the gold medal for the establishment with the longest wait times, so be sure to locate the correct counter where parking placards are processed, or politely ask a representative to direct you to it. This should drastically cut down your wait time. The fee is a minimal $5 in most states, clearly worth the convenience of a close parking space. In addition to the convenience of closer parking, some events, beaches, meters and facilities will waive the parking fees for those with handicap placards.

**Medical Devices**

If haste isn’t a factor in obtaining a supportive aid such as a wheelchair or any durable medical equipment, take the time to review your insurance policy requirements to see if the device or equipment is covered in any amount by your insurance. The process may seem tedious, requiring doctors’ documentation and physical assessments, but the cost savings is beneficial. If items aren’t immediately covered, see if your insurance offers reimbursement for items you buy out of pocket for. You have insurance and need to get everything you can to reduce stress and make managing your condition simpler. If you don’t have any luck with insurance coverage, try a secondhand medical supply store, local nonprofit organization or even Goodwill (be sure to have the items checked for safety and cleanliness if you go this route).

If you are an eligible Supplemental Nutrition Assistance Program or Medicaid recipient, a free phone is available from Assurance wireless or Q Link wireless. These companies may also provide a free phone based on income or veteran status. The catch? Use the phone! This should be easy for most, just pay attention to data, call and messaging limits.

**Public Transportation**

Need to meet with a friend for a lunch date? Maybe you need a ride to class on one of your off days. Or, maybe you have several doctor appointments, or simply just don’t feel like driving? Many cities with public transportation systems also have a program with disability access resources to provide transportation for customers who are not able to safely and efficiently navigate public transportation platforms such as the train or bus. This typically includes door-to-door transportation in a smaller more-accessible vehicle such as a van.

If using general public transportation causes extenuating stress, is too difficult to travel alone, moves too fast for your pace or isn’t as accessible as you need, ride services can help you get where you need to go with a simple application and, yes, you guessed it, a doctor’s cosign. There may also be a brief interview to assess your physical condition and your
need for this service (they won’t make you jump through hoops). With legitimate concerns for riding with general public transportation, you will be accepted into an access transportation program. Once accepted, these services will provide transportation to and from any address within specified regions. This means you have valet service from home to doctor appointments or the mall or zoo. The best part, fares are typically reduced when using these services. And, if you require the assistance of a personal care attendant, this person can travel with you for free. If you don’t require such specialized service, you may qualify for reduced fares based on your ability to navigate the public transportation platforms. There are no limits to where you may travel within the outlined area for regular transportation services. Some preparation for your trip will include having to set an appointment time for pick-up/drop-off at your desired location and exact change for paying your fare. If you decide not to use these services, you are still welcome to use general public transportation as desired, often at reduced fares.

Testing Accommodations
School is seldom a favorite place to be. It can be embarrassing and difficult to keep up with the general student body, but thankfully there is help for students who need a hand. In grade school, there are individual education plans (IEP) that allows students to work at their own pace and receive help and accommodations away from the general student body. This IEP is confidential between teachers and guidance counselors and the student. In colleges, there are a variety of resources for students. In-class accommodations can include seating in a particular spot in the classroom, or even having a notetaker to help with writing or recording notes. Testing accommodations range from extended testing time to a separate quiet testing environment. Students may also request extended time to complete assignments and projects. Locate your institution’s disability/accommodation resource center to inquire about available accommodations to help you get the A grade you deserve. Some institutions also offer testing services to examine you for anxiety, attention deficit disorder or learning disabilities. These services, however, may come with a fee. If you already have a diagnosis, you should be able to provide your most recent information from your provider to be allowed accommodations as requested.

Work Accommodations
For those in the disabled community who are fortunate enough to be employed, there are more specific accommodations that may be honored to help make your workday comfortable. Reasonable accommodations can be requested, as long as they are necessary to do your job without interrupting the business of others. At work, your request can be more specific such as a chair if you need to sit, an anti-fatigue mat if you are standing or a headset instead of a handheld phone. These are just a few suggestions, but you can request what you need depending on your work environment. Your employer may also reassign you to an equally important position that allows you to still complete your assigned work within your expertise. Support from your doctor may be requested, but it does not have to specifically identify your condition, just your requested accommodation as it relates to your limitations.

No Need to Suffer!
It is easy to feel embarrassed or ashamed when needing help from others and being dependent, especially in a society that focuses on how well we can do for ourselves. It is the duty of the able to help the unable in times of need, and there are many great resources to assist in making sure everyone is as comfortable as possible. If you’re going to have a chronic condition, you might as well use it to your advantage, right? Why suffer if you don’t have to? There isn’t a way around the unfortunate hand you were dealt, but there is a way to play your cards to your advantage.

But, please do not abuse the system to gain any of the aforementioned benefits. They are available for people who actually need them. When abuse of the system occurs, it limits available resources for assisting many disabled or less-fortunate people.

SURAYYAH MORRIS, PharmD, is an IG patient from Central Florida. As a medication therapy management and pain management specialty pharmacist, she enjoys supporting patients with chronic pain and chronic conditions to help find balance and improve quality of life.
BECOMING STRONGER THROUGH ILLNESS

Some sage advice might help those living with chronic illness find inspiration from their physical challenges and purpose in their lives.

By Matthew D. Hansen, DPT, MPT, BSPTS

THERE ARE MANY philosophies of life. In fact, it would probably be fair to say there are as many philosophies about life as there are people on Earth, because even though many of us fall into one philosophical and/or religious belief camp or another, we’ve all thought about our particular purpose and place in the universe.

Many people believe in a higher power but describe that power very differently; others don’t believe in any higher power at all. Some believe existence ends at death, while others are certain life extends beyond the grave. I’m confident most of us believe we are here for a purpose (whether it be short-term or of eternal consequences); still, some are convinced that we are here with no purpose at all.

The purpose of my commentary is not to delve into philosophy or religion, nor to try to convince readers of my own convictions or how I came to them, but to offer some perspective through my interactions with family, friends and patients who have had to deal with significant illness. My hope is the insights are provocative to many, and inspirational to those who need them the most.

Why Are We Here?

Just kidding! I promised provocative, didn’t I? Rather than asking why we are here, let’s start with the premise that we are, although some philosophers would even argue that.

As far as we know, we didn’t personally choose which physical illnesses we would have to face here on Earth, any more than someone starving and in poverty in a third-world country chose to be born into their condition rather than into a privileged ZIP code with beachfront property. Of course, a series of poor conscious decisions about our health can lead to bad consequences, but that’s different than hearing from a doctor that you have an autoimmune disease, primary immunodeficiency disease or other condition that may not have been influenced at all by one’s decisions.

So, some will ask, why was I the “lucky” one who got a chronic illness? Actually, you’re not. At least you aren’t “the one” who won — or lost — the lottery and acquired a chronic illness. According to the National Health Council, about 50 percent of adults in the U.S. have a chronic health condition, almost a third of the population is now living with multiple
chronic conditions and approximately 70 percent of U.S. deaths are due to chronic diseases (heart disease, cancer and stroke accounting for more than half of all deaths each year). Now, if you have had all three, something exceptional might be going on.

I don’t mean to sound insensitive, but I am trying to make the point that we all will have to deal with pretty significant physical challenges either now, later or now and later. That’s part of being mortal, and death is part of everyone’s experience. Of course, the mission of healthcare is to help avoid premature death when possible and to improve quality of life regardless of how much time we have.

Accept and Champion the Cause

From the time I was 11 years old, I began dealing with a couple of pretty significant chronic health conditions that initially required a prolonged hospitalization. I don’t know how many times I asked my parents, “Why me?” My situation interfered with my young athletic career and my social life, it impacted my grades in school for a time, and it wreaked havoc on my self-esteem, leading to periods of pretty serious anxiety and clinical depression.

