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Keeping Safe with Face Mask Facts and Best Practices

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Actively Participating in Your Care

MOST OF US were already feeling vulnerable and anxious in the midst of the COVID-19 pandemic that has now intensified with health officials warning it could get even worse during the influenza season, especially for those at increased susceptibility of severe illness. To address this turn of events, we deviated from our originally scheduled topic for this issue to provide you with timely guidance for these trying times. We know that while you rely on your healthcare team to manage your illness, perhaps the most important member of that team right now is you. Being an active participant in your own care is essential.

During this pandemic, it’s critical to be a strong advocate for your physical health. As we emphasize in our article “Take Charge of Your Healthcare by Becoming Your Own Advocate” (p.16), everything that was considered normal about how you take care of your healthcare needs has changed. Now, you must be extra vigilant about keeping your distance from others and getting treatment in healthcare settings. To help, Rachel Colletta, director of education resources at the Immunoglobulin National Society, provides some strategies for how you can advocate for yourself by reaching out to patient advocacy groups and getting involved in your care. She suggests learning everything there is to know about immune globulin therapy and its risk factors, ensuring your infusion providers are well-trained (also see the Let’s Talk column on p. 42) and committing to communicating openly with your specialists.

Equally important as your physical health is your mental health, particularly now when dealing with the anxiety this pandemic causes. Surayyah Morris offers support for patients with chronic conditions, providing some helpful suggestions to “rewind and settle your mind” in her article “Protecting Mental Health When Dealing with Chronic Illness” (p. 20). Her suggested approaches, including practicing breathing techniques, gratitude, meditation, yoga, self-care and more, are simple yet effective. But, she emphasizes that you shouldn’t always try to go it alone. If needed, it’s important to seek help from a professional.

Finally, we know there are many questions concerning one of the most effective protections against the COVID-19 virus: face masks. Our article “COVID-19 Mask Facts and Best Practices” (p. 24) explains who should wear a mask and how to follow the Centers for Disease Control and Prevention guidelines. We detail which types of masks are most protective and how to properly wear them, as well as describe what some are doing to overcome the secondary consequences of wearing face masks.

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.
Myths and Facts About Convalescent Plasma

By Abbie Cornett

I BELIEVE that information about appropriate medical treatments for diseases should be available to everyone. And, during the COVID-19 pandemic, this is more important than ever! Unfortunately, the emergence of COVID-19 is not the only pandemic we are facing. We are actually facing two pandemics.

The first is COVID-19, which in nine short months has taken the lives of millions around the globe and completely rewritten societal and economic norms. The second is a pandemic of misinformation about the virus and its potential treatments, which has led to confusion and fear among patients and their families.

One of the most talked about and least understood potential treatments for COVID-19 is convalescent plasma, the liquid portion of blood that is collected from patients who have recovered from this coronavirus. Roger H. Kobayashi, MD, a clinical professor at the UCLA School of Medicine and a national consultant for the Immune Deficiency Foundation, describes convalescent plasma as that derived from donors who have recovered from an illness that contains antibodies against the infection posing a threat. The assumption is the antibodies will protect the recipients who do not have the protective antibodies against the infectious threat. Yet, many myths concerning convalescent plasma exist:

Myth: Convalescent plasma is a new treatment.

Fact: Convalescent plasma has been treating various diseases for more than 100 years, including the Spanish flu in 1918, where it had mixed results.2 By administering donated plasma from donors who have recovered from a virus or bacteria, the immune system of a person infected with that same bacteria or virus who receives the donor plasma gets a boost to his or her adaptive immune system, which confers passive immunity. However, passive immunity diminishes in a relatively short time, usually a few weeks or months.3

Myth: Convalescent plasma is a recommended treatment for COVID-19.

Fact: On Aug. 23, 2020, as part of the U.S. Food and Drug Administration’s ongoing efforts to fight this pandemic, the agency issued an emergency use authorization (EUA) for the use of investigational convalescent plasma to treat hospitalized patients with serious or life-threatening COVID-19 infection.3 However, this doesn’t mean convalescent plasma is a proven treatment for the virus. Indeed, it’s important to understand that while outcomes were tracked of the some 70,000 COVID-19 patients who were treated with convalescent plasma prior to the EUA, those outcomes were not intended to be a formal clinical trial since there was no control arm. A valid study design would involve randomized placebo-controlled trials, in which half of participants are randomly allocated treatment with convalescent plasma, and the outcomes for that group are compared with the other half that receives placebo (an inactive medicine).4 Currently, no medication is recommended to treat COVID-19, including convalescent plasma.5

Myth: The donation of convalescent plasma for the treatment of COVID-19 will result in a shortage of immune globulin (IG) products for patients with immunodeficiency and autoimmune disorders.

Fact: According to Terry O. Harville, MD, PhD, medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, this myth is false! Significantly, most convalescent plasma is not being collected in donation centers that collect plasma for IG production. Convalescent plasma collection typically occurs in hospitals that have an apheresis department. Apheresis is used for the collection of donor blood components (such as a platelets or plasma), as well as for the treatment of certain medical conditions in which a part of the blood that contains disease-provoking elements is removed.6

Unfortunately for patients with immunodeficiencies or autoimmune disorders, this doesn’t necessarily mean the COVID-19 pandemic won’t have a negative impact on IG supply. Quarantining and fear of going to public places could contribute to a reduction in plasma donors.

Tragically, there is a great deal of other misinformation regarding COVID-19. So, it is up to all of us to do our due diligence when discussing medical treatments.

References

ABBIE CORNETT is the patient advocate for IG Living magazine. She can be reached at patientadvocate@igliving.com or (800) 843-7477 x1366.
Can Your Friends Handle Your Illness?

Yes, my friends are amazing. They listen to me and care about my health. A couple of them even drive me to my infusions and stay with me to keep me company. — Peggy Sue G

Noope. I actually lost a friend because she said talking about all of my medical stuff “stole her happiness.” Even most family members don’t really want to hear the details of my common variable immunodeficiency (CVID)/chronic lymphocytic leukemia life. When they ask how I am doing, all they really want to hear is “I am doing OK” even if I am not. The isolation and loneliness this life causes is almost as bad as the disease itself. Thank goodness for my CVID support group friends. — Lisa AG

I have some who do, and the rest I just let go. — Heather M

Does Your Chronic Illness Require You to Operate in Survival Mode?

I stopped and thought about just how long I’ve put a “mask” on. I’ve been doing it for over 40 years. No wonder I’m emotionally tired. — Heather M

Survival mode means just because I did something before, doesn’t mean I can do it today. Each day is a new challenge. Each infusion is new. It does not get easier with time. — Peggy SG

How Often Do You Fake Being Well?

Too many times, I have faked being well. It has taken a long time, but I finally realized I should no longer pretend. If I’m sick, I have now learned self-care is more important. It’s not about being selfish, it’s self-preservation, and I take care of me first. — Kay PB

Every day! I think that’s rather common for most with chronic illness in a variety of ways, whether it be to not worry someone, not have to discuss it yet again or a whole host of other reasons. Sometimes even for yourself you do it. — Donna G

Never. If I am not well, I tell my sister that I can’t do her online work. Or if I’m scheduled to go out somewhere or do something, I tell somebody and then I don’t go. — Rachel D

All the time. I have learned over the years when someone says “How are you,” they really don’t want to know, and that is OK. Some days, I don’t want to know how I’m doing. I usually answer “good.” If my husband is around, he will usually say, “That’s not true.” — Jenny G

Join the conversation! Connect with other immune globulin patients through IG Living’s Facebook page at www.facebook.com/IGLivingMagazine. Each day, we post interesting articles and facts, as well as thought-provoking questions you can weigh in on. These are some snapshots of what’s being discussed.
Can intravenous immune globulin therapy be started if a person has a current infection?

I am starting intravenous immune globulin (IVIG) therapy and am a lymphoma cancer survivor from 2012. For the last two-and-a-half years, I have had constant infections such as strep throat and sinus infections, but worst of all are the bacterial urinary tract infections (UTIs) caused by extended-spectrum beta-lactamases bacterial enzymes. I have never had a UTI before this, and now I suffer with symptoms of them every day. I am hoping IVIG therapy will give me back some quality of life. I believe I currently have a UTI, so I am wondering if IVIG therapy can be started.

Abbie: I spoke with Leslie Vaughan, RPh, chief operations officer at Nufactor, a Specialty Pharmacy, and she said IVIG therapy should both reduce the frequency and severity of the infections you are experiencing. However, she does recommend you speak with your treating physician before starting treatment if you think you have a current infection since patients with infections can experience worse side effects with IVIG.

Can someone with high levels of IgA antibodies be treated with a non-IgA-depleted immune globulin product?

I have common variable immunodeficiency, and I have been receiving intravenous immune globulin (IVIG) every four weeks since 1996 with good results. Because I have no IgA and a high level of antibodies to IgA, I have always been treated with a low IgA (IgA-depleted) product, and I have never had a reaction. However, a reportedly prominent immunologist told me that since even the low IgA product may have some trace amounts of IgA and I have never had a problem, I could “probably” use a standard non-IgA-depleted product. I’m questioning if this is true since the lab test report states an antibody level greater than 100 indicates a 62.5 percent chance of a severe anaphylactic reaction to the standard product, and my IgA antibody level is 120. My immunologist would like me to try subcutaneous IG (SCIG) even though there is no low IgA subcutaneous product available. She said she knows of no cases in which there have been a reaction, and the belief is the body filters out any IgA content when treated subcutaneously.

Abbie: According to Vaughan, current recommendations include use of a low IgA product for individuals who have IgA antibodies and/or prior reactions to IG products. If your immunologist believes it would be clinically appropriate to proceed with a standard non-IgA-depleted product infused intravenously, the best course of action may be to receive the infusion in a controlled clinical setting with physicians on site in case of an anaphylactic reaction. Whether SCIG is an option, there are a few literary articles that outline the safe use of SCIG in patients with low IgA and IgA antibodies. So, considering the presence of IgA antibodies, receiving the first dose in a controlled clinical setting would be warranted.

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

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Type II Hypersensitivity: Non-IgE Antibody

By Terry O. Harville, MD, PhD

IN PREVIOUS issues, we have discussed true allergic disease caused by IgE antibodies, which is known as type I hypersensitivity in the Gell and Coombs classification system of histopathologic reactions (type I, type II, type III and type IV hypersensitivities). Here, we will transition to a discussion of type II hypersensitivity. Confusingly, types II, III and IV hypersensitivity may be present at the same time, but they are not allergic reactions, so categorizing them as specific diseases can result in obfuscation rather than clarification. However, understanding the mechanics of what occurs can help to clarify their pathophysiology. Fortunately, the type II hypersensitivity mechanism of action is relatively straightforward.

Type II hypersensitivity occurs when an IgG, IgA and/or IgM (but not IgE) antibody binds directly to a cell surface or structural component in the body and results in disease symptoms or manifestations. It is not hypersensitivity in the common use of this word; it is named so only in context of the Gell and Coombs classification system. Further, it is not an allergic reaction by typical definition.

Type II hypersensitivity is a relatively common pathophysiology of autoimmune disease. The most common manifestation occurs when subclasses 1, 2 and 3 of IgG and IgM activate complement and result in damage to cells, tissues and organs. There are multiple examples:

• Autoimmune hemolytic anemia, which occasionally occurs in patients with common variable immune deficiency (CVID), is a condition in which autoimmune antibodies develop and bind to specific proteins found on the surface of red blood cells (RBCs). The autoimmune antibodies can activate complement and result in the lysis or breakdown of the RBCs.
• Autoimmune thrombocytopenia is caused by the binding of antiplatelet antibodies to proteins on the surfaces of platelets. Further, it is an unfortunately all-too-common autoimmune complication in patients with CVID.
• Pemphigus (vulgaris and foliaceus) is a condition in which IgA antibodies are directed at the adhesion molecules necessary to bind and hold cells together. The pemphigus IgA antibodies cause a loss of adhesion between the cells, causing them to separate and cause blistering on the skin.
• Goodpasture’s disease occurs when antibodies (primarily IgG) bind to the basement membrane, a form of connective tissue that helps hold the capillaries and alveoli of the lung in the correct configuration and the capillaries and kidney cells of the glomeruli in the correct positioning. Antibodies to the basement membrane disrupt these areas and capillary integrity, and patients suffer from bleeding in the lungs (hemoptysis) and bleeding from the kidneys (hematuria).
• Other conditions in which autoimmune antibodies bind directly to cells and exert disease include myasthenia gravis and autoimmune hyperthyroidism.

Another form of type II hypersensitivity occurs when certain IgG antibodies are directed toward a donor’s HLA proteins, which is one cause for rejection of a transplanted organ. Anti-HLA IgG antibodies commonly develop in women during pregnancy, when the mother makes antibodies to the father’s HLA type different from her own. If these women donate plasma, these antibodies may be found in plasma that is processed into intravenous immune globulin (IVIG) or subcutaneous IG (SCIG). In modern plasma processing, these antibodies are typically precluded from the final IVIG and SCIG products; however, they may occasionally be present. If so, and if the anti-HLA antibodies are directed toward the HLA type of the IVIG recipient, a disease process called TRALI (transfusion-related acute lung injury) may occur. Therefore, TRALI is a form of type II hypersensitivity that can occur with the infusion of IVIG and is not an autoimmune disorder. Fortunately, this can be readily treated in most instances. (More on this and other issues associated with hypersensitivities and immunodeficiencies, as well as issues with antibodies in IVIG and SCIG, will be discussed in future columns.)

In summary, type II hypersensitivity occurs when IgG, IgA and/or IgM antibodies directly bind to the surfaces of cells or components in the body, in some instances with the ability to activate complement (IgG1, IgG2, IgG3 and IgM), and result in cell, tissue or organ damage.