I remember that Mom’s standard response to my inquiries of “Why me?” was to say, “I don’t know, Honey, but at least you don’t have [fill in the blank with some other horrible health condition]. Try to focus on all you do have.” Did Mom’s advice help? Not in the least. Mom is great at so many things, but not all of her attempts at counseling have hit the mark. Maybe it would have been better if she just focused on the “count your blessings,” instead of taking an approach that made me feel guilty for feeling lousy. It also made me think my friends’ moms were using me as the example whenever their kids felt down about getting a grass stain on their pants or a C on a test. “Don’t worry, Honey, at least you’re not Matt Hansen!”

In retrospect — another blessing of life — I learned several things from my early experience with chronic health issues. First, Mom was doing the best she knew how, and her motivations were pure. Even those who have personally dealt with debilitating illness often don’t know how to help someone else who is passing through a similar trial, and oftentimes, given our individuality, the situations aren’t similar, but we try to insist they are.

Secondly, I now realize that as difficult as those years were, they had a huge impact on who I am today and on the direction of my career. I don’t even know that it was a conscious decision at the time. It just felt natural. I knew I wanted to help people, which I’m sure was borne from my experience of being helped by so many caring people, as well as my encounters with those who never should have been in healthcare. I wanted to be sure others who were struggling had the best help available.

Many people find purpose in life from struggles they have either overcome or have come to accept. Accepting doesn’t mean giving up, it means understanding the illness is a reality; trying to do everything one feels is right to improve their condition; supporting efforts to eradicate the disease through volunteerism, donations and/or political advocacy; teaching others about the disease to increase public awareness; and/or supporting others who live with the condition.

Benefits of Our Illness to Others

It probably sounds funny to refer to an illness we’re struggling with as a benefit to others, but there are oftentimes consequences of our trials that have positive implications in the lives of friends, family and others — many of whom are never seen or heard of by the person they are helping to support.

My wife and I used to live in the state of Washington. We ran a productive therapy staffing agency and were at a crossroads. If we stayed where we were much longer versus moving back to our home state of Utah to be closer to family, we knew we would be rooted in Washington. Not that we would have minded staying there; we loved the state, our home, our neighborhood and friends! However, after trying to have children for years and finally just welcoming our first two to the family, moving to be closer to grandparents, siblings and young cousins was an active discussion.
As life would have it, not many months passed before we were presented with what we considered to be deciding factors. First, our business’ primary contract announced its need for us was going to be reduced by 40 percent over the next three months and moving forward. We could have found new contracts without much difficulty, but then we received the phone call. My mother discovered a lump in her breast that proved to be breast cancer. We probably would have been able to make a lot more money staying in Washington, but we wanted our children to know their grandparents, and we wanted to be there to support Mom. So, within a matter of weeks, we transferred our contracts, found renters for our home, loaded up the U-Haul and moved in the basement of my in-laws while we figured things out, including where I was going to land professionally because the work environment in the Salt Lake Valley was very different from what it was in Western Washington.

Mom’s now a breast cancer survivor of seven years. She went through chemotherapy, radiation and a double mastectomy. I can’t imagine not having been here to help support her through that time. But, being here was not only a benefit to her; it blessed me and my family in many ways. To start, I learned it is possible to pick up, move on and start over. I gained more faith in myself. I grew a greater appreciation for my wife and how supportive she is. My children were exposed to some of the ugly facts of life, but in a safe environment full of love. We all had the opportunity to serve, thereby becoming less selfish. In a chain of events that continues to unfold, my career and personal purpose in life have evolved and progressed. And, I was finally given the opportunity to share the same advice with my mom that she had given me as a teenager. Actually, I never had to, because Mom never once asked “Why me?”

**Life Lessons**

If you’re someone who believes in an afterlife, it’s probably not too difficult to envision how our trials, including physical illness, can help to refine and shape our souls. Life presents so many opportunities to learn and develop important lessons and characteristics from our struggles, even our sufferings.

If you’re someone who doesn’t believe in an afterlife, then YOLO (you only live once), so why would you want to dwell every day on what you can’t do or what opportunities you don’t have, versus what you can do with what you have?

Human history is replete with stories of women and men who had a dream, an opportunity, the talent and heart to accomplish their dream, and then life happened and took the opportunity away. Whatever the tragedy was — physical, mental, financial, personal — it really doesn’t matter. Some people allow themselves to become victims and fall into despair. Others rise above and acknowledge that even though the opportunity or even talent may have been lost, it doesn’t mean they have to give up.

By learning to live with chronic illness, we can become stronger.

If you don’t lose heart or can rise above and find it again, there may be another way to accomplish the dream, or a new dream may be just as valued. For me, the National Basketball Association ship sailed long ago. However, I have many other dreams, and I’m working on them. Even if you aren’t able to use your body to accomplish a dream, think of all you can use. We all have unique gifts. Maybe yours is your mind, your charisma, your vision and/or your faith.

By learning to live with chronic illness, we can become stronger. We can become wiser. We can become more patient, more caring, more grateful, more observant. We can become a better self.

Though I would never wish my past or future trials on anyone, I would not trade them for anything either. They have made me who I am, and they will form the person I am becoming.

To paraphrase Mr. Spock from “Star Trek”: Live and prosper, regardless of how long the journey may be.

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

**Reference**

Understanding
Idiopathic Inflammatory Myopathies

These often-debilitating muscle diseases cannot be cured, but they can be effectively managed with diagnostic tools and the growing number of treatments.

By Ronale Tucker Rhodes, MS

IDIOPATHIC INFLAMMATORY myopathies (IIMs), also referred to as myositis, affect between one and eight in every one million people in the U.S. This group of disorders, characterized by inflammation of the muscles used for movement (skeletal muscles), usually appears in adults between ages 40 years and 60 years or in children between ages 5 years and 15 years, although it can affect anyone at any age. And, for some unknown reason, two types of IIMs affect twice as many women as men, while the other type is more common in men.

The first description of myositis occurred in 1863 by German pathologist Ernst Leberecht Wagner who described a patient with a rare muscle disease with significant cutaneous lesions. Later, in 1887, Professor Heinrich Unverricht published an account of the muscle disease in a 27-year-old stonemason who developed acute onset of weakness, stiffness and pain in the proximal arm, leg and back muscles, followed by swelling of the face and extremities, low-grade fevers and a bluish rash over his eyelids. After several weeks, the patient died of pulmonary arrest, with an autopsy showing fluid-filled lungs and swollen proximal muscles. When Dr. Unverricht reported a second case in 1891, he coined the disease dermatomyositis (DM). And although polymyositis (PM) was described by Dr. Wagner in 1863, recognition of PM as a distinct entity was attributed to John N. Walton, MD, and Raymond D. Adams, MD, in 1938. In 1975, Drs. Walton’s and Adam’s observations were confirmed by Anthony Bohan, MD, and James B. Peter, MD, who developed the diagnostic criteria for PM and DM.

Since that time, a great deal has been learned about IIMs, with diagnostic criteria and treatments continuing to evolve, resulting in better long-term outcomes for patients.

What Are IIMs?

As mentioned previously, another term for IIM is myositis. The “myo” root meaning muscle, combined with the “itis” suffix meaning inflammation, describes an inflammatory muscle disease. IIMs are a group of diseases that involve chronic muscle inflammation that leads to destruction of muscle tissue causing
muscle weakness and, in some cases, muscle pain.\textsuperscript{6}

There are three main types of chronic (long-term) myopathies: PM, DM and inclusion body myositis (IBM). PM is a disease in which the inflammatory cells of the immune system directly attack muscle fibers,\textsuperscript{7} and onset generally occurs between ages 30 years and 60 years, but it is rarely seen in persons younger than 20 years old.\textsuperscript{6} DM is a disease in which the inflammatory cells of the immune system attack the small blood vessels that supply muscles and skin,\textsuperscript{7} with average age of diagnosis at 40 years old. DM also affects children (known as juvenile DM) with an average age of onset between 5 years and 14 years old.\textsuperscript{8} IBM is a disease that appears to be partly inflammatory and partly a degenerative muscle disease, with disease symptoms usually beginning after age 50 years.\textsuperscript{7}

**Causes of IIMs**

IIMs are thought to be caused by a combination of genetic and environmental factors. Yet, “thought” is the key word, which is why IIMs are also referred to as idiopathic because it means the specific cause of the disorders is unknown.\textsuperscript{2} However, it is known IIMs are autoimmune disorders in which the immune system attacks its own muscle fibers, blood vessels, connective tissue, organs or joints.\textsuperscript{6}

While IIMs aren’t genetic disorders, genetic factors can increase the likelihood of an IIM developing.\textsuperscript{7} For instance, researchers have identified variations in genes belonging to a family of genes called the human leukocyte antigen (HLA) complex, which helps the immune system distinguish the body’s own proteins from those made by foreign invaders such as viruses and bacteria. In addition, researchers are studying variations in other genes related to the body’s immune function to determine how they contribute to the risk of developing IIMs.\textsuperscript{7}

The actual triggers of IIMs are believed to be environmental factors such as infection, medications and even ultraviolet light.\textsuperscript{2} For instance, HIV-positive individuals and those with a virus called HTLV-1 can develop an IIM. And, drugs suspected of contributing to an IIM include articaine (a local anesthetic), penicillamine (to lower copper levels in the body), interferon-alpha (to treat cancer and hepatitis), cimetidine (to treat ulcers), carbimazole (to treat thyroid disease), phenytoin (to treat seizures) and growth hormone.\textsuperscript{7}

Lastly, most cases of IIM are sporadic, meaning they occur in people with no family history of the disorder. However, some individuals with an IIM have close relatives with autoimmune disorders.\textsuperscript{2}

**Symptoms of IIMs**

Symptoms of PM, DM and IBM are different.

PM affects skeletal muscles (involved in body movement) on both sides of the body, and symptoms differ for each person.\textsuperscript{4} Typically, muscles become weak and gradually get weaker over a period of weeks or months. Muscles in the hips, thighs, upper arms, top part of the back and shoulder area, as well as those that move the neck, are most affected. In addition, there may be tenderness in the affected areas.\textsuperscript{7} Left untreated, muscle weakness can lead to difficulty with extending the knee, stepping down or climbing stairs, fixing hair, placing things on a high shelf, raising the head off the bed when lying down, swallowing and speaking.\textsuperscript{6,7} Some individuals also develop arthritis, shortness of breath, heart arrhythmias or congestive heart failure.\textsuperscript{6}

Once considered PM with a rash, DM is now known to have fundamental differences; in fact, the skin (dermat) manifestations make it a distinct disorder.\textsuperscript{7} A skin rash, which precedes or accompanies progressive muscle weakness, appears patchy with purple or red discolorations, usually developing on the eyelids and on muscles used to extend or straighten joints (knuckles, elbows, knees, toes). Additionally, red rashes may occur on the face, neck, shoulders, upper chest, back and other locations, and there may be swelling in the affected areas. Other than rashes, adults may experience a low-grade fever, inflamed lungs and sensitivity to light.\textsuperscript{6} And, similar to PM, the muscles most affected by weakness include the shoulders, upper arms, hips, thighs and neck, and swallowing muscles can be involved.\textsuperscript{7}
to push themselves up from a prone position.\(^8\)

IBM affects different muscles than PM and DM. Progressive muscle weakness usually begins on both sides of the body in the wrists and fingers, causing difficulty with pinching, buttoning and gripping objects, as well as atrophy in the forearm and quadriceps muscles, causing falls and trips. In approximately half of people with IBM, difficult swallowing occurs due to involvement of throat muscles.\(^6\)

PM affects skeletal muscles (involved in body movement) on both sides of the body, and symptoms differ for each person.

### Complications of IIMs

In addition to symptoms of PM, DM and IBM, patients may experience other complications, including:

- Antisynthetase syndrome, a set of symptoms that typically occur in those with one of several specific autoantibodies known as antisynthetase antibodies, which are immune system proteins that target and attack tRNA synthetase enzymes. Eight antisynthetase antibodies have been identified so far as being significant in myositis diseases. This syndrome typically occurs in patients whose average age at the onset of disease is 50 years, and it occurs twice as often in women than men. Symptoms associated with antisynthetase syndrome include interstitial lung disease (approximately 75 percent of people); muscle inflammation; inflammatory arthritis; fever (about 30 percent of people); Raynaud’s phenomenon (a condition in which spasms of the arteries cause episodes of reduced blood flow, typically involving the fingers and toes); and mechanic’s hands (thickened, dry, cracked skin on the sides of the fingers and palms, which can be painful).\(^9\)

- Dystrophic calcinosis, the abnormal collection of calcium salts in or under the skin and in muscles or tendons, even when levels of calcium in the blood are normal. Calcinosis appears as hard, irregular nodules (lumps) in or under the skin in any area of the body, which can be especially uncomfortable when they appear on the face, around joints or on pressure points such as the buttocks, feet or wrists. While this occurs in approximately 20 percent of adult patients with DM, it appears in as many as 70 percent of those with juvenile DM.\(^10\)

- Cancer-associated myositis, a malignancy that develops within a year or two of a myositis diagnosis. It is believed the malignancy activates the immune system, which stimulates the development of the autoimmune disease. Cancer-associated myositis occurs most frequently in patients with DM (it’s estimated as much as 20 percent to 30 percent of DM patients will develop cancer), but only half as frequently in PM patients. And, it is very rare in children or those with IBM.\(^11\)

- Cardiovascular disease, inflammation in the heart (called myocarditis) that occurs in the same way it does in skeletal muscles, which can lead to fibrosis (scarring).\(^12\)

- Dysphagia, difficulty swallowing food or fluids usually caused by weakness in the muscles of the throat. This occurs in about one-third of myositis patients, and while it can occur in all forms of myositis, it is most common in those with sporadic IBM and juvenile DM.\(^13\)

- Infection, caused by medications that suppress the immune system, placing individuals at an increased risk for developing a serious infection. For the vast majority of myositis patients, long-term immunosuppression is the only treatment for their muscle and skin symptoms.\(^14\)

- Interstitial lung disease, a group of diseases that affect the tissue and spaces (interstitial) around the air sacs (alveoli) in the lung. When these spaces are obstructed by inflammation that is untreated for too long, it can result in pulmonary fibrosis in which the lungs are scarred causing serious breathing problems. Except for IBM, this disease is the most common and serious complication of IIMs, occurring in an estimated 30 percent to 40 percent of patients.\(^15\)

- Overlapping autoimmune diseases, when patients experience the whole range of clinical symptoms and laboratory findings of two well-defined autoimmune diseases at the same time — even those not part of one or the other disease. Overlap syndromes tend to appear more in patients with DM and PM.\(^16\)

- Rhabdomyolysis, a serious, acute condition that results from rapid death of muscle tissue, causing cells to release their contents into the blood stream. When the kidneys are unable to remove this waste quickly enough, renal (kidney) failure may result. Muscle symptoms that occur as a side effect of statin medications can also cause this complication, especially when taken in high doses.\(^17\)
Diagnosing IIMs

A myopathy is often suspected when individuals have trouble performing tasks that require muscle strength or when they present with rashes or breathing problems. When this happens, a neurologic history and exam, as well as laboratory tests, are conducted.

A history and exam will look at key diagnostic factors, including difficulty with motor tasks, muscle weakness, muscle atrophy and heliotrope rash with eyelid edema. Other diagnostic factors include frequent falls, fatigue and generalized malaise, weight loss and shortness of breath.\(^{18}\)

Tests include:\(^{19}\)

- A blood test to measure the various muscle enzymes and myositis-specific antibodies
- An electromyogram (often referred to as an EMG) to gauge electrical activity in muscle
- A biopsy of a weak muscle, which involves removing a small piece of muscle tissue (considered the gold standard)
- Magnetic resonance imaging to look for abnormal muscle

In addition, tests are often conducted to rule out cancer in DM and, sometimes, PM since these IIMs may be linked to cancer.\(^{19,20}\)

The original diagnostic classification criteria were developed by Drs. Bohan and Peter in 1975. However, these criteria were for PM and DM only; they did not recognize IBM. The Bohan and Peter criteria included: 1) symmetrical proximal muscle weakness; 2) typical rash of DM (a distinguishing feature for DM and PM); 3) elevated serum muscle enzymes; and 4) myopathic changes on electromyography. A definite diagnosis of DM requires all four criteria (including rash) and a definite diagnosis of PM requires all four criteria (without rash). Drs. Bohan and Peter also established a clinical classification system, including 1) primary idiopathic PM, 2) primary idiopathic DM, 3) juvenile DM, 4) PM or DM with malignancy and 5) PM or DM with associated collagen vascular diseases.\(^{3}\)
Because the Bohan and Peter criteria didn’t include IBM, the International Myositis Classification Criteria Project, a group of more than 100 myositis experts from rheumatology, dermatology, neurology and pediatric rheumatology, led by Professor Ingrid Lundberg and her research team at the Karolinska Institutet, developed new classification criteria for myositis in 2017, which are now accepted by the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR).