In the next issue, we will continue with discussion of Gell and Coombs hypersensitivities.
Dealing with Frustration or Impatience

By Erika Lawrence, PhD

WE ARE MORE than six months into a pandemic that has left us all stuck at home and under chronic strain. Most of us are home with our partners, kids, roommates or other family members with little meaningful face-to-face contact outside of those people. And, while we love the people closest to us, we have been trapped in the house for a long time! So, how can we best deal with the frustration or impatience we feel?

What Does Impatience or Frustration Look Like?

Impatience is the inability to stay calm while you’re waiting for an outcome you need or want. There are three ways frustration or impatience can loom large in our lives: when we are dealing with other people’s needs, wants, demands, or when others fall short of our expectations, needs or wants; when major and chronic stressors impact our lives; and when we face daily circumstances beyond our control.

Actually, impatience is closely tied to frustration. When we are impatient or frustrated, we may notice a variety of physical symptoms such as clenching our hands, tensing our muscles, breathing shallow and fast, and having difficulty sitting or standing still. Emotionally, we may feel irritable and/or nervous. Behaviorally, we may try to rush though tasks or make snap decisions.

How Can We Best Manage Our Frustration or Impatience?

First, specific people, words or situations are most likely to spark your frustration, so make a list of them. Becoming aware of when you are likely to feel frustrated is the first step toward learning how to manage your responses.

Then, next time you get frustrated or impatient:

1) Pay attention to your basic biological needs. Are you hungry? Tired? Dehydrated? Many of us get frustrated or impatient when we haven’t eaten or slept.

2) If that does not work, write down your specific thoughts, much like making a grocery list (e.g., “S/he is not listening to me.” “I’m being disrespected.” “I can’t handle this.”). Do not journal; that will only make it worse. Simply list the thoughts one at a time, then step back and look at the list.

3) Next, write down any other feelings or memories. Perhaps your frustration over this situation is reminding you of another time. Again, just list them (e.g., “memory of trying to home-school for math yesterday”; “memory of the last time someone cut me off in traffic”). Do not write out or take your mind through a whole narration of the memory; that will only make your frustration worse.

4) Let your thoughts and feelings be there! This is the hardest step for most. Many of us try to talk ourselves out of our thoughts or feelings or tell ourselves not to think/feel that way. Maybe other people have told you to stop being mad or to “snap out of it” or to “look on the bright side.” Does that work? Or does it leave you even more frustrated than before?

5) Think about your values. What matters to you? Is it treating others with respect, showing compassion to your children or asserting your own needs? We tend to lose sight of the things that truly matter to us and get “tunnel vision” when we get frustrated.

6) Manage your physical symptoms. You may want to do some deep breathing or muscle relaxation exercises, or scan your body for tension and then focus on tensing and relaxing that part of your body.

7) Choose a behavior. Without judging the options, brainstorm a list of choices you could make right now. Then choose which one(s) to do. For example, you could snap or yell at someone, but that may not be in line with your values. Or, you could try to remove yourself or avoid the situation, but that might not fix it.

Taking these steps is difficult when you are frustrated. And, while I can’t guarantee this process will always work, it may be worth trying.

References

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Immune Globulin Dosing: Actual Versus Ideal or Adjusted Body Weight

By Michelle Greer, RN, IgCN, and Joseph DiStefano, RPh, IgCP, CSP

RECENTLY, STUDIES have examined whether methods for determining dosing strategy for immune globulin (IG) treatment could result in cost savings and better clinical outcomes and quality of life. Specifically, the studies sought to answer whether dosing should be based on adjusted (AdjBW) or ideal body weight (IBW) versus actual body weight (ABW), especially in obese individuals.

IG Dosing Guidelines

IG therapy dosing is based on body weight in kilograms. Since one kilogram is equivalent to 2.2 pounds, pounds are divided by 2.2 to convert them into kilograms. Dosing also depends on a variety of other factors, including diagnosis, physician preference, tolerability and response to therapy. For some diagnoses, physicians may prescribe a high dose of IG therapy throughout the course of therapy. For instance, patients treated with intravenous IG (IVIG) for autoimmune blistering diseases typically start with 2 grams per kilogram of body weight each month and continue this dose for the entire length of treatment. For other conditions, dosing may start high and then taper as response to treatment is noted and/or reported. For instance, dosing for chronic inflammatory demyelinating polyneuropathy (CIDP) patients might begin at 1 gram per kilogram to 2 grams per kilogram and then be tapered to 0.5 gram per kilogram to 1 gram per kilogram monthly. Conversely, in some patients, dosing may begin low and remain that way for life. This is often the case for primary and secondary immune deficiency patients in whom the dose is typically around 0.4 grams per kilogram monthly. It should be noted that for CIDP and immune deficiencies, these dosing guidelines can be prescribed for both IVIG and subcutaneous IG administration.

Proposed New IG Dosing Guidelines

Currently, U.S. Food and Drug Administration-approved dosing for all IG products is based on ABW, and traditionally, ABW is used to determine the formal study of the processes of absorption, distribution, metabolism and excretion of medicinal products (e.g., how the body chemically processes medicines). As one publication states, the proposed pharmacokinetic differences between lean and obese patients and the opportunity to reduce costs has led to the proposal that obese patients should receive proportionally lower doses of IG once a certain threshold is reached. Indeed, initial data shows comparable outcomes using lower doses, so the question is: With a product such as IG, for which supply can ebb and flow based on plasma donations and other factors, is it a good idea to begin dosing patients based on IBW or AdjBW to better manage and protect product supply? Especially now, when plasma donations have decreased over the last many months due to the COVID-19 pandemic, this could be a beneficial way to prescribe less product yet achieve the same clinical outcomes. Additionally, depending on where patients are infused, there could be significant clinical cost savings.

In 2017, a study compared the effectiveness of using a precision-dosing strategy (IBW or AdjBW) versus a traditional-dosing strategy (ABW) for IVIG therapy in patients with hematologic malignancies or those undergoing hematopoietic stem cell transplant. The retrospective cohort
study included 209 IVIG patients, 125 of whom were dosed using ABW and 84 of whom were dosed using IBW or AdjBW. The primary outcome was infection rate within 30 days of IVIG administration. Secondary outcomes included 60-day infection rate, immunoglobulin G (IgG)-level response (IgG higher than 400 mg/dl) and realized and potential IVIG savings. Results showed no difference in 30-day infection rate between precision- and traditional-dosing strategies (15.5 percent versus 16 percent, respectively). Similarly, no difference was identified in the 60-day infection rate between groups (23.2 percent versus 19.8 percent, respectively). Levels of IgG obtained after IVIG repletion showed a treatment response rate of 86 percent in both groups. In addition, use of a precision-dosing strategy achieved $2,600 per month in institutional savings with the opportunity for an additional $4,600 per month in savings with complete adherence to this dosing strategy.

Another retrospective study conducted at MD Anderson Cancer Center examined all IVIG doses administered in adults ages 18 years and older from January 2011 through January 2016. Total body weight (TBW) and height at the time of administration were used to calculate prescribed dose (grams per kilogram), IBW and AdjBW. Three dosing methods were then analyzed: 1) Use of AdjBW if TBW was greater than 120 percent of IBW, 2) AdjBW for all doses and 3) IBW for all doses. Outcomes included potential IVIG use averted, direct drug cost savings and reductions in outpatient infusion times for each method. Of the 9,918 doses administered to 2,564 patients over five years, which represented an average usage of 75,994 grams per year, the study found that if dosing methods 1, 2 and 3 had been used, the annual use of IVIG would have decreased by 21.9 percent (16,658 grams per year), 24.2 percent (18,371 grams per year) and 35.9 percent (27,252 grams per year), respectively. This translated into average annual cost differences of $2.37 million, $2.62 million, and $3.89 million, and average annual outpatient infusion time savings of 841 hours, 920 hours, and 1,366 hours, respectively. The researchers concluded IVIG dosing optimization through use of alternative dosing weights represents a significant source of waste and cost reduction.

Although only a few studies have examined these different IG dosing guidelines, there are numerous unpublished anecdotal reports since dosing can be prescribed based on the prescriber’s preference or a pharmacist’s recommendation. Plus, it is always desirable for patients to be prescribed the lowest effective dose of any treatment. This is especially true for IG treatment for which infusions can be lengthy and the frequency of therapy can be years or lifelong. Additionally, with higher dosing, the potential for side effects becomes greater. Most notably, because the risk for cardiovascular adverse events is higher in obese individuals, lower doses that can generate the desired clinical response are always preferred.

### Improving Clinical Outcomes and Quality of Life

There are many benefits of IG dosing based on AdjBW or IBW, especially in obese patients to reduce the risk of adverse events, including serious ones. But, overall, benefits include use of the minimally effective dose to achieve the desired clinical outcome; improved quality of life due to shorter infusion periods; potential cost savings for patients due to smaller doses and less nursing time; overall cost savings for the healthcare industry; and optimization of the drug supply to help the greatest number of patients, especially during periods of decreased plasma donations. Indeed, adopting IG dosing guidelines based on AdjBW or IBW for all patients could magnify these benefits and may be worthy of consideration.

### Table. Calculating Body Weight

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<tr>
<th>Adjusted body weight (AdjBW)</th>
<th>(AdjBW) = IBW + 0.4(ABW – IBW)</th>
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<tbody>
<tr>
<td>Ideal body weight (IBW)</td>
<td>(estimate using kg):</td>
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<tr>
<td>• Males: IBW = 50 kg + 2.3 kg for each inch over 5 feet</td>
<td></td>
</tr>
<tr>
<td>• Females: IBW = 45.5 kg + 2.3 kg for each inch over 5 feet</td>
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### References

Hizentra Study Shows Positive Results for Expanded Infusion Parameters

In a clinical study that examined faster infusion rates and higher volumes than currently approved for CSL Behring’s Hizentra (immune globulin subcutaneous [human] 20% liquid) (SCIG) in patients with primary immunodeficiency disease (PI), researchers report positive results from the expanded infusion parameters for most patients.

In the multicenter, open-label, parallel-arm, nonrandomized Hizentra Label Optimization (HLLO) study, 49 adult and pediatric patients with PI who were on a stable dose of Hizentra therapy were enrolled in either a pump-assisted volume cohort (15), pump-assisted flow rate cohort (18) or manual push administration flow rate cohort (16). Assignments were based on prior experience with pump-assisted infusions or manual push administration infusions. The primary endpoint was the responder rate within each cohort and for each infusion parameter level; secondary endpoints examined safety and tolerability.

For the two pump-assisted cohorts, responder rates were defined as the percentage of patients who successfully completed greater than or equal to 75 percent of planned infusions. For the manual push administration cohort, responder rates were defined as the percentage of patients who completed at least 60 percent of infusions at each flow rate and who completed the full dose per scheduled infusion without interruptions for any reason.

Each cohort tested increasing infusion levels of Hizentra. After four consecutive weeks of receiving the same infusion level, qualifying participants moved to the next higher infusion level. Each infusion parameter level was tested for four weeks, after which responders were switched to the next level. The cohorts included:

- Pump-assisted volume cohort (weekly infusions with volume per injection site of 25 mL, 40 mL and 50 mL)
- Pump-assisted flow rate cohort (weekly infusions with flow rate per injection site of 25 mL/hour, 50 mL/hour, 75 mL/hour and 100 mL/hour)
- Manual push administration flow rate cohort (two to seven infusions per week with flow rate per injection site of 0.5 mL/minute (30 mL/hour), 1 mL/minute (60 mL/hour) and 2 mL/minute (120 mL/hour).

Results for the pump-assisted cohort results showed:

- Responder rates for the pump-assisted volume cohort were 86.7 percent (25 mL) and 73.3 percent (40 mL and 50 mL), while responder rates for the pump-assisted flow rate cohort were 77.8 percent (25 mL/hour and 50 mL/hour), 66.7 percent (75 mL/hour) and 61.1 percent (100 mL/hour).
- Dose and volume adherence rates were greater than or equal to 90 percent in all patients of the volume cohort, and 83.3 percent in patients in the flow rate cohort (less than 90 percent in three patients).
- Mean serum immunoglobulin G (IgG) trough levels (g/L) were similar between day one and end of the study for the volume cohort and the flow rate cohort.

Results for the manual push administration cohort results showed:

- Responder rates were 100 percent (0.5 mL/minute and 1 mL/minute) and 87.5 percent (2 mL/minute), with 98.5 percent to 100 percent of infusions completed per the planned schedule.
- Compliance rates were greater than or equal to 90 percent in all patients but one.
- Mean serum IgG trough levels were similar between day one and the end of the study.

The final analysis examined the rate of treatment-emergent adverse event (TEAE) frequency, type, intensity or duration across all three cohorts. Low rates of TEAEs per infusion were observed across all cohorts, with mild to moderate infusion site reaction being the most common. Specifically, TEAE rates per infusion were 0.145, 0.228 and 0.085 in the pump-assisted volume cohort, pump-assisted flow rate cohort and manual push administration flow rate cohort, respectively. There was no clinically meaningful difference in TEAE frequency, type, intensity or duration among the three cohorts, and rates of TEAEs per infusion did not increase with expanded infusion parameters.

“Understanding the needs of patients with PI and how we can improve their treatment experience by optimizing our therapies is a critical focus for us,” said Mittie Doyle, MD, vice president of research and development, Immunology Therapeutic Area at CSL Behring. “This study is yet another example of how we are delivering on our promise to patients with PI to help them better manage their health challenges.”