These classification criteria “are based on a probability score model that provide the best sensitivity and specificity to distinguish myositis from other mimicking conditions, and include 16 variables, including age of illness onset, type and location of muscle weakness, skin manifestations, laboratory findings and muscle biopsy findings. The criteria rely on a weighted score that sums the 16 variables, and allow for flexibility in that not all variables need to be assessed to classify a patient as having myositis.”

These new criteria include the major subtypes of myositis, covering DM, PM and IBM in adults, and juvenile DM in children. A probability score of greater than or equal to 90 percent corresponds to definite IIM, and a probability of greater than or equal to 55 percent corresponds to probable IIM and is recommended as a minimum standard to classify a patient as having an IIM. For patients without the characteristic skin manifestations and those with characteristic DM skin findings without muscle involvement, a skin biopsy is recommended. Further, subclassification of IIM can be made after a patient has been classified as having IIM using the 2017 EULAR/ACR classification criteria, and is based on a

Table 1. PM and DM Treatments

<table>
<thead>
<tr>
<th>Medication/Treatment</th>
<th>How It Works</th>
</tr>
</thead>
<tbody>
<tr>
<td>Corticosteroids prednisone tablets (Deltasone); intravenous methylprednisolone sodium succinate (Solu-Medrol)</td>
<td>Dampens inflammation and immune response by interfering with processing of antigens and with early triggering of T-cell and B-cell production and later proliferation of B cells and T cells. These cells are produced by the immune system in autoimmune diseases such as PM and DM.</td>
</tr>
<tr>
<td>Azathioprine (Imuran)</td>
<td>Interferes with proliferation of B cells and T cells.</td>
</tr>
<tr>
<td>Methotrexate (Rheumatrex, Folex, Mexate)</td>
<td>Interferes with proliferation of B cells and T cells.</td>
</tr>
<tr>
<td>Cyclosporine (Neoral, Sandimmune)</td>
<td>Keeps T cells from stimulating production of more T cells and B cells (“upstream” of azathioprine and methotrexate action).</td>
</tr>
<tr>
<td>Cyclophosphamide (Cytoxan)</td>
<td>Interferes with proliferation and activity of B cells and T cells.</td>
</tr>
<tr>
<td>Mycophenolate mofetil (CellCept)</td>
<td>Interferes with proliferation of B cells and T cells.</td>
</tr>
<tr>
<td>Tacrolimus (Prograf, old name FK506)</td>
<td>Keeps T cells from stimulating production of more T cells and B cells (“upstream” of azathioprine and methotrexate action).</td>
</tr>
<tr>
<td>Hydroxychloroquine sulfate (Plaquenil)</td>
<td>Mechanism not understood; used in arthritis, lupus, malaria; can be used to reduce steroid dosage in myositis, particularly in children.</td>
</tr>
<tr>
<td>Intravenous immune globulin</td>
<td>Has complex actions on immune system such as providing antibodies against patient’s own antibodies; interfering with immune-system reaction to antibody-marked cells; interfering with blood-transported chemicals released by immune system; interfering with activation and maturation of T cells and B cells.</td>
</tr>
<tr>
<td>Plasmapheresis</td>
<td>Removes antibodies and proteins made by the immune system from the blood and returns “cleansed” blood to the patient.</td>
</tr>
</tbody>
</table>
classification tree, taking into account patient age at illness onset, presence of characteristic rashes of DM, patterns of weakness and muscle biopsy features. Subclassifications include PM, DM, IBM, amyopathic DM or juvenile DM.21

Treating IIMs

Unfortunately, IIMs can’t be cured, but symptoms can be treated with medication (Table 1), physical therapy, exercise, heat therapy, orthotics and assistive devices, and rest.

For PM and DM, the first line of treatment is high doses of corticosteroid drugs such as prednisone either orally or intravenously to reduce inflammation. With corticosteroids, blood muscle enzymes tend to return to normal between four weeks and six weeks after treatment, with most patients regaining muscle strength in two months to three months.6,19 For those who don’t respond well to corticosteroids, immunosuppressant drugs such as azathioprine and methotrexate can help reduce inflammation. Other immunosuppressive drugs that can treat inflammation associated with PM and DM include cyclosporine A, cyclophosphamide, mycophenolate mofetil and tacrolimus.

In severe cases of PM and DM when other treatments have failed, biologic therapies such as rituximab, tumor necrosis factor inhibitors such as infliximab or etanercept, or intravenous immune globulin (IVIG) can be used. It should be noted that periodic treatment with IVIG can increase the chances of recovery.

For IBM, there is no evidence-based course of treatment, and corticosteroids and immunosuppressive drugs are generally ineffective. However, in a small number of cases, IVIG and immunosuppressive drugs have had a slight, but short-lasting, beneficial effect.

Physical therapy is recommended for PM, DM and IBM to prevent muscle atrophy and maintain muscle strength and range of motion. And, extended bed rest is discouraged since it may cause muscle atrophy, decreased muscle function and joint contractures.

Other suggestions include consuming a low-sodium diet to help reduce swelling and cardiovascular complications, occupational therapy to assist with tasks such as feeding, bathing and dressing and, in rare instances, surgery to remove calcium deposits that cause nerve pain and recurrent infections.6

Lastly, it is recommended individuals with DM avoid sun exposure during peak sunshine periods and use sunblock and protective clothing to avoid exacerbating the skin aspects of the disease.7

Long-Term Prognosis of IIMs

The prognosis for IIMs varies. Most cases of adult DM resolve with therapy. With early treatment, survival rates are as high as 80 percent and 73 percent at five years and eight years, respectively. However, in those with heart problems, the disease is usually more severe and resistant to therapy. Other poor prognostic indicators include recalcitrant disease, delay in diagnosis, older age, malignancy, fever, asthenia-anorexia, pulmonary interstitial fibrosis, dysphagia and leukocytosis. For juvenile DM, one-third of individuals recover, one-third have a relapsing-remitting course of disease and the other third have a more chronic course of illness. Poor prognostic indicators in juvenile DM include late onset of treatment, initial treatment with a dosage of prednisone that is too low, recalcitrant disease and pharyngeal involvement. In addition, up to two-thirds of juvenile DM patients develop severe complications of calcinosis cutis with mortality rates between 3 percent and 10 percent.6,8

Most people with PM respond well to therapy; however, those with more severe disease do not, and they may have significant disability. Some PM patients become malnourished due to difficulty swallowing, and there is increased risk for falls that can lead to hip and other bone fractures, disability or death. In rare cases, individuals with severe and progressive muscle weakness can develop respiratory failure or pneumonia.6

With IBM, the older the age of onset, the faster the loss of strength and mobility. After 15 years, most require assistance with basic daily routines, and while some are able to walk, those more severely affected may need a wheelchair. And, since IBM is generally a slowly progressive disease, life expectancy isn’t significantly affected; however, most clinicians agree that IBM can be an indirect cause of death due to aspiration pneumonia in patients with difficulty swallowing and, in rare causes, respiratory failure due to respiratory muscle weakness.1
Research Abounds

A host of IIM research is underway, and as of this writing clinicaltrials.gov lists 180 studies, mostly investigating new drugs and diagnostic tools. Some newer agents being studied to treat IIM that may lead to better results include:22

• Anakinra, a recombinant human IL-1 receptor antagonist used commonly for treating rheumatoid arthritis. Two studies of 15 patients treated for 12 months with Anakinra showed a beneficial clinical response in at least half.