Medicare Part B Now Covers Cutaquig for Adult PI Patients

The Centers for Medicare and Medicaid Services (CMS) has modified the External Infusion Pump Local Coverage Determination to include Cutaquig 16.5%, Octapharma’s subcutaneous immune globulin (SCIG), for use with electric and mechanical pumps for adult primary immunodeficiency disease (PI) patients. The Medicare Part B coverage determination went into effect Sept. 6, 2020. For any item to be covered by Medicare Part B, it must be eligible for a Medicare benefit category, as well as meet all other applicable Medicare statutory and regulatory requirements. Since Cutaquig is administered through an external infusion pump, an item of durable medical equipment, it is covered for Medicare beneficiaries with PI under the Part B durable medical equipment benefit of the Social Security Act.

“Cutaquig is an important CMS-approved addition to the available supply of SCIG products for adult primary immune disease patients,” said Octapharma USA President Flemming Nielsen. “As the supply of immunoglobulin products faces challenges in the near future, the addition of Cutaquig to the list of products available for Medicare patients comes at the right time. Octapharma has increased its production of Cutaquig by more than 45 percent over the last year, so we have strong supply to meet patient needs.”

LOOKING BACK on the year 2020, one can’t help but acknowledge the stark differences from the beginning of the year to the present. Individuals with chronic illnesses have likely faced unprecedented situations and overcome obstacles never before thought possible. The COVID-19 pandemic started as a whisper with barely a mention in the news, then very quickly ravaged our country and our world. To date, the virus has infected more than 38 million people and taken more than one million lives globally.

Our frontline healthcare professionals have worked tirelessly to care for those in need, many sacrificing their own lives. With so much of our healthcare resources focused on controlling the spread of the virus and managing the acutely ill, patients with lifelong medical conditions have had to cope with issues generally left to healthcare professionals. Armed with limited information, patients treated with immune globulin (IG) therapy and their caregivers have to ensure ongoing medical needs are met and lifesaving treatment is continued.

Almost everything related to healthcare that was considered normal a year ago has had to be wholly reimagined, from healthcare visits and routine lab work to where, when and how IG therapy can be safely administered during the pandemic.
pandemic. With so much information and misinformation changing daily, life is chaotic. And, it is well understood that immunocompromised patients and those with pre-existing conditions are at higher risk of developing complications related to COVID-19. Indeed, patients who rely on IG therapy understand now how critical it is to maintain a consistent treatment schedule during this pandemic.

Because IG therapy is highly specialized and individualized, it requires a team of healthcare professionals, including physicians, pharmacists, nurses and others. And, it is more critical than ever for patients to be involved in the decision-making processes that impact their lives, pandemic or not. After all, who understands better the day-to-day struggles and successes of living with a lifelong therapy such as IG? When patients and their families are engaged in decision-making early and ongoing throughout treatment, good things happen. Patients have better outcomes and infusion experiences, and healthcare professionals are better informed and, therefore, can better care for patients. Following are strategies patients can use to advocate for themselves during these times.

The Power of Patient Advocacy Groups
The concept of patient advocacy is not new. In the 1920s, nurses were an integral part of establishing advocacy and patient rights in the nursing code of ethics. During World War II, Great Britain developed its own advocacy group known as the Citizen’s Advice Bureau. It wasn’t until the 1970s, however, that advocacy groups as we know them today were formed. There are currently hundreds of advocacy groups, many supporting patients and families who receive IG therapy. And, while becoming your own advocate in no way replaces these valuable resources, it does empower you to take a more informed and active role in your care.

Getting Involved
Whether you are new to IG therapy or have been receiving it for years, it is never too late to arm yourself with the knowledge you need to become your own advocate — which requires you gain enough information to be an active participant in decisions about your healthcare.

Patients diagnosed with an illness treated with IG therapy are barraged with unfamiliar medical terms and information from the moment of diagnosis, continuing throughout the treatment process. In general, patients rely on the expertise of their healthcare professionals to make decisions related to the course of therapy, route of administration, choice of product, site of care, etc. Patients may feel anxious and unsure of what is best for them. Feeling overwhelmed with a new diagnosis and the thought of lifelong therapy may prevent some patients from being able to make informed decisions. But, becoming active, gaining the knowledge you need and getting involved as early as possible in the decision-making processes can help to integrate this therapy seamlessly into your life.

Treatment Planning
Preparing for treatment with your healthcare providers before starting IG therapy provides a strong foundation for building a relationship of trust and mutual respect. This includes providing your full health history and discussing cultural, financial and lifestyle issues to help create a clear treatment path — one that suits your lifestyle and creates an atmosphere of confidence for all.

You can prepare for discussions with your healthcare team in advance by arriving with a list of questions or concerns to be addressed during each visit. Prioritize the list to optimize the limited time you have to spend with your provider.

The COVID-19 pandemic has demonstrated that telemedicine is an effective way of communicating with healthcare providers. If you are unable to see your provider directly, inquire about scheduling a virtual appointment, which can be an excellent option to share feedback with your healthcare team. If you receive care at home, utilize your specialty pharmacists and nurses as sounding boards to discuss how your therapy is going. It may be helpful to keep a treatment journal to document dates of infusions, side effects before or after treatments, blood draws, future treatments, etc.

Pre-Existing Conditions/Risk Factors
You must advise your healthcare provider if you have a history of diabetes, renal insufficiency, cardiac issues, deep vein thrombosis or blood clots in the lungs. Such critical information will help your provider determine the correct IG product or route of administration. Additionally, if you experience any of these conditions while receiving therapy, notify your provider immediately. All IG products come with certain risks, so communication with your healthcare team should be open and occur often. Oftentimes, the IG prescriber is not the primary care provider and may not be familiar with your full medical history. Consequently, if you see multiple specialists, make sure all providers know your health history and communicate with one another.
IG Brands

Currently, numerous IG brands are available, and they are not considered generic or interchangeable. All IG products contain essentially the same amount of antibodies, but the similarities stop there. Products vary in concentration, stabilizing ingredients and manufacturing processes, to name a few important differences. By thoroughly understanding your medical history, your provider can determine the IG product best suited to you.

Once an IG product is chosen, the current standard of practice is to keep patients on their optimal product. At some point, however, it may become reasonable or necessary to change brands. Reasons for product transitions include shortages, insurance mandates or changes, and product discontinuation. You may also request to change products if you are experiencing continued side effects despite efforts to reduce or stop them. When transitioning between different IG brands, special administration guidelines and protocols must be followed to ensure patient safety and reduce side effects. This further emphasizes the importance of clear communication between patients and providers.

The safety of IG products has been well established. However, should a problem with product arise, the patient notification system, a joint effort between manufacturers and consumers, notifies patients and other registrants of voluntary or mandatory recalls or withdrawals of products from the market. This system is confidential and easily accessible by visiting patientnotificationsystem.org. The U.S. Food and Drug Administration (FDA) also provides information about IG product shortages at www.fda.gov/vaccines-blood-biologics/safety-availability-biologics/cber-regulated-products-current-shortages.

Knowing which products have impending shortages can make the transition to an alternate product less stressful. Importantly, starting a new product is like starting therapy all over again, so the first dose of any product should be administered cautiously.

Route of Administration

IG products can be administered intravenously (IVIG) or subcutaneously (SCIG). IVIG is administered by a healthcare provider directly into the vein, usually every three to four weeks. When compared with SCIG, IVIG can be associated with a higher rate of side effects such as headache, feeling tired or unwell, chills and back pain.

SCIG can be administered by a patient or caregiver into the subcutaneous tissue (under the skin), usually more frequently (one or more times per week). SCIG has a lower rate of side effects described above, but it can cause injection site reactions such as swelling, itchiness, redness, etc., which typically resolve with time. In some cases, it may not be suitable for patients unable to self-administer.

There are advantages and disadvantages to both routes of administration. For example, a college student living away from home may desire optimal independence with therapy administration and choose SCIG. Conversely, a vision-impaired patient would most likely not be the right candidate for SCIG, but rather better suited for IVIG.

The route of administration can also be changed at any point during your course of treatment. Resistant side effects, skin site reactions or irritations, frequent infections or worsening of symptoms may be signs a change is needed.

Cultural Considerations

Inform your healthcare provider of any cultural considerations that may impact your treatment. Examples include religious beliefs about receiving blood products, religious or spiritual practices, language barriers or literacy issues. Communicating about these issues early will help avoid misunderstandings.
Side Effects and Adverse Reactions
Talk with your healthcare team about possible side effects associated with IG therapy and management strategies should they occur. Headache, flu-like symptoms, fatigue and skin site reactions are the most common side effects patients experience. Some side effects may occur during the infusion; others may occur hours or days after the infusion. Many IVIG reactions can be addressed by drinking more fluids prior to infusions, slowing infusion rates and administering premedications. Side effects associated with SCIG can be managed by proper needle insertion technique and changing needle length, rate of infusion and other factors.

While patients may believe side effects are inevitable, this is not necessarily true. Speaking up early and often about how you are feeling before, during and after treatment is critical. Sometimes interventions as simple as staying well-hydrated before the infusion or slowing the infusion rate can substantially impact how treatment is tolerated.

Evaluating Therapy Effectiveness
Setting expectations before starting IG therapy is critical. Talk with your healthcare team about when an improvement in health might be noticed, and what tools are used to evaluate the effectiveness of therapy. Determine a follow-up schedule with your provider, and make appointments in advance. Inquire if routine lab work is needed, and schedule these appointments at predetermined intervals.

Getting to Know Your Infusion Specialist
Because IG therapy is such a specialized field, the healthcare professional treating you should be familiar with its complexities. And, getting to know your infusion specialists is a necessary step in becoming your own advocate. Nursing experience and familiarity with IG products and administration can vary widely. The Immunoglobulin National Society (ig-ns.org) offers the only nationally recognized certification exam for IG clinicians: the Ig Certified Nurse (IgCN) and the Ig Certified Pharmacist (IgCP). Clinicians who seek and obtain certification in this field have highly specified knowledge and skills. It’s important to inquire if your treating nurse has this certification, about his or her level of experience with IG therapy and how often he or she administers these products. These questions should be asked routinely and freely of any nurse administering IG therapy.

Communicating with Your Team
Remember, your team is there to support you. Becoming your own advocate makes you the captain of your team, which will give you confidence and control throughout the turbulence of managing your illness and treatment. As team captain, you must keep your appointments, whether in person or virtually, and be well prepared with the required information to make your visits more productive. Leaving appointments with all questions answered and a plan for the future puts everyone involved at ease.

Becoming your own advocate requires practice and persistence. As this may be an entirely new way of communicating with your healthcare team, and change can be difficult, start slow if you must. Initially, begin discussions with the member of your healthcare team with whom you feel the most comfortable. Ask questions, express concerns and make suggestions. Take control of your care; it is your time.

References
3. Immunoglobulin National Society. IgNS Standards of Practice, Edition 2.1

RACHEL COLLETTA, BSN, CRNI, IgCN, VA-BC, is director of educational resources at the Immunoglobulin National Society, a professional organization dedicated to the advancement of immune globulin therapy across clinical indications and areas of practice.
Protecting Mental Health When Dealing with Chronic Illness

Some simple yet effective techniques can help patients relax and improve their mental and emotional state.

By Surayyah Morris, PharmD
MORE THAN 45 million Americans are living with a mental illness.\textsuperscript{1,2} As one of the most common causes of disability, it is important to address what causes mental illness and how to manage it. This is especially true for those with a chronic illness, which increases the chances of developing a mental illness such as depression and anxiety. Indeed, the connection between physical health and mental health is significant, which is another reason proper mental healthcare is important when also battling a chronic illness.

The physical pain of having a chronic illness alone is enough to drive anyone crazy. Add in complicating factors such as family, friends, work, school, medications, healthcare and everyday life, it can become overwhelming to deal with how much there is to handle. That’s why, despite life’s ongoing situations, people must find a way to balance mental health and navigate wide-ranging feelings and emotions appropriately. And, while that’s easier said than done, it is still possible to live a happy, joyful, positive life and manage illness effectively. Through trial and error, speaking to other patients and consulting with doctors, I have gathered beneficial resources that will help many chronic illness warriors begin to live a balanced life by incorporating practices that are simple yet productive to keep them on the right mental track.

The following are some suggestions to help rewind and settle your mind. Not everything works for everyone, but I encourage you to try them first and make adaptations to better fit your needs and your lifestyle.

First, Breathe!

Inhale for four seconds, hold it for four seconds and exhale for four seconds. Repeat.

Breathing is the gold mine for controlling emotions. Notice that when you are angry, frustrated or nervous, your breath accelerates. When you are calm, relaxed and stress-free, your breath is deep and long. Controlling your breath not only helps you to physically reduce tension by forcing your heart rate to slow down, it also helps to mentally relax. There are many variations of breathing techniques, but below are a couple of the simplest exercises to help get you started.

Belly breaths:
1) Lie on your back on a flat surface or sit in an upright position with your back erect, knees bent and feet planted firmly on the floor.
2) Place your hand on your belly.
3) Inhale with your hand on your belly, ensuring you feel the rise of your belly only (not up through your chest, but out through your belly).
4) Exhale by blowing the air back out and letting your belly deflate back to its original position.
5) Repeat for just a few minutes.

4-4-4 breathing:
1) Inhale through your nose for four seconds.
2) Hold your breath for four seconds.
3) Exhale out of your mouth for four seconds.
4) Repeat.

Taking a few moments out of your day to practice breathing or to reset after a stressful situation is something that is simple, yet super beneficial to your mental health.

Gratitude

Gratitude is the antidote to negative emotions like fear, hate, anger and sadness. The practice of gratitude rewires your brain and shifts your mindset to a more positive one. Finding things to be grateful for forces you to focus on what is good and transforms your perception of undesirable situations. It may seem like another daunting task to add to your daily to-do list, but I promise a daily gratitude will shift your mood in the right direction. All it takes is a single gratitude each day, although you can have as many as you’d like. It can be any time of day. You can log your gratitude in a diary, an app or just keep it on your mind for the day. And don’t stress yourself trying to think of an over-the-top, holy, super meaningful thing that makes you grateful. It can be something as simple as clothes to wear, a car for transportation or your amazing caregiver. From time to time, my gratitude is actually my immune globulin medication because without it, I’d be a complete train wreck, so I’m always grateful for access to it.

Meditate

Meditation is a technique that can positively impact your mental health. Daily practice increases your dopamine levels, which positively affects motivation, concentration and pleasure responses. Because there are numerous meditation techniques and exercises, it will take trial and error to find the best one for you, but it will be worth the mind-shifting benefits. You can begin with guided meditations using an app like Headspace, Balance or Calm. Guided meditations explain what to do, are easy to follow and are customizable with technique and time. As you become more comfortable, you can begin unguided meditation on your own. Start with just a few minutes, and work your way up to longer meditations as needed.
Yoga

If you asked me five years ago to do yoga, I’d laugh hysterically and politely decline. Yoga was the single most boring thing I could ever think of doing — like the ultimate trigger to an underlying attention deficit disorder every time I tried it. Now, I can’t live without it because I have experienced its mental and physical benefits, and it has improved several aspects of my life. The benefits of yoga include and certainly aren’t limited to:

- Easing depression and anxiety
- Improving balance and flexibility
- Reducing inflammation
- Improving circulation and heart health
- Reducing stress
- Improving mood and fatigue
- Reducing chronic pain

Finding time to incorporate yoga into your week can have a positive impact on your overall well-being, especially your mental health. Use YouTube videos for guidance in the comfort of your home, or attend yoga classes at your local gym if you want a more social environment (Zoom classes count if you’re not ready to go out in public yet).

Self-Care

Become more aware of what is worth expending energy toward. Many times, we become wrapped up in things we feel we are supposed to be doing at the expense of our sanity, energy and well-being. It is just as important to take care of yourself as it is to take care of everyday responsibilities and even others. What will you pour into others if you have nothing left in you to pour? So, what does self-care look like?

- Finding a hobby you enjoy
- Taking a walk to clear your mind and get your body moving
- Pampering yourself with a nice bath
- Performing simple daily hygiene (brushing teeth, washing up, combing hair, getting facials)
- Meditating, yoga, breathing exercises, mindfulness
- Saying “no” when necessary

Create a Routine and Set Goals

Creating a routine is an excellent way to promote good mental health. The routine should consist of things you would do every day consistently. Incorporate a daily goal into your routine to help define an activity that gives you a sense of purpose. Your routine can look something like:

- Washing your face and body in the morning and/or evening
- Meditating for five minutes
- Eating a balanced meal
- Exercising/practicing yoga
- Taking your medications on time
- Organizing your to-do list
- Acknowledging your gratitude for something
- Getting some fresh outside air or going for a walk

Setting goals is another way to ensure you establish a sense of purpose. If you have a chronic illness that keeps you more sedentary and homebound than the average person, then it is beneficial to create goals. Short-term goals are great to accomplish on a daily basis. For example, cross two things off your to-do list. Long-term goals are great to work toward over an extended period and provide a sense of accountability. For example, take an online course to learn a new skill. You can begin by creating goals for your health, career and relationships. Committing to your goal and achieving it is a rewarding experience and sure to bring you a great sense of accomplishment. It boosts your mood and prevents you from being idle. But, remember, your goals should be SMART:

- Specific
- Measurable
- Attainable
- Realistic
- Time-bound
Laugh

When I used to have long hospital stays, my youngest sister would always visit to keep me company. She would bring her iPad and make me watch these random “try not to laugh” videos on YouTube (similar to “America’s Funniest Home Videos”) except they were never-ending; she’d play them for hours. I realized just how much joy something so simple created for her. It was only a matter of minutes before I was giggling for hours right along with her, and I genuinely felt better. Soon enough, I started watching on my own when I needed to add some joy to my day.

Not only did I watch silly videos, I also started watching motivational videos. I just searched for a motivational video and listened to the positivity and encouraging words. Give it a try, and after a few minutes, you’ll begin to feel encouraged and motivated to keep pushing forward, to tackle daily tasks or to just have a more positive mindset. This doesn’t have to be a daily habit. Whenever you feel yourself on a downward mental spiral, head to YouTube and search for motivational videos and pick any one. I guarantee you will feel happier afterward, even if just for the moment. Sometimes all you need is that little push to keep you going through the rest of the day.

Netflix

Netflix is a necessity. And by Netflix, I’m referring to all streaming services that aren’t cable: Hulu, Firestick, Roku, Amazon Prime, etc. To keep your mind off of reality, streaming services provide an endless number of shows and movies for you to indulge in on your not-so-good days or if you just need to take a mental health day and keep your stress levels low. The art of distraction in this way is useful to allow your mind to focus on something entertaining and lets your emotions decrease in intensity if you feel overwhelmed.

Talk to Someone

Your mental health is important and cannot always be managed alone. Talk to someone you trust, or seek help from a professional like a psychologist, psychiatrist, therapist or counselor. Just an hour a week will work wonders after a short period of time. Expressing your concerns or just speaking about your life provides an outlet so you’re not holding on to your emotions. If you prefer to engage in other ways, there are several organizations dedicated to maintaining your mental health that provide resources for all ages and backgrounds:

• National Alliance on Mental Illness (www.nami.org)
• Mentalhealth.gov
• Ok2talk.org
• The Mighty (themighty.com)

If you are not ready to speak to someone, journaling your thoughts is another great outlet. Writing down what’s on your mind is a safe and effective way to clear your head without actually speaking to a person. While it does not replace talk therapy, journaling is an alternative to help you release thoughts that may be overwhelming you.

Benefits of Addressing Mental Health

While it may be difficult to address mental health concerns, the benefits of tackling any issues or preventing them in the first place certainly outweighs the risk of adding the stress of poor mental health. Remain conscious of your mental and emotional state, and always be honest with yourself and others about how you are feeling.

References


SURAYYAH MORRIS, PharmD, is an autoimmune small fiber neuropathy patient from Central Florida. As a medication therapy management and pain management specialty pharmacist, she enjoys supporting patients with chronic pain and chronic conditions to help find balance and improve quality of life.
FACE MASKS ARE the new norm. The Centers for Disease Control and Prevention (CDC) is calling on Americans to wear face masks to reduce the spread of COVID-19, and face masks are now mandated at indoor public spaces, as well as outdoor public spaces where social distancing of six feet is not possible, in 30 U.S. states (with exceptions for certain ages and conditions). Yet, the fashioning of face masks began in mid-March when the lack of personal protective equipment (PPE) for frontline workers made headlines — far before CDC’s recommendation. Feeling the need to do their part to help essential workers fight the pandemic and keep themselves, their friends and families safe, people began making face masks at home, often creating their own patterns with elaborate designs and some with size adjustments. And that was just the beginning. As of this writing, some 15 major retailers, including Old Navy, Disney and Anthropologie, have debuted their own lines of stylish masks. But, while face masks are a part of today’s wardrobe, do they all meet the criteria to qualify as PPE? And, are individuals properly donning them to ensure they are protective against the coronavirus?

Should People Wear a Face Mask?
Unfortunately, at the start of the pandemic, experts were unaware of the extent individuals with COVID-19 could spread the virus before symptoms appeared. It was also unknown that people could have COVID-19 and be asymptomatic, and that both those with and without symptoms could unknowingly spread the virus to others. In fact, a new study found 78 percent of people who tested
positive for COVID-19 showed no symptoms when they were swabbed.\textsuperscript{3} However, once these discoveries were made, the World Health Organization and CDC recommended wearing face masks to slow the spread of the virus.\textsuperscript{1}

Many people believe wearing a mask is to protect themselves. But, while wearing a mask can provide personal protection against inhalation of harmful pathogens and particulates, they are mostly intended to protect people around them. This is known as source control. “Masks are used to contain your respiratory secretions and protect others from you,” explains Leann Poston, MD, medical content contributor for Invigor Medical.\textsuperscript{3} Most importantly, masks help protect people with higher risk of severe illness from COVID-19 such as older adults, obese individuals and those with compromised immune systems, as well as workers who frequently come in contact with other people such as in restaurants, stores and healthcare offices.\textsuperscript{4}

Sadly, myths about wearing face masks are still running rampant, even ones maintaining wearing a mask is dangerous. The first claim is that face masks make people breathe too much carbon dioxide and not enough oxygen. Yes, the cells in a person’s body need oxygen, and with each breath, a person uses a little bit of that oxygen and exhales a little bit of carbon dioxide. So, if a person were trapped inside of an enclosed space for a long time, then it would be possible for the carbon dioxide levels to rise in that space. But, the tiny amounts of carbon dioxide that a person breathes out are not trapped by a cloth mask or a surgical mask. Masks can sometimes make it difficult to breathe, especially for people with certain health conditions, but that is not a result of carbon dioxide buildup. The second claim is that people can get sick from breathing their exhaled viruses. And, this one really doesn’t make any sense because if a person is breathing out the COVID-19 virus, that person is already sick, so how could the mask pose any further danger?\textsuperscript{5}

Indeed, the proof for why people should wear a face mask is in the science. In May, a study published in The Lancet analyzed 172 studies from 16 countries and six continents to determine ways to slow the spread of the pandemic, as well as other coronaviruses, including SARS and MERS. Specifically, the researchers looked at three measures: face masks, social distancing and eye protection. They found that without a mask, social distancing or any other preventive measures, the risk of transmitting the coronavirus is 17.4 percent. However, with a mask or respirator, the risk dropped to 3.1 percent. It should be noted, though, that the study found social distancing to be the most effective strategy. With less than 1 meter (3.2 feet) of distance and no other protective measures, the risk of transmission was 12.8 percent, yet with more than 1 meter, the risk was only 2.6 percent. Eye protection offered benefit with little risk.\textsuperscript{6}

In July, CDC published details of a study that found two hairdressers in Springfield, Mo., who were infected with coronavirus did not infect any of 139 clients they worked with, probably because they wore face masks. According to a statement by CDC, “At this critical juncture when COVID-19 is resurging, broad adoption of cloth face coverings is a civic duty, a small sacrifice reliant on a highly effective low-tech solution that can help turn the tide favorably in national and global efforts against COVID-19.”\textsuperscript{7}

Who Should Wear a Face Mask?

Who should wear a face mask depends on official state mandates and recommendations by CDC.

As of the end of July, 30 states had statewide mask mandates, 18 states had some mask mandates and only two states had no face mask requirements (Idaho and South Dakota). In the 18 states without statewide mandates, many cities and counties have enacted mandates.\textsuperscript{8}

Many of the states requiring face masks follow CDC’s guidelines (see below); however, others have both looser and stricter mandates. For instance, Arkansas requires only individuals 10 years and older to wear face masks, whereas California now requires all individuals to wear them, even those younger than 2 years old.\textsuperscript{9} Individuals should check with their local cities, counties and states to determine their specific mandates.

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**Sadly, myths about wearing face masks are running rampant, one of which is that wearing a mask is dangerous.**

CDC has published specific guidelines about who should wear a face mask. For the general public, CDC recommends all people 2 years of age and older wear a mask in public settings, especially in areas with high transmission levels. The guidelines encourage people to wear masks in public places, such as grocery stores, pharmacies and other essential businesses. The recommendations also suggest wearing masks when social distancing is not feasible, such as on public transportation or in crowded settings. While face masks can help slow the spread of COVID-19, it is important to remember that vaccines are the best way to prevent the virus from spreading. Effective vaccines are currently available for COVID-19, but they require ongoing research and development to be effective against new variants of the virus. In conclusion, wearing face masks is an essential public health measure during the COVID-19 pandemic, and they will continue to play an important role in controlling the spread of the virus until a vaccine is widely available.---
settings and when around people who don’t live in their household, especially when other social distancing measures are difficult to maintain. CDC also recommends people who know or who suspect they might have COVID-19 to wear a mask when around people or animals even within their own homes. In addition, those caring for someone who is sick with COVID-19 at home or in a non-healthcare setting should also wear a mask.

Because wearing a mask may not be possible in every situation and for some people, CDC recommends adaptations and alternatives be considered. Following are some examples:

- People who are deaf or hard of hearing (or those who care for or interact with someone who is hearing impaired) could consider using a clear mask. People with intellectual and developmental disabilities, mental health conditions or other sensory sensitivities are encouraged to consult with their healthcare provider for advice about wearing a mask.

- For younger children who may have trouble wearing a mask properly, particularly for an extended period, wearing a mask could be prioritized at times when it is difficult to maintain social distancing. People engaging in high-intensity activities such as running that would have difficulty breathing when wearing a mask should consider a location with greater ventilation and air exchange where it is possible to socially distance. And, people who work in settings where a mask may increase the risk of heat-related illness or cause safety concerns should consult with an occupational safety professional to determine which mask would be most appropriate.

- There are also people who should not wear a mask, according to CDC. These include children younger than 2 years old; anyone who has trouble breathing; anyone who is unconscious, incapacitated or otherwise unable to remove the mask without assistance; and those who engage in activities that may cause the mask to become wet such as when swimming, which can make it difficult to breathe.4

Are All Face Masks Created Equal?

It’s no secret that at the start of the COVID-19 pandemic, PPE was hard to come by. Just conducting a search for face masks on Amazon.com turned up “unavailable; reserved for healthcare personnel.” This shortage, of course, prompted people to begin making their own face masks not only to distribute to essential workers, but also for friends and family. Some individuals even started face mask businesses, and then large companies joined in. But, it’s important for people to understand that not all face masks are deemed equal, with some providing better protection than others, and others more dangerous than wearing nothing at all.

According to CDC, individuals should wear cloth face masks, reserving surgical and N95 masks for healthcare workers. However, now that there is less of a shortage, people are seen in various locations wearing the latter. So, what exactly are the differences between the types of face masks?