• Alemtuzumab, a humanized monoclonal anti-CD52 antibody that causes an immediate and severe depletion in the peripheral blood lymphocytes. In a study of 13 sporadic IBM patients, alemtuzumab infusions slowed down disease progression up to six months and improved muscle strength. And, a long-term follow-up study on a treatment-resistant case showed marked improvement in muscle strength 12 weeks into a single treatment cycle with alemtuzumab that lasted approximately three years.

• Belimumab, a human monoclonal antibody directed against B lymphocyte stimulator, which is a TNF-related cytokine implicated in B-cell maturation and development. It was approved for treatment of systemic lupus erythematosus in March 2011.

• Sifalimumab, an anti-IFN-monoclonal antibody. Neutralization of the type-I IFN gene signature by sifalimumab resulted in coordinated suppression of T cell-related proteins such as soluble IL-2RA, TNF receptor 2 and IL-18.

In addition, many organizations are involved in IIM research. The National Institute of Neurological Disorders and Stroke at the National Institutes of Health (NIH) is working to identify the causes of muscle weakness to discover effective treatments, and develop objective, image-based methods for describing the muscle damage associated with inflammatory muscle disease. NIH is also studying childhood-onset PM and DM to learn more about their causes, immune system changes and associated medical problems. And, since there are no therapies approved by the U.S. Food and Drug Administration to diagnose IIMs, researchers are looking for better, less-invasive ways of diagnosing these disorders. Other areas of study include the genetic diversity of IIMs and their susceptibility to certain drugs or vaccines. MDA is also studying the IIMs occurring spontaneously in dogs to see if they offer insight into disease mechanisms. And, in IBM, MDA is investigating mice with an IBM-like condition.17

While IIM research progresses to discover more about these puzzling muscle diseases to create better treatments leading to improved outcomes, and enhanced diagnostic criteria to enable early diagnosis, the quest continues for the answers to optimal prognosis. 

RONALE TUCKER RHODES is the editor of IG Living magazine.

Table 2. Resources for IIMs

| • American Autoimmune Related Diseases Association: aarda.org |
| • Arthritis Foundation: arthritis.org |
| • Muscular Dystrophy Association: mda.org |
| • The Myositis Association: myositis.org |
| • National Institute of Arthritis and Musculoskeletal and Skin Diseases: niams.nih.gov |
| • National Institute of Environmental Health Sciences: nih.gov |

References


Making a difference in Our Patients’ Lives.

Specialty Solutions in Chronic Care
- Immune Globulin
- Factor
- Infliximab

Nufactor is committed to exceptional customer service, product and patient safety, and secure product availability and affordability. Excellence is our standard, and we’ve earned the most respected name in homecare. Our customers know we care about them, and that makes all the difference.

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Profile: Pamela Mills

By Trudie Mitschang

Wearing face masks was nothing new for Pamela Mills when they became a nationwide essential due to COVID-19. The common variable immune deficiency (CVID) patient was used to routinely donning a mask when visiting the doctor or running errands to avoid getting sick. So, when the demand for masks began to spike in early 2020, Pam used her crafting skills to begin making them — and giving them away. Her efforts were designed to help others navigate the strange new “not normal” post-pandemic world, a world quite familiar to anyone battling a primary immune deficiency (PI). Pam eventually began selling the masks and an array of other products at her online business The Craft Sage (www.facebook.com/thecraftsage).

Trudie: How did you get into the craft business?

Pam: I have been crafting since I was a child. It was something I could do on those lonely days when I was home sick with asthma. I did crafts in scouting and while waiting at the doctor’s office, or in the family RV on the way to a campsite. It seemed I was always doing something creative, and I loved keeping my hands busy. Years later, after my diagnosis with CVID, I was told I couldn’t work anymore. I tried doing many different jobs such as tutoring. Eventually, I got a position as a freelance art instructor for a company called Creatsy Art Studio. At the time, I needed a business name and some cards to help advertise my classes, and I wanted something that showed the range of my creative abilities. The term “sagacious” came to mind, and that’s how my business The Craft Sage was born!

Trudie: Tell us about the unique features of your face masks.

Pam: I was diagnosed with CVID more than 10 years ago, and I’ve been wearing masks ever since. I wear them when I go to the doctor’s office or when I’m around people who may be sick. When I started designing masks, I created them with a feature that allows for charcoal-activated filters to go inside a special pocket. The filters help absorb potentially contagious droplets, similar to an N95 mask.

Trudie: How soon after COVID-19 began were you inspired to market masks to the general public?

Pam: Last year, I saw handmade bags on Pinterest, and I decided to try and sew them. These bags became dice bags that I gave to my gamer friends for Christmas. I was making more dice bags and trying to sell them when the coronavirus began infecting people in the United States. My sister Jamie, who lives in Oklahoma, told me the pharmacy where she worked wasn’t able to get enough masks due to the pandemic, and she suggested that I start making them. My mother-in-law Kris contributed her sewing machine and quilting fabrics to help me, and I was able to purchase some interfacing, wire and elastic. Kris and I tried several styles of masks, and Jamie eventually tested them at her job. Finally, I looked at the masks I had purchased over the years for myself to wear during cold and flu season for inspiration. That’s when I put together a pattern for what I call “the pocket mask,” because of the filter pocket that fits a pm2.5 carbon filter.

Trudie: What are your personal and business goals?

Pam: My business goals are to keep things simple and fun. Kris and I are doing this to keep us busy while we are self-isolating during COVID-19. It is also something positive to do for our...
community, and it relieves some of the stress and worries we have. I do not plan to grow my business too big because I need to prioritize my health and manage my stress levels. I would like to incorporate some of my other crafts and artwork soon, and I also have plans to begin making YouTube video tutorials for crafting classes.

Trudie: Do you think you will continue selling masks?
Pam: The masks don’t actually fit into my long-range business plan. If I were to truly sell them for my cost and include the time it takes to make each one, it would be so expensive no one would buy them. The money we are earning for each mask right now is simply going to help my family with our bills at this time.

Trudie: What has this journey with chronic illness taught you about yourself?
Pam: I am resilient. I can endure so many things, and I will get through the next health crisis.

Trudie: What is your current treatment plan?
Pam: I receive intravenous immune globulin (IVIG) every two weeks. Last year, I was getting my IVIG every four weeks and I would get a severe “crash” sometimes. This crash would make me feel a terrible malaise for anywhere from several days to as long as two weeks. I talked with my doctor, and we tried subcutaneous infusions for a while, but I didn’t like stabbing myself in four to five spots each week, so I asked to go back to IVIG. That is when we decided to split my dosage and frequency in half so I now receive 20 grams every two weeks. This change has been great, and it has reduced my crash to only a day or so, which is manageable.

Trudie: How involved are you with the Immune Deficiency Foundation (IDF)?
Pam: I attended a couple of IDF events many years ago when they were held in San Jose, Calif. I also went to the IDF National Conference the year it was held in Anaheim, Calif. I was only able to attend it because I was able to get a scholarship. My mother accompanied me as my aide. Even though my mother had been a nurse for about 50 years, she was surprised at what she learned about PIs and how I probably should have been diagnosed as a child instead of when I was 39 years old.

Trudie: What advice would you offer other patients?
Pam: Ask questions and do your own homework about your disease so you are able to talk intelligently to doctors and medical personnel about your health. I have seen so many doctors who don’t know or remember what PIs are and how that can impact their treatment plans for you. You have to be your own advocate. If you can, try to teach your partner about your health so he or she can advocate for you if you are unable to represent yourself. I also carry a sheet of paper with me that has all my medical information on it. It’s the first thing I give to doctors.

Trudie: How do you stay motivated, positive and encouraged?
Pam: Like many CVID patients, I deal with depression on a constant basis. It’s hard to deal with the lack of finances, inability to work, not feeling up to being with friends or attending social events, and much more that comes with CVID. I take my antidepression medications regularly, and I also see a therapist once a month.