Fabric (cloth) masks. A fabric mask is intended to trap droplets that are released when the wearer talks, coughs or sneezes, but they offer the lowest protection from particles and fluid in the immediate environment. They are easy to find or make and can be washed and reused. While all kinds of materials can be used to make them, the best material is a tight-weave 100 percent cotton cloth. The natural fibers in cotton tend to have more three-dimensional structure than synthetic fibers, which are smoother, says Christopher Zangmeister, PhD, a researcher at the National Institute of Standards and Technology who co-authored a new study that tested how well dozens of different materials filtered. According to Dr. Zangmeister, while two synthetics, including one that’s 100 percent polyester, did well, most synthetics ranked near the bottom.1,10

The edges of these masks are not designed to form a seal around the nose and mouth, which means leaks may occur around the edges of the mask when a user inhales, and the masks may not effectively filter small particles from the air. Unlike other categories of masks intended for medical purposes (surgical masks and N95 respirators), these masks are not required to meet standards for fluid resistance, particle and bacterial filtration efficiency, flammability and biocompatibility before distribution.11

In addition, a good option for fabric masks is a built-in pocket where a filter can be placed. Polypropylene filters derived from plastic are best because they can hold an electrostatic charge, meaning they use the power of static electricity. The static-cling effect traps incoming and outgoing droplets, says May Chu, MPH, an epidemiologist at the Colorado School of Public Health who co-authored a paper on the filtration efficiency of household mask materials. According to Chu, a two-layer tight-weave cotton mask alone can filter out about 35 percent of small particles, but adding a polypropylene filter can boost filtration efficiency by as much as another 35 percent.
Actually, how well the mask seals to a person’s face will also determine its ability to filter out particles. It’s best to choose masks that cup tightly to the face or those that have pleats or folds. Masks with a flat front design are less effective.10

*Surgical masks.* Also called a medical mask, a surgical mask is a loose-fitting disposable mask that protects the wearer’s nose and mouth from contact with droplets, splashes and sprays, and that also filters out large particles in the air. Many are made of paper, although some are made with polypropylene.1,10

Surgical mask edges are also not designed to form a seal around the nose and mouth, again resulting in possible leaks. While surgical masks must meet certain standards for fluid resistance, flammability and biocompatibility before distribution, they may not effectively filter small particles from the air. However, they do provide additional protection to the wearer against large droplets, sprays or splashes of bodily or other hazardous fluids. They also protect individuals from the wearer’s respiratory emissions.11

While these masks haven’t been approved by the U.S. Food and Drug Administration (FDA) to protect against the coronavirus, they can provide protection when N95 masks are unavailable. The important thing to understand about surgical masks is they, as well as fabric masks, don’t protect wearers when they are in prolonged contact with others.1,10

*N95 masks.* An N95 mask is a type of respirator that offers more protection than a surgical mask because it can filter out both large and small particles when the wearer inhales. They are made of many layers of fine polypropylene fibers, which use the power of static electricity to filter out 95 percent of very small (0.3 micron) particles. Some N95 masks have valves that make them easier to breathe through, but this allows unfiltered air to release when the wearer exhales, so some locations have banned them.1,10

N95 masks are designed to fit tight to the face and to form a seal around the user’s nose and mouth.11 In fact, healthcare workers must be trained and pass a fit test to confirm a proper seal before using an N95 respirator in the workplace. And, while they are intended to be disposable, testing is being conducted to find ways to disinfect them for reuse.1,10

All N95 masks are tested and certified by the National Institute for Occupational Safety and Health (NIOSH) based on physical and performance characteristics, including filtration efficiency.11

*Surgical N95 respirators.* A surgical N95 respirator is both certified by NIOSH as an N95 respirator and cleared by FDA as a surgical mask. It helps reduce particles inhaled by the wearer (like a respirator) and reduce particles expelled by the wearer, and it resists fluids (like a surgical mask). In addition, a surgical N95 respirator must meet the same FDA standards for fluid resistance, flammability and biocompatibility as a surgical mask before commercial distribution.11

In August, a new study conducted by Duke University researchers used a simple optical measurement method to evaluate the efficacy of masks. The method is inexpensive and can be built and operated by nonexperts, allowing for rapid evaluation of mask performance during speech, sneezing or coughing. In the study, researchers created a black box, which had laser beams that created a sheet of light inside the box, with a hole in the front of the box for individuals to spit into. A person then wears a face mask and speaks into the direction of the laser beam in the black box. Droplets that propagate through the laser beam scatter light, which is recorded on a cell phone camera.

Fourteen commonly available masks or mask alternatives, one patch of mask material and a professionally fit-tested
Hizentra is an Ig* therapy that provides proven PI protection with the convenience of self-administration, so you can focus on everyday living

*Ig=immunoglobulin

Important Safety Information

WARNING: Thrombosis (blood clots) can occur with immune globulin products, including Hizentra. Risk factors can include: advanced age, prolonged immobilization, a history of blood clotting or hyperviscosity (blood thickness), use of estrogens, installed vascular catheters, and cardiovascular risk factors. If you are at high risk of blood clots, your doctor will prescribe Hizentra at the minimum dose and infusion rate practicable and will monitor for signs of clotting events and hyperviscosity. Always drink sufficient fluids before infusing Hizentra.

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

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

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

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

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

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

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

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

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

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

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

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

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

You can also report side effects to CSL Behring’s Pharmacovigilance Department at 1-866-915-6958.

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

BRIEF SUMMARY OF PRESCRIBING INFORMATION

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

WARNING: THROMBOSIS

See full prescribing information for complete boxed warning.

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

INDICATIONS AND USAGE

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

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

For subcutaneous infusion only.

DOSAGE FORMS AND STRENGTHS

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

CONTRAINDICATIONS

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

WARNINGS AND PRECAUTIONS

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

ADVERSE REACTIONS

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

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

DRUG INTERACTIONS

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

Based on March 2020 revision

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N95 mask (Figure) were tested with this measurement method. Here are the results:

- First place: fitted N95 mask that didn’t have an exhalation valve
- Second place: three-layer surgical mask
- Third and fourth places: cotton-polypropylene-cotton mask and two-layer polypropylene apron mask
- Fifth through 11th places: four different two-layer cotton pleated masks and one one-layer cotton pleated mask (number seven was another type of N95 mask with an exhalation valve, and number nine was a one-layer Maxima AT mask)
- 12th place: knitted mask
- 13th place: bandana
- 14th place: fleece mask

The researchers concluded the number 12th and 13th finishers could actually be worse than wearing no mask at all.\textsuperscript{12,13}

**How to Properly Wear a Face Mask**

No matter the type of face mask, there are some clear do’s and don’ts about how to properly wear one. First, it’s difficult for some to get used to wearing a face mask, so wearers should start slow by wearing it at home for a short time and slowly increase the time until it feels more comfortable. Second, if the mask is uncomfortable or if it is too difficult to breathe through, other mask options should be considered.

Following are some specific tips for how to put on and take off a face mask:\textsuperscript{14}

- Wash or sanitize hands before and after putting on and taking off the mask.
- Place the mask over the mouth, nose and chin.
- Tie the mask behind the head, or use ear loops to make sure it’s snug with no open flaps.
- If the mask has a wired side meant to adhere to the shape of the nose, press down on it over the nose for a more comfortable fit.
- Don’t touch the mask while wearing it.
- Switch to a clean mask if it gets dirty or wet.
- Remove the mask by untying it or lifting off the ear loops without touching the mask or face.
- Wash hands immediately after removing the mask.
- Regularly wash the mask with soap and water by hand or in the washing machine.
- If the mask is not washed immediately after use, place it in a clean resealable plastic bag.

In addition, wearing a mask incorrectly renders it useless. Following are seven mistakes often made:\textsuperscript{15}

- Not paying attention to which side of the face mask goes outside versus inside
- Wearing the same face mask all day
- Pulling down the face mask to speak to someone
- Touching the face mask with gloves on
- Not covering the nose while wearing a face mask
- Wearing a damp face mask
- Wearing the wrong kind of face mask

**The Secondary Consequences of Face Masks**

While the public health benefits of wearing face masks far exceed the social costs, says Tyler Cowen, a professor of economics at George Mason University and writer for the blog Marginal Revolution, “If we want mask-wearing to be a stable norm, we may need to protect against or at least recognize some of its secondary consequences, including the disorientations that masks can produce.” For instance, the majority of masks hide individuals’ mouths, which means their smiles, frowns and other nonverbal gestures and expressions that connect people can’t be seen. “If nothing else, our smiles cannot be seen under our masks, and that makes social interactions feel more hostile and alienating, and it may lower immediate levels of trust in casual interactions,” explains Cowen.\textsuperscript{16}

The Reveal mask by ClearlyHuman is designed to ease some of the secondary consequences of wearing face masks.
Indeed, it is often cited that 55 percent of communication is visual. That figure comes from research conducted in 1971 by Albert Mehrabian, PhD, professor emeritus of psychology at the University of California, Los Angeles, who is best known for his publications on the relative importance of verbal and nonverbal messages. Dr. Mehrabian’s research concluded communication, on a face-to-face basis, is thought to consist of three separate elements: words (what is said), tone of voice (how the words are said) and body language. Words (the literal meaning) account for 7 percent of the overall message; tone of voice accounts for 38 percent; and body language accounts for 55 percent. Therefore, through face-to-face communication, the nonverbal communication becomes the most powerful mode of communication when conveying feelings or attitudes.\textsuperscript{17}

A quick Internet search for clear masks turns up just a handful such as The Smile Mask (rafinova.com), Italian Smile Face Mask and Masked with a Smile (etsy.com) and ClearMask (theclearmask.com). However, it should be noted that the clear masks available today are nonmedical grade, which means they are essentially the same as a cloth face covering, except with a piece of flexible plastic or vinyl in the center to show the mouth. And, while they can provide some protection as outlined above, they have a much lower standard of filtration, protecting against only 50 percent to 70 percent of particles.

A clear mask that is set to debut in December is the Reveal mask by ClearlyHuman (ClearlyHuman.com). Reveal is a transparent protective mask that consists of four components: 1) a clear, durable, anti-fog polycarbonate shell; 2) a soft silicone gasket for a comfortable fit and seal; 3) an adjustable and easily replaceable strap; and an N95-level filter cartridge for maximum protection. It comes in three sizes: small (3.3 inches), medium (3.7 inches) and large (4.1 inches).

According to Richard Holbrook who is in charge of the company’s product design and development and brand strategy, the mask is being created by a team with decades of experience in product design, engineering and manufacturing. When the pandemic hit, he and colleagues whose core business was put on hold wanted to apply their talents and resources to help people through the pandemic and beyond. The company received a PPE loan to invest in the Reveal mask, as well as keep the team working. Holbrook, like so many people, is at high risk of developing severe complications from COVID-19 and, therefore, has to be hypervigilant about protecting himself. The Reveal mask is an N95 mask, but what really sets it apart from other N95 masks is its design. Rather than locating the filter on the front of the mask, which allows for the air that is being breathed to come straight in and out, the Reveal filter is located on the bottom next to the chin so that breath is inhaled and exhaled up
and down to improve protection. “We decided to change the course of exhalation and have it go down and your inhalation up so the mask is a deflector, as well as a filtration device,” explains Holbrook. “We hope to do better than the N95 rating.” In addition, the mask is designed to be reused, and can be safely and easily cleaned. Even the medical-grade silicone rubber seals can be cleaned with mild soap and water.

The company is currently applying for the CDC’s NIOSH certification to verify it achieves an N95 filtration rating, and it plans to apply for FDA approval of the mask as a respirator. Yet, while it is unlikely the Reveal mask will have this certification and approval by the time it is available in early December due to government regulator backlogs, they will have a certifying rating confirmed by independent lab tests.

It Comes Down to Social Responsibility

Wearing a face mask is one of the easiest things people can do to prevent the spread of COVID-19, and face masks are mandated in 30 states. According to a Pew Research Center survey, more than 85 percent of U.S. adults reported wearing a face mask most of the time in stores and businesses during the month of August. Yet, there remain some who refuse to wear one.

A recent study conducted in Brazil, which has the most cases of COVID-19 after the U.S., found people who reported “antisocial traits” such as low levels of empathy and high levels of callousness and risk-taking were less likely to comply with COVID-19 prevention measures such as wearing a mask and social distancing. The study authors suspect that people who have low levels of empathy and antisocial tendencies may have fewer concerns about exposing themselves and others to risks. A separate study of U.S. adults found those who display high levels of antisocial traits were more likely to endorse behaviors that would “knowingly expose others to risk.” Conversely, people who are empathetic believe it’s their social responsibility to self-isolate, practice hand hygiene and wear a mask, the Brazil study authors wrote.

But while all types of face masks provide some amount of protection, understanding which types of face masks provide the most protection is paramount, especially for high-risk populations. “Do you want to take a 5 percent risk or a 45 percent risk?” asks Holbrook. “Since this is an invisible killer, no one can look around and ask ‘What’s my risk level now?’”

Nevertheless, wearing a face mask is only one way to protect against COVID-19 infection. “We’re all experiencing pandemic weariness, and we are going to start letting our guard down,” says Holbrook. “I would really caution anybody who considers themselves at a higher risk to try to remain vigilant and keep their guard up and not let fatigue get the better of them.” Indeed, masks are not a catch-all solution. The best protection is wearing a mask, social distancing and washing hands frequently. “Rather than viewing [mask wearing] as the sole way you’re staving off infection, it should be seen as a tool in a larger group of anti-infection measures,” says David Cutler, MD, a family medicine physician at Providence Saint John’s Health Center in California.

References


coronavirus-coronavirus?


RONALE TUCKER RHODES, MS, is the editor of IG Living magazine.
Understanding Cytopenias in Primary Immunodeficiency Diseases

Cytopenias are common comorbid conditions in patients with primary immunodeficiency diseases, especially in those with common variable immune deficiency, and diagnosis can be challenging. And, while most respond to typical treatments, it can be difficult to provide definitive therapy in some.

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

IN GENERAL, primary immunodeficiency diseases (PIs) are characterized by dysfunctions of the immune system that result in undue susceptibility to infections. The majority of PIs involve some form of antibody deficiency. In the past, it was believed immunodeficiency was a consequence of decreased immune function and autoimmunity was essentially the opposite caused by overactive immune function. However, as the specific genes involved have been identified, it is now recognized that mutations in the same gene can result in immunodeficiency in some persons, autoimmunity in others and immunodeficiency and autoimmunity in the same individual. Therefore, with certain gene mutations, antibody deficiency and cytopenias may occur together due to altered gene function. Hence, these are not mere “low immunity” and “high immunity” situations.

What Are Cytopenias?

The word “cytopenia” originates from the Greek language; essentially, “cyto” means “cells,” and “penia” means “lack of.” So, when someone develops a cytopenia, it means that person is lacking specific cells. Most people recognize the term “anemia” has to do with low red blood cell (RBC) levels or counts. But, cytopenia is an encompassing word for each of the cell types that can be decreased. Leukopenia (or leukocytopenia) is caused by a general decrease in leukocytes or white blood cells (WBCs). Neutropenia is caused specifically by a reduction in neutrophils (a specific type of WBC). Lymphopenia (or lymphocytopenia) occurs when another category of the WBCs, known as lymphocyte counts, are low. Thrombocytopenia is caused by a reduction in platelets (cells used for clotting blood). And, pancytopenia
occurs when all the major types of cells normally found in blood are low. Clinically, cytopenias can be divided into two categories: 1) decreased or “lack of production” of the cells in the bone marrow (central production problems) and 2) removal, loss or destruction of the cells from the circulating blood (peripheral loss or destruction problems) (Table).

Why Do Cytopenias Occur?
All of the blood cell types (RBCs, WBCs and platelets) originate from primordial cells in the bone marrow. Stem cells give rise to progenitor cells, which in turn produce the specific blood cells to be released into the circulation (the peripheral blood) (Figure). Stem cells and progenitor cells primarily remain in the bone marrow (to continue to regenerate new blood cells). Thus, one mechanism that results in cytopenias is a lack of formation of the specific cell type in the bone marrow. This is aptly known as “lack of production.” Certain antibiotics, chemotherapies, poisons and toxins can impair cell growth and result in cytopenias (for example, it is common for a person treated for cancer to need RBC and platelet transfusions since chemotherapy impairs growth of those cells in the bone marrow). Also, viral infections (especially parvovirus B19, Epstein-Barr virus, cytomegalovirus, adenovirus, human immunodeficiency virus, etc.) can impair cell development in the bone marrow. In PI patients, the inability to clear an infection can add further risks for the development of cytopenias due to impaired growth of cells in the bone marrow. Further, in response to an infection or because of autoimmunity, the immune system can block cell development or kill developing cells in the bone marrow. Therefore, lack of production due to a variety of reasons can result in reduction of specific cell types, multiple cells types or all cell types (pancytopenia), which results in a reduction of the cells in the circulating blood (cytopenia).

The other major cause of cytopenias is the suitably named “peripheral removal, loss or destruction” of cells from the circulating blood. In this situation, the blood cell types have normally formed and matured in the bone marrow, but after they are released into the blood circulation, they are removed, lost or destroyed. The most typical reason for this is autoimmunity antibodies directed against the specific cell type, resulting most commonly in autoimmune thrombocytopenia (AIT), followed by autoimmune hemolytic anemia (AIHA). In AIHA, RBCs are removed and/or destroyed by autoantibodies directed to components found on the surfaces of the RBCs. This process applies to all of the cell types lost in the circulating blood.

The three main autoimmune cytopenias are 1) autoimmune thrombocytopenia (AIT), sometimes referred to as ITP for idiopathic thrombocytopenic purpura, idiopathic thrombocytopenia or immune thrombocytopenia, caused by the loss of platelets in blood; 2) AIHA; and 3) autoimmune neutropenia (AIN), caused by the loss of neutrophils in the blood. These are also the most common types of autoimmunity found

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<th>Table. Categories of Cytopenia</th>
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<tr>
<td><strong>Cell Type</strong></td>
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<tr>
<td>White Blood Cells (Leukocytes)</td>
</tr>
<tr>
<td>Lymphocytes</td>
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<tr>
<td>Neutrophils</td>
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<tr>
<td>Red Blood Cells</td>
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<td>Platelets</td>
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The general names used in the different categories are listed.
*Leukopenia is generally a problem with production of WBCs in the bone marrow, and autoantibody-mediated destruction in the circulating blood is not typically found, except for AIN.
**Lymphopenia is more typically a problem of production in the bone marrow. Autoantibody destruction of lymphocytes is not commonly found.
***AIT may be found in the literature named as ITP, originally for idiopathic thrombocytopenic purpura or idiopathic thrombocytopenia (idiopathic means the cause is not known) and later (and more commonly used more recently) as immune thrombocytopenia (meaning autoantibody-mediated disease is occurring).
in three PI conditions: 1) common variable immunodeficiency (CVID) (AIT in 14 percent, AIHA in 7 percent and AIN in less than 1 percent), 2) autoimmune lymphoproliferative syndrome (AIT in 51 percent, AIHA in 23 percent and AIN in less than 1 percent) and 3) combined immunodeficiency (AIHA is very common among infants with purine nucleoside phosphorylase deficiency).

In summary, cytopenia indicates reduced numbers of a specific type of blood cell either due to lack of production in the bone marrow (from multiple causes) or removal, loss or destruction of a specific cell type from the circulating blood. The latter typically occurs due to the production of autoimmunity antibodies, and they are typically labeled as autoimmune “specific-cell-type-penia, specific-cell-type cytopenia or specific-cell-type anemia” (for example, neutropenia, thrombocytopenia and RBC anemia, respectively). Reduction in all cell types is known as pancytopenia.

Signs and Symptoms of Cytopenias

Signs and symptoms depend on the type of cytopenia, as well as the underlying condition causing it. Typical symptoms associated with AIHA include fatigue, headache, dizziness, fainting and poor exercise tolerance. Frequently, patients look pale. They can develop a yellow discoloration known as jaundice due to the excessive breakdown of the RBCs and conversion of hemoglobin into bilirubins that accumulates in the skin and white of the eyes. In addition, the spleen may become enlarged as it traps the autoantibody-coated and damaged RBCs. The heart and lungs can become stressed due to excess work to compensate for the decreased capacity to carry oxygen in the blood caused by the reduction of RBCs.

In those with AIT, symptoms are typically easy bleeding and bruising, and difficulty with stopping bleeding. Bruising may occur in unusual areas or without known trauma to the area. Notable is the finding of petechiae in the skin, a rash of pinpoint-sized red dots caused by small hemorrhages into the skin. The capillaries (smallest blood vessels) essentially have a blow-out like the inner tube of a bicycle, and the blood leaks out. Nosebleeds may occur, and the gums may easily bleed. More serious internal bleeding may occur such as in the brain, which could result in a hemorrhagic stroke, one of the main concerns with very low platelet counts. For those with autoantibody-coated or peripheral destruction of platelets, an enlarged and tender spleen (splenomegaly) is common due to the accumulation of platelets removed from the circulating blood.

AIN results in increased susceptibility to bacterial and fungal infections due to reduced numbers of neutrophils. Fever is a frequent finding, but other signs of infection may also be present. Patients with AIN commonly have ulcers or sores in the mouth, esophagus or intestines, and the gums may become inflamed and red.

Cytopenias may be an indicator of an underlying problem such as cancer or leukemia, rather than a consequence of autoimmunity. In such cases, there is a lack of production in the bone marrow, likely due to the overgrowth of cancer cells. Several conditions commonly associated with cytopenias include multiple myeloma, Hodgkin’s lymphoma, non-Hodgkin’s lymphoma and other bone marrow diseases such as aplastic anemia and myelodysplasia. Additionally, vitamin B-12 deficiency, folic acid deficiency, chronic liver disease, viral infections, hepatitis, malaria and blood diseases that destroy blood cells such as paroxysmal nocturnal hemoglobinuria (PNH) result in cytopenias.
These underlying conditions more typically affect the growth and development of cells in the bone marrow caused by a lack of production rather than peripheral loss or destruction, except in the case of PNH.

**Diagnosing Cytopenias**

Diagnostic testing begins with a complete blood count that measures the WBC counts and RBC counts. A separate platelet count is also performed. Additional and follow-up testing depends on the patterns observed. For example, an isolated somewhat low platelet count, but not so low to be of concern for excessive bleeding, with normal WBC and RBC counts that seem to be occurring after a recent viral illness, may be followed for signs of bleeding, with a repeat in the blood and platelet counts after a few days. Platelets live for only a few days in the bloodstream, so they are continuously made in the bone marrow to replenish the blood. An interruption due to a viral illness or toxic event can lead to a temporary decrease that may last only a few days, and it is typically considered medically inconsequential. This condition occurs quite commonly, but typically resolves quickly, unlike the other pathologic processes discussed.

A slight decrease in neutrophils with normal RBC and platelet count may also be a consequence of recent viral illness and may be followed to determine whether it is temporary in a patient with no signs and symptoms of infections. Neutrophils have a short lifespan in the bloodstream and require constant replenishment from the bone marrow. A viral illness or toxic insult can result in reduction in the blood due to lack of production in the marrow, but usually resolves within a short period. As such, periodic checks of the WBCs can be performed following for evidence of recovery. Likewise, this is a commonly occurring process, unlike the other pathologic conditions being discussed.

Decreased RBCs with otherwise normal WBC and platelet counts may indicate a nutritional or vitamin deficiency. Specific characteristics of the RBCs can offer clues. Smaller, less dark RBCs can be seen in iron deficiency. Large, less dark RBCs can be seen with folic acid and vitamin B-12 deficiencies. Additional individual studies can be performed to confirm the deficiencies. RBCs live for about 120 days in the bloodstream. Hence, loss of RBCs or lack of production of RBCs can take longer to replenish than neutrophils or platelets. Therefore, if iron-deficiency anemia exists, supplemental iron would be given, and the counts would again be measured after a few weeks to assess the benefit.

Mild individual cytopenias may be evaluated and managed as discussed above. However, more severe or combinations of cytopenias require more intense and possibly more invasive investigations. Finding RBC anemia due to low counts but no changes in the size or color of the RBCs (so-called normochromic, normocytic anemia) may indicate lack of production or loss of RBCs (for example, from hemorrhage into the gastrointestinal tract). If the platelet count is also low, and there is also possibly a low neutrophil count, performing a bone marrow aspiration and bone marrow biopsy is considered imperative to determine if leukemia or lymphoma is present, or if a condition such as myelodysplasia (a marrow full of cells, but not the correct maturation to release into the blood) or a marrow empty of cells as found in aplastic anemia is present.8

Examining the bone marrow aspirate and biopsying each for the developing cell types to determine which are diminished, whether there is early development and then blockage of further development, and whether there is presence of excess eosinophils and erythrophagocytosis (also known as hemophagocytosis, which is macrophages engulfing developing RBCs) can also be helpful for diagnosis by narrowing the disease possibilities.

Other assays are available to help determine if the anemia is a consequence of marrow production issues, peripheral loss or destruction issues. A Coombs test determines if there are antibodies directed to the RBCs (testing for AIHA). Additionally, tests are available to identify the specific targets of the antibodies on the RBC surfaces. In particular, identifying these targets can be useful when providing transfusion therapy. Hemoglobin analyses can determine if a hemoglobinopathy such as sickle cell disease or thalassemia is present. And, with specific testing, other hereditary conditions such as hereditary spherocytosis can be identified.

There are also specific assays for antibodies directed toward platelets. Antibodies can be made to specific adhesion molecules on platelets that result in AIT. Identifying these can be helpful for providing specific platelets for transfusion therapy. Likewise, there are assays to determine which anti-neutrophil antibodies may be present, resulting in AIN.

**Treating Cytopenias**

Treatment can be divided into acute treatment to deal with the most immediate issues and chronic therapy for treating the underlying issues and attempting to resolve the cytopenia.
Acute therapies may involve transfusing the missing cell type. When the issue is lack of production in the bone marrow, transfusion will provide temporary improvement for the patient. When the cytopenia is a result of peripheral loss or destruction due to autoantibodies, unfortunately transfusion can actually worsen the cytopenia. Clearly, a careful diagnosis is crucial.

Transfusion of neutrophils can be performed by those familiar with this process, but it is typically unavailable. Leukocytes, which contain neutrophils and lymphocytes, can be obtained from a single donor by a process known as apheresis. Essentially, a person’s blood is directed through an IV catheter into a machine that separates the leukocytes (WBCs) from the RBCs. Through a separate IV catheter, the RBCs are infused back into the patient, while the WBCs are collected in the machine. Alternatively, leukocytes can be obtained from individual blood donations at donation centers. Typically, the WBCs from several individual donations are pooled together to obtain sufficient neutrophils for infusion. The leukocytes will be irradiated with gamma radiation to prevent the lymphocytes from growing and dividing after transfusion, since this could result in a disease process known as graft-versus-host disease (GvHD), leading to severe complications and possible death. Since the neutrophil lifespan is relatively short, leukocyte transfusions need to be performed essentially every day or every other day to be successful in fighting and clearing infections.