Trudie: Do you have a personal support system?
Pam: My partner James helps me so much when I get down. He is understanding and willing to just talk with me and help me to focus on positive things. Throughout my life, my mother has preached to “just think positive,” and I try to keep that focus. I also try to focus on things that I have control over and not those I can’t do anything about. For example, I can’t control the pandemic, but I can do something positive to help the situation by making masks to reduce the spread of infection.

Trudie: What gives you joy?
Pam: Helping others is a wonderful feeling. Finally finishing a project or painting gives me the most wonderful sense of accomplishment and pride. And, of course, my family. My mother-in-law Kris, partner James, our dogs Rockystar, Angel and Oberon, and our cat Spock all bring joy into my heart and make me smile.

Editor’s note: Pam is offering a 15 percent discount to IG Living readers who enter the code Dazzle during checkout.
PATIENT PERSPECTIVE

We’re Worth It

By Stacey Philpot

I GET A bit grumpy when I’m not feeling well. Knowing this, my 8-year-old daughter will sometimes intuitively ask, “How are you feeling, Mom?” Often, I’ll reply with a minimized summary of what’s not quite right. I might answer, “You know, I’m not feeling that well. I can’t seem to get over this cold” or “I’m running a bit of a fever, and I think I might rest after I drop you off.” Or, “Mommy wants to sleep for the next three years. Do you ever feel so tired you can’t think straight?” More than once lately, she’s commented, “But, Mom, you’re always sick.” It certainly feels true.

While waiting for approval from a new insurance company, I experienced a delay in my infusions. I had already been catching a new virus every week through the winter months, and now my immune system was particularly vulnerable. Every day, I was battling a fever, new symptoms, increasing fatigue and an overwhelming desire to hibernate until spring. Yet, the demands of life had not stopped. I felt cranky. I needed to make a few adjustments to protect my immune system and improve my mood. Here are a few of the changes I made:

1) I listened to my body. Even though it sometimes meant I might have dirty dishes in the sink or more to do later, I rested when I needed it. Even when it meant I might disappoint someone because I had to say no or cancel plans, I put my body first. This is hard to do, but our bodies are worth it.

2) I was intentional about where I spent time. This meant I alternated going to the gym and working out at home. There are few things I love in the world quite like my gym yoga class. I enjoy the gentle stretching and the way it wakes my body up, as well as the socialization. But even the smallest amount of exercise pushes my body, putting it at risk for infections and viral activity. No matter how careful I am at the gym, I can usually count on catching something within 24 to 48 hours. For this reason, I alternate between my beloved gym yoga class and living room floor yoga with a YouTube instructor. My body still gets the movement in my living room, but without the germs.

3) I let the light in. In the winter months, I begin to wonder if I will ever see the sun again. I need to find light wherever I can. I drive with my sunroof open, throw open all the blinds and access as much natural light as I can. Letting the light in may also include listening to uplifting messages, watching favorite shows and leaving the house when it would be easier to remain in isolation.

4) I connected with others. Whether this is in online groups or lunches with friends, I need to laugh, have meaningful conversations, hear about my friend’s bathroom remodel and look at baby pictures. One of the tools I use for this is a walkie-talkie app. Friends leave voice messages for me that I can listen to any time. Likewise, I can respond whenever I’m feeling up to it without a lengthy phone conversation. Sometimes, I need to hear the sound of someone else’s voice, but I don’t feel up to talking. This is an excellent way for me to stay connected.

Feeling less like ourselves emotionally can be a normal part of sickness. However, if we notice we no longer enjoy the things or people we once did, have disrupted sleep and eating patterns, have feelings of hopelessness or can’t shake that “not-quite-ourselves” feeling, it’s probably time to talk with someone. Just as there is no shame in seeking help for our physical symptoms, there is no shame in seeking help for our emotional symptoms. Our bodies and our hearts deserve the best possible care. Talking about our emotional symptoms can require courage, but we’re worth it.

STACEY PHILPOT is an author, goofball and avid reader. You can find her blog at chronicallywhole.com, where she shares her journey of making the most of a life touched by common variable immunodeficiency, Lyme disease and rheumatoid arthritis.
RESEARCH SHOWS smartphone apps can help people living with chronic illness achieve remission, maintain overall wellness and cope with disease. And, mHealth (mobile health) is only going to keep advancing. Here are my picks for the 21 must-download apps now.

To manage chronic illness, try apps created for the disease you’re dealing with:
• Cancer: chemoWave (chemowave.com; free for iOS and Android)
• Chronic pain: Curable (curablehealth.com; free for iOS and Android)
• Idiopathic pulmonary fibrosis: patientMpower (info.patientmpower.com/patient-support-programs/#breaker-psp; free for iOS and Android)
• Irritable bowel diseases: mySymptoms (skygazerlabs.com/wp; $2.99 for iOS and Android)
• Primary immunodeficiency: IgCares (www.cutaquigus.com/igcares-patient-support-and-resources/igcares-mobile-app; free for iOS and Android)
• Rheumatoid arthritis: MyVectra (trackmyra.com; free for iOS and Android)

To track symptoms:
• Flaredown (flaredown.com; free for iOS, Android and the web)

To get a handle on mental health:
• Moodnotes (App Store or Microsoft Store; $3.99 for iOS only)
• Sanvello (sanvello.com; free for iOS and Android)

To learn how to cope with chronic illness:
• SuperBetter (superbetter.com; free for iOS and Android)

To improve anxiety and sleep better:
• Calm (calm.com; free for iOS and Android)
• Sleep Cycle (sleepcycle.com; free for iOS and Android)

To speak with a licensed therapist:
• Talkspace (lp.talkspace.com; free for iOS and Android, although users must pay for appointments)
• 7 Cups (7cups.com; free for iOS and Android)

To keep up with taking and refilling meds:
• Medisafe (medisafe.com; free for iOS and Android)
• MyTherapy (mytherapyapp.com; free for iOS and Android)

To make life more accessible, try apps that can help you better navigate the world:
• Instacart (instacart.com; free on iOS, Android and the web)
• Wheelmap (wheelmap.org; free on iOS, Android and the web)
• Lyft (lyft.com; free on iOS and Android)
• Bathroom Scout (App Store or Google Play; $0.99 on iOS and Android)
• Be My Eyes (bemyeyes.com; free for iOS and Android)

All my suggestions are based on recommendations from individuals with chronic illness: real app users and people living and thriving with diseases, thanks to these technical innovations. Whether you’re working toward a diagnosis, seeking to find the right treatment or looking to maintain your current level of wellness, these excellent tools will help you get healthy and stay healthy.

BRENDA KIMBLE is a writer and caregiver based in Austin, Texas.
CHILDREN WITH a primary immunodeficiency (PI) are used to taking certain precautions to prevent infections. These may include taking prophylactic antibiotics, taking antibiotics during the winter months or avoiding crowds during influenza season. But kids with some forms of PI may have added risk when it comes to the health of their teeth and gums. For this reason, PI children and their parents should be extra vigilant with their dental care, and their dentists (who may not be very familiar with their condition) should be informed of the risks associated with PI and oral health.

The type of PI a child has is directly related to specific dental concerns. For example, patients with an antibody deficiency such as X-linked agammaglobulinemia (XLA) may suffer from gingivitis, candidiasis (yeast infections such as thrush) or recurrent aphthae (small ulcers that occur in groups in the mouth or on the tongue). And, patients with DiGeorge syndrome are susceptible to enamel hypomineralization (softening and discoloration of the enamel), cavities and altered eruption patterns. Other oral or dental concerns that can occur with various PIs include ulcers, increased viral infections, periodontitis (inflammation of the tissue around the teeth), abscesses and oral lichenoid lesions.

High Fever and Tooth Enamel Defects

In addition to the type of PI, multiple childhood infections that cause high fevers can lead to enamel damage in young teeth. According to Jae Seon Kim, DDS, a dentist at Pacific Modern Dentistry in Seattle, Wash., high fevers make teeth weak and prone to bacterial attack. “High fever tampers with the calcification process of the enamel in young teeth,” he explains. “This causes the enamel to be discolored and have an irregular surface. The rough patches on the enamel are more susceptible to decay.”

According to Dr. Kim, when enamel is weak, it is easily worn out by gastric juices, and small amounts can break away as children chew, grind, brush their teeth or drink carbonated beverages. Also, high fever may cause teeth to be permanently or temporarily discolored. “The discoloration can affect the tooth in parts or the entire tooth,” he says. “In infants, metabolic diseases easily cause the fevers that cause an imbalance in the calcium and protein content of the enamel. This imbalance consequently affects the color of the teeth.”

Before my oldest son was diagnosed with XLA at 3 years old, he suffered from multiple infections such as bronchitis and recurrent ear infections. He had five fevers over 105 degrees Fahrenheit in a four-month span. By the time he was 12 years old, an X-ray showed decay in the apex of his tooth, and he required a root canal. We now know the fevers that occurred while his permanent teeth were still forming had a direct effect on the strength of his tooth enamel, which led to decay. There is a visible line...
running across the upper third of each of his teeth, marking the point at which the fevers occurred. The root canal, unfortunately, will not be his last. For this reason, the negative impact of PI on dental health is just one more reason why early detection is so important.

Infections and Complications after Dental Procedures/Surgeries

Dental surgeries such as root canals and wisdom tooth extractions can be painful and dangerous for anyone, but for those with PI, they can carry a higher risk. According to David Dart, DDS, a dentist at the University of California Santa Barbara Dental Care Center, people who suffer from immune deficiencies are at greater risk for sepsis following invasive dental procedures since bacteria are introduced into the bloodstream from the mouth. Dr. Dart recommends that before any dental procedure, PI patients should be sure their blood IgG level is above 600 mg/dL. This will lower the risk of infection following invasive dental procedures, he says, which include any “procedures that involve substantial local anesthetics and/or deep drilling into teeth, which can cause significant bleeding in the oral cavity.” He recommends PI patients are carefully monitored prior to, during and after any invasive dental procedure, and that the patient’s dentist and medical team work together while planning the dental treatment to ensure the procedure is conducted safely. Sometimes, depending on a patient’s risk, it is necessary to prescribe antibiotics before and/or after an invasive dental procedure.\

Sam’s Story

Sam is a 15-year-old with XLA. In July 2019, he had his two lower wisdom teeth removed. The oral surgeon consulted with his hematologist to see if any extra precautions needed to be taken due to his XLA. It was decided he would be treated as a normal patient and would receive just one day of prescribed antibiotic before surgery. The surgery went well, but approximately 10 days after surgery, the right side of his face became swollen, although he had no fever or pain. The oral surgeon deemed he had an infection, started Sam on antibiotics and then drained the abscess. A culture indicated the infection was typical of the bacteria that can cause infection after wisdom tooth extraction. Sam was checked daily and began to recover while taking a 10-day course of antibiotics. Shortly after, he attended sleepaway camp, but after just three days, he had to be hospitalized for one night due to intense stomach discomfort. At the hospital, the antibiotics were stopped.

At 35 days postsurgery, Sam’s face again became swollen on the right side, but again he had no fever or pain. Another visit to the oral surgeon revealed another infection at the site of the wisdom tooth extraction. The surgeon was puzzled, and said it was rare for a second infection to occur in the same spot. He consulted with Sam’s hematologist and prescribed a different antibiotic and again drained the abscess. This time, Sam healed well, and the infection did not return. Whether Sam’s XLA contributed to his first infection is unclear since infection after wisdom tooth extraction isn’t unheard of. What was odd, though, was the second infection. Sam’s mother, Leah, wonders if a longer course of antibiotics before the surgery and after might have prevented this. Obviously, the decision to prescribe antibiotics before or after dental surgery is one that should be made by the child’s dentist along with the doctor who regularly treats the child’s PI.

Teach Good Oral Hygiene

Because of their higher risk of infections, PI children are more susceptible to dental complications than their peers. Good oral hygiene should be taught at a young age whether a child has an immune deficiency or not, but it is especially important for kids with PI. Parents should make a point of examining their children’s mouth and gums regularly for signs of decay or inflammation (bright red gums, bleeding when brushing/flossing). Also, they should schedule dental exams at least twice a year, and make sure the child is brushing and flossing regularly while also avoiding sugary foods and drinks.

Starting with their children’s first dental exam, good communication between parents, the children’s dentist and primary healthcare providers is crucial for optimal dental and oral health.

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

References
JUST RELAX!

That’s easy to say when we’re not in the midst of a pandemic that has turned the world upside down. For many individuals, particularly those in the high-risk population such as patients who receive immune globulin infusions, just stepping outside during these unsure times can lead to heightened stress and anxiety. Still, even in the face of extremely traumatic scenarios, there are numerous ways to relax the mind and body.

Fight or Flight

When the human body faces stressful situations, a flood of hormones is released, and the decision is made to fight or flee. This response to stress evolved as a survival mechanism, enabling humans to react quickly — without overthinking it — to life-threatening situations. While these innate reactions can help in certain scenarios, they can also cause the body to overreact to nonserious situations such as public speaking, traffic and career woes.

When the body is under “attack,” it releases cortisol. This so-called stress hormone is momentarily helpful, but not when it sticks around. For individuals under constant stress, lingering cortisol can lead to an array of side effects, including depression, weight gain, irritability and heart disease.

Training Day

Ready to ward off the undesirable side effects of stress and anxiety? Then grab those mental resistance bands and start flexing those brain muscles. It’s entirely possible to teach the human brain, even for the population more genetically prone to anxiety, to respond less destructively to stress.

Herbert Benson, MD, first introduced the theory of the “relaxation response” in the 1970s in the Harvard publication Stress Management: Approaches for Preventing and Reducing Stress. In the report, the cardiologist discussed what were then unusual practices: meditation, yoga, tai chi and qigong. Dr. Benson also highlighted other relaxation-inducing techniques, including breath focus, body scan and guided imagery. Breath focus simply involves disengaging the mind from distracting thoughts and focusing. Taking calming breaths can be particularly helpful when blood pressure rises due to anger or anxiety-inducing moments. A body scan combines breathing exercises with muscle relaxation. By focusing on one part of the body or a group of muscles, the brain mentally releases physical tension in those specific areas.

Count Your Blessings

It’s an old adage, but daily recognition of your blessings can promote peace and productivity. In his book You Are Awesome, Neil Pasricha writes that a simple two-minute morning ritual can make all the difference in how the day pans out. The Toronto-based author cracks open his journal every morning and writes out three prompts: “I will let go of,” “I am grateful for” and “I will focus on.” The idea behind these prompts is that setting regret and anger free, acknowledging what you’re thankful for and expressing your daily intentions invites more happiness into your life and helps you cope better with stress while avoiding distractions and temptations.

Sleep Soundly

Sleep is essential to everyone’s health and well-being. Although the amount of necessary sleep varies, adults aged 18 years to 60 years need at least seven hours a day for optimal health, according to the American Academy of Sleep Medicine. Getting less than seven hours of shut-eye a day is associated with an increased risk of obesity, diabetes, high blood pressure, heart disease, stroke and mental issues.

Unfortunately, one in three American adults aren’t snoozing enough on a regular basis, according to the Centers for Disease Control and Prevention’s 2016 Morbidity and Mortality Weekly Report. Stress, sleep disorders, alcohol, diet and bedtime routines are some of the major obstacles that stand in the way of a good night’s sleep. And these days, with the constant buzz of smartphones and the stress of the day lingering, it’s difficult to wind down. Experts recommend turning off notifications, putting the phone away a few hours before bedtime and creating an electronic-free sanctuary in the bedroom that promotes sleep and relaxation.

Take Advantage

Although we are currently living in a world of uncertainty, taking advantage of these relaxation techniques can lead to a more peace-filled presence. So, jot down some inspirational words, practice those breathing techniques, brew up that cup of chamomile and just relax.