For AIHA, identifying the autoantibody targets on the RBCs can be helpful. RBCs from a donor who does not have the target on their RBCs can allow for successful transfusion. This is also true for platelet transfusions in AIT. In some cases, too many targets have antibodies directed against them or a donor lacking the targets is not available. In these circumstances, successful RBC and platelet transfusions can be accomplished by slowly infusing the cells (sometimes called a "slow drip") simultaneously with a slow infusion of intravenous immune globulin (IVIG). The IgGs in the IVIG infusions can sometimes mitigate the effects of the patient’s autoantibodies, allowing for successful transfusion.

Otherwise, corticosteroids have been the first-line therapy for cytopenias essentially since they became available. While corticosteroids can provide benefit in most patients for most types of cytopenias, recurrence is likely when attempts are made to taper the dosing of the corticosteroids. Thus, therapy can be successfully begun but not typically completed, and there are major adverse long-term side effects with prolonged corticosteroid usage.

For the eventual discontinuation of corticosteroids, the clinical routine is to use a “steroid-sparing” agent. Traditionally, this agent was primarily azathioprine. Alternatively, cyclophosphamide was used. Ironically, both of these toxic medications can cause bone marrow suppression-associated cytopenias, which can prevent the situation from improving. Mycophenolate usage has mainly replaced these older medications, but it can still result in bone marrow suppression. Therefore, the irony is these medications may help improve the typical autoimmune cytopenia (circulating blood loss or destruction), but they may result in impaired development of the cells in the bone marrow and cause lack-of-production cytopenias. Physicians have to carefully monitor and adjust medication dosages to achieve the desired benefit. Another downside for PI patients is that this approach to treatment is immunosuppressive. As a result, a patient with an already compromised immune system may have even more impairment of immunity with risk for infections. Other immunosuppressive agents (for example cyclosporine A, tacrolimus, sirolimus, etc.) that have been successful in preventing graft rejection in the transplantation setting and have been effective in treating a variety of autoimmune disorders have been successfully used to treat cytopenias. Mostly, these help with the immune process impairing the development of cells in the bone marrow. With multiple options available, treatment can be individualized to the patient, which may provide better outcomes.

A typical treatment approach includes a trial of corticosteroids to try to rapidly halt the process (either immune-mediated suppression of cells developing in the bone marrow or autoimmune attack of cells in the circulating blood), with transfusions as needed. If a cytopenia is returning with a decrease in corticosteroid usage, then a secondary medication such as mycophenolate (or other medication individualized to the needs of the patient) is added. This approach will typically be used for a patient who is not too severely affected by the cytopenia.

Alternatively, many doctors may immediately proceed with high-dose IVIG (2 g/kg). This is an overall safe process, and it does not have inherent marrow suppression as a side effect. Further, high-dose IVIG helps to support the immunity rather than suppress it as other medications can, and thereby it can reduce the risk for infections that occur with immunosuppressive therapies. In addition, due to the month-long half-life of IgG, the dosage may provide prolonged benefit in some. Therefore, if the cytopenia is immune-mediated suppression of growth of cells in the bone
RBCs, which then “clog up” all the sites to which platelets with AIT. It is thought that the anti-D binds to the patient’s immune globulin can improve the platelet count in those or D positive blood type, infusing anti-D (or anti-Rho) “D” antigen. It has been found that if someone has the Rho making it more complicated, this is also referred to as the positive refers to being Rho positive. Unfortunately, When a blood type is mentioned, for example A positive, infectious complications.

remain on penicillin or a similar medication to help prevent immunity causing cytopenias. Typically, for maximal benefit, a combination of IVIG and rituximab is used.

For patients with AIHA or AIT not responding to therapeutic interventions just described, splenectomy has been performed in the past. The spleen acts as a filter collecting most of the RBCs and platelets coated with autoantibodies, taking them out of circulation. With the spleen removed, even though autoantibodies still coat the RBCs or platelets, they remain in circulation. Since the spleen is critical for immune protection, patients are immunized with the pneumococcal and meningococcal vaccines prior to splenectomy. Further, they may need to remain on penicillin or a similar medication to help prevent infectious complications.

Rho is a protein component found on the surfaces of RBCs. When a blood type is mentioned, for example A positive, the positive refers to being Rho positive. Unfortunately, making it more complicated, this is also referred to as the “D” antigen. It has been found that if someone has the Rho or D positive blood type, infusing anti-D (or anti-Rho) immune globulin can improve the platelet count in those with AIT. It is thought that the anti-D binds to the patient’s RBCs, which then “clog up” all the sites to which platelets coated with autoantibodies would otherwise be removed, thus preserving the platelets in the circulation. This occurs since the normal RBC count greatly exceeds the normal platelet count in the blood, so there are many more RBCs to clog the removal sites before a platelet finds its way to one of them. As a cautionary note, the RBC count may decline by 5 percent to 10 percent with this therapy, and therefore, must also be carefully monitored. In some cases, RBC transfusion is given before anti-D infusion to prevent RBC anemia (thus another irony; RBC anemia is caused via antibody infusion to preserve circulating platelets). Indeed, prior to splenectomy for AIT that does not respond to other therapies, anti-D is infused. If there is improvement in the platelet count, splenectomy may be successful. However, if there is no improvement, then splenectomy will likely not resolve the thrombocytopenia and should be deferred.

When someone develops a cytopenia, it means that person is lacking specific cells. Thrombopoietin is a protein growth factor used to stimulate the production of platelets in the bone marrow when thrombocytopenia is caused by a lack of production, rather than due to circulating loss or destruction. When pre-leukemia, leukemia or lymphoma is present in the marrow interfering with platelet production, treating the underlying condition can result in improved platelet counts. For example, treatment with venetoclax, approved by the U.S. Food and Drug Administration to treat relapsed/ refractory chronic lymphocytic leukemia (CLL), resulted in improvement of platelet counts in CLL patients. Likewise, erythropoietin (protein growth factor for RBC development) can be used for RBC anemia due to lack of production.

When neutropenia occurs, the first assessment is to determine whether it is a production issue or antibody-mediated peripheral loss or destruction issue. In any case, corticosteroids are frequently used as first-line therapy. However, the downsides of corticosteroids are the adverse side effects and recurrence of disease when tapering this therapy. If production appears to be the problem, recombinant
human granulocyte colony stimulating factor can be used to increase the counts in the blood.\(^8\) Dosing amount and frequency are determined by how the neutrophil count responds and if infections occur, with higher and more frequent dosing for poorer improvement in the counts and infections that persist.\(^9\) Unfortunately, immunosuppressive therapy, IVIG, splenectomy and rituximab have not been shown to be as beneficial for most patients with AIN.\(^8\)

The more we learn, the clearer it becomes that PI patients have a predisposition to developing autoimmunity.

Patients who are refractory to treatment may require hematopoietic cell transplantation, especially if there is an underlying condition such as leukemia causing the cytopenia.

### Long-Term Prognosis

The more we learn, the clearer it becomes that PI patients have a predisposition to developing autoimmunity. For those with CVID, autoimmune cytopenias are a common problem. Autoimmunity may be exhibited as activity against RBCs, WBCs and/or platelets in the bone marrow, which may be mediated by T lymphocytes. Alternatively, autoimmune antibodies may develop that result in destruction, loss or removal of RBCs, WBCs and/or platelets from circulating blood. In some cases, both lack of production in the bone marrow and loss or destruction in circulating blood can occur, further complicating diagnosis and treatment. In some cases, though, despite the apparent commonality of a specific cytopenia with a specific immunodeficiency, it can be more difficult to determine or understand the root of the cytopenia, and consequently, more time and in-depth evaluations may be required. Moreover, this potentially translates into a more entailed treatment regimen.

Since patients with immune problems can be at higher risk for malignancies, these must be considered early in the diagnostic process and thoroughly evaluated to identify and treat them as soon as possible.

The eventual success of treatment regimens also depends on other comorbidities and their treatment. Comorbidity complexities result in more diverse treatment needs. In 2018, 990 patients with CVID in the USIDNET Registry were listed with diagnoses consistent with autoimmune cytopenias. Additionally, those with autoimmune cytopenias were likely to have other conditions, including interstitial lung disease, enteropathy, hepatic disease, lymphoproliferation, lymphoma, granulomatous disease and organ-specific autoimmunity, all resulting in an increased risk of morbidity and mortality.\(^11\)

For most, the therapies discussed are effective, but for PI patients, successful therapy tends to be IVIG and rituximab. When treatment is successful, most patients have no major restrictions on their daily activities. However, patients with chronically low platelet counts may have refrained from activities with a higher risk of injury such as contact sports.\(^4\)

Ultimately, the evaluator of such patients must consider and recognize that multiple processes may be occurring, which may not appear at first to be related due to the different cells, tissues and organs involved. Indeed, an underlying immunodeficiency with potential for the development of autoimmunity may be the issue, requiring aggressive pursuit of diagnosis and treatment that will generally result in overall better outcomes.\(^9\)

### References


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Download the *IG Living* eBook today—now available for iPad, Nook and Kindle!

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Let’s Talk!

Profile: Luba Sobolevsky

By Trudie Mitschang

Trudie: Tell us about the industry need that IgNS was created to fill.

Luba: IgNS started because of a critical need in our field for education, care standards and practice resources. Almost all clinical areas such as immunology, oncology and neurology have accepted standards of practice that practitioners follow. But IG is prescribed for almost 100 different clinical indications, and it is very difficult for any single association focused on a disease state to come up with universal standards and guidelines. IG therapy practice also includes a multi-disciplinary care team and practice model, so it’s no wonder it took so long for an organization to take the lead to develop resources for education and application.

Trudie: How did IgNS start?

Luba: When IgNS was started in 2012, we conducted a landmark clinician survey that showed more than 90 percent of healthcare professionals never received any systematic education in IG therapy, they did not have any resources or standards of practice to reference, and there were no guidelines for basic policies and protocols. Additionally, there was no way to assess a clinician’s level of expertise, knowledge or skill. Because IG is one of the most complex therapies available, it is unacceptable for IG clinical practice to not be guided by standards. It is not possible to maintain good practice and ensure patient safety without standardized practice, education and other critical resources. And, that was the main impetus for starting IgNS.

Trudie: How has your work evolved?

Luba: In the years since our inception, we’ve established a very robust educational platform for clinicians (nurses, pharmacists and physicians) through our annual conference and our online content and resources. We have also developed the first-ever clinically and legally defensible IgNS Standards of Practice for IG therapy. This means every provider or organization, whether it’s a specialty pharmacy, independent infusion center, private practice or hospital, has access to comprehensive guidelines for treating patients with different diagnoses in different settings. In addition, IgNS has developed a very unique certification program that focuses strictly on IG therapy, called the Ig Certified Nurse and Ig Certified Pharmacist, so there is now a tangible way to assess clinical skills and for patients to gauge their nurses’ expertise. The four-letter credential after their name — IgCN for nurses and IgCP for pharmacists — is how patients can easily recognize a clinician as an IG therapy expert.

Trudie: How does this framework help patients?

Luba: In a very practical sense, it means no matter where patients receive care or who comprises their clinical team, their care should remain consistent and of the highest standard. It gives patients peace of mind and empowers them to set high expectations, and it empowers clinicians to hold themselves accountable for the level of care they provide.

Trudie: How are you measuring success?

Luba: We know from tracking exams that the resources and education we have put in place are making a big difference in clinician skills and knowledge. For patients, we started the IgNS Patient 360 Conference that runs in conjunction with the annual IgNS National Conference that convenes more than 1,000 clinicians, industry members, patients and advocacy groups who work together to improve practice and patient care. We wanted to learn more about this trailblazing organization from its Founding Executive Director Luba Sobolevsky.

SINCE THE Immunoglobulin National Society’s (IgNS) inception nearly a decade ago, the organization has focused on improving the care of patients receiving immune globulin (IG) therapy. IgNS has accomplished many things, including creating the first-ever Ig Standards of Practice to guide and standardize IG clinical practice; establishing an IG certification for nurses and pharmacists that ensures expertise and protects patients; providing thousands of hours of education to keep up with the changes in this complex field; and hosting the IgNS National Conference for clinicians and the IgNS Patient 360 Conference that convenes more than 1,000 clinicians, industry members, patients and advocacy groups who work together to improve practice and patient care. We wanted to learn more about this trailblazing organization from its Founding Executive Director Luba Sobolevsky.
Let's Talk!

Luba: This year’s virtual event took place in October and is still available on-demand until December 31.

Trudie: How else does IgNS collect data?

Luba: An important part of the IgNS Patient 360 Conference is an annual survey that teaches us about patient experiences with IG therapy, as well as their major needs and obstacles, and that allows us to gauge how things are changing and evolving. The survey includes questions about diagnosis and treatments, where patients receive IG and the route of administration, side effects, insurance issues, as well as patient perceptions about their clinical team’s knowledge, skill set, communication and collaboration. Specific questions like this help us measure where the key gaps are so we can focus our educational and resource efforts in those areas. We have been conducting this survey now for three years, and we intend for it to continue. As a clinician, I know if we are not listening to patients, we cannot be effective in our practice.

Trudie: Tell us about your professional background.

Luba: I am a pharmacist who has worked in different areas of practice, both in clinical settings and in the pharmaceutical industry. I understand patient needs and the obstacles patients may experience, as well as the difficulties and obstacles clinicians might experience in terms of education and resources. My background has helped me gain important insight into the research and development of IG therapy, as well as the complexity of manufacturing, bringing products to market and delivering them to patients. In my role at IgNS, my background helps me to see the big picture, establish important collaborations across our industry and be a more effective advocate of the IgNS initiatives.

Trudie: What do you find most rewarding about what you do?

Luba: Most rewarding are creating IG therapy resources that are vitally needed for clinicians and patients, developing something that did not previously exist and witnessing practice improvements. From having 100 participants at our first conference to more than 1,000 this year, I know the work we do directly improves the standard of care and quality of life for patients. As a clinician, you cannot ask for anything more rewarding. That is what drives me every day.