HEATHER BREMNER CLAVERIE is a contributing writer for IG Living magazine.
**Brew and Chill**

Herbal products such as a Cup of Calm, a blend of passionflower, chamomile, lavender and catnip, all known as “nervines” due to their nervous system-supporting elements, can help individuals unwind. If looking to sleep, the valerian blend from Nighty Night Extra can help. The company also offers an array of mood-enhancing herbal teas. Prices vary; [www.traditionalmedicinals.com](http://www.traditionalmedicinals.com)

**Relax Your Peepers**

With the Tranquileyes Travel and Sleep Kit by Eye Eco, a relaxed sleeping environment is possible. This kit includes goggles that are padded with comfortable memory foam and a set of removable inserts that can be soaked in hot water prior to sleep to help soothe tired eyes. The pads can be frozen to ease migraines or puffiness. $49.00; [www.eyeecco.com/tranquileyes-travel-and-sleep-kit-lavender-.html](http://www.eyeecco.com/tranquileyes-travel-and-sleep-kit-lavender-.html)

**Shopping Guide to Relaxation Aids**

**Clear Your Mind**

Apps that help infuse relaxation into the daily routine are all the rage, but the Headspace app boasts a variety of guided exercises, sleeping aids, videos, articles and more to help lead the way to happier, more centered lives. Free two-week trial or $69.99/annually; [www.headspace.com](http://www.headspace.com)

**Become a Brainiac**

The Muse headset gives users an accurate real-time report of the brain’s activities. Using advanced signal processing, it interprets the user’s current mental state and helps improve meditation sessions. Starting at $224.99; [choosemuse.com](http://choosemuse.com)

**Knead Out Stress**

The Brookstone Shiatsu Neck and Back Massager with Heat and Automated Programs brings that massage therapist into one’s own home. Three levels of intensity will soothe aching muscles with the deep-kneading massage nodes and relieve tightness. $249.99; [www.amazon.com/Brookstone-Shiatsu-Massager-Automated-Programs/dp/B01LXFK3NQ/ref=sr_1_1?dchild](http://www.amazon.com/Brookstone-Shiatsu-Massager-Automated-Programs/dp/B01LXFK3NQ/ref=sr_1_1?dchild)

**Breathe to Relieve**

Essential oils can be inhaled to help relieve anxiety and stress by adding them to a diffuser or applying them to the skin when mixed with a carrier oil such as coconut or almond oil. There are a variety of oils available from sleep-promoting lavender to nervous system-calming jasmine. Prices vary; [www.nowfoods.com/essential-oils](http://www.nowfoods.com/essential-oils)
One way individuals with chronic illness can take charge of their lives and achieve better health and well-being is through the art of storytelling. Social science and emerging neurobiological research reveal storytelling helps people heal, cope and communicate, lifting their mood and regaining control of their lives’ narratives. With practical, science-backed guidance and relatable human stories, *The Healing Power of Story* offers readers an opportunity to improve health and deepen human connections that serve as the foundation of healing through engaging with and telling their stories.

### What’s Missing from Medicine: Six Lifestyle Changes to Overcome Chronic Illness

**Author:** Saray Stancic, MD  
**Publisher:** Mythos & Ink

In *What’s Missing from Medicine: Six Lifestyle Changes to Overcome Chronic Illness*, Dr. Stancic shares what she’s learned about the power specific lifestyle changes can have for those living with chronic illness. Her medical practice is dedicated to prevent, treat and even reverse chronic illnesses such as heart disease, diabetes, irritable bowel syndrome, Crohn’s disease, lupus, Parkinson’s disease and many others, all by using tools grounded in science and backed up by medical studies. Dr. Stancic is highly critical of the medical community’s lack of success when it comes to treating chronic illness. Therefore, this book is both an invitation to a better life, as well as a clarion call for the medical establishment to make lifestyle changes an integral part of the practice of medicine.

### The Joyful Caregiver: 8 Steps to Prevent Caregiver Burnout

**Author:** Josephine Grace  
**Publisher:** Morgan James Publishing

Author, speaker, teacher and caregiver Josephine Grace uses “The Graceful Process” within *The Joyful Caregiver* to bring ease and comfort to caregivers and their loved ones as they get the care and help they need to fight their cancer or another debilitating disease. For those who are serious about dedicating a portion of their life to the service of a family member with an illness, this book will help caregivers learn how to help their loved ones beat their chronic disease through clear and informative practice; communicate clearly with doctors and prevent medical errors with their care; get extra support and resources when they need them; care for themselves in the process and be guilt-free; make decisions coming from love rather than fear; and stay strong and give their loved ones the support and care they need, no matter how hard it gets.

### Super Sick: Making Peace with Chronic Illness

**Author:** Allison Alexander  
**Publisher:** Mythos & Ink

Superheroes don’t face the challenges of the chronically ill, which include socially inappropriate topics like mental illness, sex and diarrhea. Allison Alexander, who has struggled with a chronic illness since she was a child, wants to see herself in her heroes and searches for examples of sick characters in pop culture. She weaves her own painful experiences with stories from other chronic sufferers, engaging with how society values healthiness, how doctors don’t always have answers, and how faith, friendship and romance add pressure to already complicated situations. If readers are a fan of Marvel, Harry Potter, Final Fantasy and other stories from pop culture, they may find some familiar references inside. Journey through sage stories as Alexander makes peace with her illness despite a culture that suggests she’s worthless unless she’s healed.
“You can lament what is lost to you, whether it’s opportunity, a person or your health, but clinging to anger is no way to experience life.” — Rebecca Zook in “Life Lessons,” excerpted from *Chronic Inspiration*.

Download a daily dose of inspiration with this heartfelt compilation of writings on life with chronic illness. From coping strategies and parenting tips to “from the trenches” advice on dealing with family and friends who simply don’t get it, these personal stories are sure to uplift, challenge and inspire. Honest and candid, *Chronic Inspiration: Heartfelt Perspectives on Life with Chronic Illness* gives voice to those who refuse to let their diagnosis define who they are or what they can accomplish.

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

— Jenny Gardner

*Chronic Inspiration* can be purchased on iTunes, Amazon and Barnes and Noble.com
### Ataxia Telangiectasia (A-T)
- **WEBSITES**
  - A-T Children’s Project: www.atcp.org

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

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

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

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

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

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

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

### Multiple Sclerosis (MS)
- **WEBSITES**
  - All About Multiple Sclerosis: wwwMULT-sclerosis.org/index.html
  - Multiple Sclerosis Association of America: mymsaa.org
  - Multiple Sclerosis Foundation: www.msfound.org
  - National Multiple Sclerosis Society: www.nationalmssociety.org
- **ONLINE PEER SUPPORT**
  - Friends with MS: www.FriendsWithMS.com
  - MSWorld’s Chat and Message Board: www.msworld.org
  - Overcoming Multiple Sclerosis: overcomingms.org/community

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

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

### Primary Immune Deficiency Disease (PI)
- **WEBSITES**
  - Immune Deficiency Foundation: www.primaryimmune.org
  - Jeffrey Modell Foundation: www.info4pi.org
  - The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/Pages/index.aspx
  - American Academy of Allergy, Asthma & Immunology: www.aaaai.org
  - International Patient Organisation for Primary Immunodeficiencies (IPPO) — UK: www.ipopi.org
  - New England Primary Immunodeficiency Network: www.nepin.org
  - Rainbow Allergy-Immunology: uhhospitals.org/rainbow/services/allergy-immunology
- **ONLINE PEER SUPPORT**
  - IDP Friends: www.idfpfriends.com
  - Jeffrey Modell Foundation Facebook Page: www.facebook.com/JMFworld
  - IDP Peer Support Program: www.primaryimmune.org/idp-peer-support-program
  - Michigan Immunodeficiency Foundation: www.primaryimmune.org/en/nonprofits/2432e2b2a159425e06ce8d8a0709cbe-michigan-immunodeficiency-foundation-monroe

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

### Stiff Person Syndrome (SPS)
- **WEBSITES**
  - American Autoimmune Related Diseases Association Inc.: www.aarda.org
  - Genetic Alliance: www.genetecalliance.org
  - Living with Stiff Person Syndrome (personal account): www.livingwithspss.com
  - Stiff Person Syndrome: www.stiffpersonsyndrome.net
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