Trudie: What are your goals as you look ahead?

Luba: It’s been a difficult year, to say the least. We have a high-risk patient population, many of whom are immunocompromised. We are facing a decline in plasma donations due to COVID-19. IgNS was one of the first organizations to launch a plasma collection campaign called “It’s My Turn” that appeals to all members of our society to donate plasma and help save lives. Many people did not know, for example, that plasma donation centers are considered essential, and they have remained open throughout the pandemic. The decline in donations has a real possibility of impacting IG supply within 12 months to 18 months, and we must do all that is in our power to prevent a product shortage. We are fortunate to have great industry partners — companies and advocacy groups that have endorsed the campaign, and thousands of individuals who have stepped up to donate plasma and partner with us to educate the public about plasma donation.

Trudie: What distinguishes IgNS from other associations?

Luba: IgNS is very unique. We believe education must reflect real practice. Our resources are comprehensive and robust and have a practice-based approach. We are a multidisciplinary association that includes all types of healthcare professionals. We welcome and serve as a conduit to clinicians, business professionals, advocacy and trade groups, manufacturers and, of course, patients. It is very special that patients are part of our organization and conferences. We have the opportunity to connect patients with advocacy groups and experts who provide education and help resolve their major concerns. Forming a larger IG patient community regardless of diagnosis is very important; it leads to better support, better understanding about patient experience with IG therapy and more effective advocacy efforts.

Trudie: How do you engage with clinicians and patients throughout the year?

Luba: We have ongoing live educational events, a robust online educational platform for members, podcasts and publications, and we are very active on social media. I encourage everyone to visit our website and learn more about IgNS.

Editor’s note: To learn more about IgNS and to access their annual conference on-demand, visit www.Ig-NS.org.

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.
Patient Perspective

Purpose in Pain
By Stacey Philpot

AT 19 YEARS old, my witty and protective older brother, Matt, with his kind wise brown eyes lay in a rented hospital bed in the bedroom next to mine. He wore an adult diaper and his favorite St. Louis Cardinals shirt while my parents washed his hair with dry shampoo and then wiped down each of his shriveled, lean limbs. Cancer had engulfed his body like wildfire across the parched ground in a matter of months.

I stood in the doorway that morning wondering what in the world was happening as a monitor told us he’d stopped breathing, his lips turned blue and he left that pain-filled shell behind. I hadn’t known it was possible to love someone so deeply or to hurt so profoundly; it felt as though a limb had been torn away.

My mom asked me to call the numbers on a handwritten list to inform each household that my best friend on this Earth had left the world that day. One by one, I dialed the numbers, broke the news and comforted each person as they tried to absorb the shock. That was the day I learned finding purpose by helping others can guide us through our pain. Each time I comforted someone, I found the gaping hole in my chest was a little less achy.

This year, we’ve seen a great deal of hurt in our nation. We’ve likely experienced anxiety, pain and loss in our lives on top of our illnesses. How do we cope with it all? Perhaps, we can:

1) Take care of others. When nothing makes sense, the pain is unbearable, the waves of grief come crashing in or the uncertainty is more than we can bear, I’ve found taking care of others provides a positive, productive outlet for my pain and restlessness. When others are comforted, have their needs met and know someone cares, I find my pain dissipates. Having a project gives me a sense of purpose and control in moments when it can feel as though everything is spiraling out of control. Using our talents and skills on behalf of others gives us something productive to focus on and helps our communities. This might mean volunteering at a shelter, babysitting for a friend, knitting blankets for the cancer center, tutoring online, teaching a class or supporting a friend who recently received a new diagnosis. Whatever it looks like, staying busy allows us to find purpose in pain.

2) Take care of ourselves. Whether it’s going for a walk, traveling to a new location, enjoying time with a pet, practicing yoga, being intentional about what we eat, checking in with friends, reading a good book, talking with a therapist, enjoying a new hobby or taking the time to grieve, taking care of ourselves is critical all the time, but especially when we are in pain.

3) Take care of our world. Gardening, recycling and reducing waste can provide outlets for proactive energy that are enjoyable and good for the environment. Whether we make small changes to our lifestyle or lead a group in our area, taking care of our world can be one way we ease pain and bring joy.

All of us go through seasons of pain and loss in our lives. Collectively, we’ve experienced a great deal of loss this year. Each of us is searching for ways to cope. Hopefully, we’re finding supportive friends to hold us up when it all feels like too much and to encourage us when we feel like we can’t continue. Perhaps we can use these moments of pain to refine our caretaking skills and find purpose that eases the pain.

When others are comforted, have their needs met and know someone cares, I find my pain dissipates.

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.
Considering Relocating? A Few Tips!

By Michelle Searle

I GREW UP in a town just outside of Fort Lauderdale, Fla., and although I love it there, I knew I wanted to leave when it was time to go to college. I was ready and excited to live independently, meet new people, have new experiences and make a home for myself in a new city. So, I attended college across the state, a two-hour drive from my hometown. After college, I followed my dream and moved across the world, a nine-hour plane and two-hour bus ride away from my hometown to a city in Italy. Since I have a primary immune deficiency (PI), it was scary to think about moving somewhere I wouldn’t know anybody, and where I wouldn’t have my family or friends in case I needed help or my doctors who know my medical history. Now, having moved twice, I have some advice for others with PI who are considering relocating but are unsure where to start. Following are some of the things I considered before moving, as well as important lessons learned about moving out of your comfort zone:

Consider your medical needs. I’m terrified about anything having to do with medicine, so when I was growing up, my mom administered my infusions. But as college neared, I knew it was time for me to take care of my own medical needs, which meant ordering my supplies and setting up and administering my infusions. I made this transition months before leaving for college while my mom would be there in case I needed help and to build confidence. I also took over the role of scheduling my appointments, making sure I always had the correct paperwork and going to see doctors by myself. It’s important to consider what things you’ll have to learn to do for yourself well before you’re on your own.

Doctors, specialists and medications, oh my! Depending on where you are relocating, it might be possible to keep the same doctors and specialists. If that’s not the case, it’s important to find and connect with new doctors ahead of time. When I relocated for college, I kept my immunologist and would go to see her when I returned home for visits. However, when I moved to Italy, I had to find a new immunologist months prior to moving, and I had to set up an appointment for the week I arrived. While the Immune Deficiency Foundation is a great resource to find immunologists in the United States, the Jeffrey Modell Foundation can help you find immunologists worldwide.

Medications must also be considered. For instance, can your medicines be mailed to the new location, or can a large amount be prescribed for you to take to the new location? Will you need to locate an infusion center or a new specialist?

Money, money, money. Living with a chronic illness is expensive. Insurance, medications, doctor visits, tests and therapy are just some examples of possible expenses. What’s more, depending on your chronic illness, you may not be employable. Therefore, when relocating, you must evaluate if you’ll be able to work, what the job market is like, where income will come from and whether medical expenses will increase or decrease.

Consider your mental health. Where we live can have a big impact on our mental health. Therefore, it’s essential to evaluate what kind of environment will bring you joy. For instance, do you desire weather that inspires you to go to the beach and watch the waves, or would you rather watch the snow fall while you’re curled up in front of a fireplace? Or maybe what matters to you is living somewhere that will enable you to engage in a favorite hobby or that has great support groups and therapists. It’s also important to consider whether the culture makes you feel included or excluded. Our bodies and minds connect, so make certain both needs are met.

Wherever you decide to move or even if you’re still deciding if you want to move, ensure your physical, mental and medical needs are covered at the new location. Remember: Your chronic illness is only one part of you. If you want to move somewhere new and experience the unique challenges that come with moving, you absolutely can! It will just take some extra planning.

MICHELLE SEARLE is a teacher from South Florida who was diagnosed with common variable immunodeficiency at 11 years old. She is currently living in Italy where you will most likely find her eating pizza or trying to make friends with the local cats.
Family Balance When Some Siblings Are Chronically Ill and Others Are Unaffected

By Jessica Leigh Johnson

IN MY family, we have three boys, all living with a primary immunodeficiency (PI). As a caregiver, it can be a bit overwhelming to have not just one child with a chronic illness, but three. Common tasks like ordering monthly infusion supplies or administering at-home infusions just take longer — three times longer than for only one child. But even though the responsibilities and worry of raising three chronically ill children are multiplied, I’m often thankful my boys have each other as they go through life. They understand each other. They understand what it’s like to be sick or the disappointment of missing out on something because of a medical issue. When one son goes to the lab for a blood test, the others are there, too, waiting for their turn to be poked. I realize it would be easier on me if only one (or none) of my sons had PI, but it might not be easier on them.

In a family where only one child suffers from chronic illness, how does that child feel? Does he or she feel singled out or that the situation is unfair? Does that child feel envious of his or her unaffected siblings? And where does that child turn for support? As parents, how can we find the right balance within the family when so much of our attention seems focused on our chronically ill kids?

Encouragement for Parents

First of all, parents should not feel guilty for paying a little extra attention to the needs of their chronically ill children. All kids have needs, and those who suffer from chronic illness have a few extra ones. They may require more of their parents’ attention, especially when it comes to their health — and that’s OK. Every child deserves to feel loved and to have his or her needs met.

But sometimes caring for a chronically ill child can become all-consuming. Issues like limited treatment options, the cost of medical care and concern about the future can give parents tunnel vision and cause them to lose touch with the needs of their other family members. They can become so obsessive over caring for their child with a chronic illness or disability that it negatively impacts their lives — particularly their relationships with their spouses or their other children.

Parents should never allow dealing with their child’s illness to overwhelm them. Parents aren’t able to solve all family problems associated with their child’s illness by themselves, and they need to be careful not to become isolated from others. Help and support can be found from physicians, counselors, faith-based groups, friends and other parents of children with the same or similar chronic illnesses. If family problems arise from taking care of a child with a chronic illness, parents need to ask for help.

Focus Special Attention on the Unaffected Siblings

The unaffected children also need to know their feelings and needs are just as valid and important as the needs of their chronically ill siblings. Every member of a family is valuable and deserves special attention from time to time. It is important that parents are intentional about building and maintaining close relationships with their unaffected children. Although, as caregivers, we may be physically tired or mentally exhausted after a day at the clinic or after a lengthy phone call with the insurance company, we must be careful not to use our child’s chronic illness as an excuse to put off siblings’ needs.

For example, if an unaffected sister comes to one of her parents with friend
troubles or homework she doesn’t understand, that parent should not reply with a statement, “I’m too tired to deal with this right now because I’ve been caring for your brother’s medical issues.” Statements like this are sure to build resentment between siblings. Instead, while parents do need to be honest when they’re feeling overextended, they also need to be intentional. Saying, “I’m not able to talk right now, but how about I take a short rest and we look at your homework in an hour?” lets the unaffected child know that her parent has not forgotten about her needs, and intends to help her. But parents must be true to their word and keep that homework appointment an hour later.¹

Scheduling special outings for healthy siblings is another way to help them to feel the chronically ill child doesn’t get all of his or her parents’ time. In our family, we only have one daughter, and she does not suffer from PI like her brothers. Being the only girl has given her opportunities for special outings such as girls’ weekends and sleepovers, which her brothers didn’t get to participate in. These times help her to feel special (and give her a little time away from the boys), since over the years, she has had to make sacrifices because of her brothers’ condition.

**Emotional Support for the Child with Chronic Illness**

No one should have to go through struggles alone, and chronic illness can often be a battle. It’s crucial for a child’s emotional health and well-being that he or she has someone to talk to about the unique issues that arise when living with a chronic illness. In families where more than one sibling is affected, it’s easier for children to find an empathetic listening ear. It’s natural for them to talk to one another about their feelings or their symptoms. But when only one child suffers from the illness, he or she turns to someone else in the family such as a parent or older, unaffected sibling, and that person won’t always be able to relate.

To foster an environment where kids are comfortable talking about their chronic illness, parents should encourage them to speak freely and ask questions. Let them share their frustrations and emotions regarding the issues they face, even if the parents don’t personally experience the same things. Parents can ask questions, too, and that will help the child feel like family members truly care about what’s going on in that child’s life. Also, connecting children to others who are affected by the same condition — through specific illness-based social networks and patient advocacy groups — can be a great source of encouragement and an invaluable resource, provided all online activity is closely monitored by parents.

**Encourage Family Unity**

Obviously, building relationships within the family is important for siblings to stay connected going forward into adulthood. The more activities families do together, the less a child will feel that a wall between siblings exists, discouraging division in relationships because of his or her own chronic illness or that of a sibling. Fostering family cohesiveness creates less of a me versus them feeling.

In my own family, now that my daughter has left home to attend her first year in college this year, I really hope she doesn’t look back on her childhood and feel like she got slighted or missed out on things because of her brothers. I hope that as a family, we focused enough on doing things together with all of our kids such as attending sporting events together and taking family vacations. I also hope we’ve given our daughter the freedom to do some things on her own — such as having her own YMCA membership, a job and the opportunity to participate in sports — without feeling tethered to her brothers or held back by their condition. I’m confident we’ve given our children plenty of opportunities to spend time together and with us as parents that had nothing to do with whether they did or did not have a chronic illness. These shared experiences are something they will all have in common, something they did together, regardless of their health circumstances. And those memories are the ones we as parents hope they carry with them for a long time to come.

**References**


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**GETTING DRESSED.** A trip to the market. Making dinner. For most, these are simple tasks that don’t require much thought or energy. But for individuals living with chronic illnesses such as primary immunodeficiency diseases, autoimmune disorders or chronic pain, completing these tasks every day can often be a challenge. That’s where assistive devices or technology come into play. Thanks to these tools and technologies — which run the gamut from something as simple as a pill organizer or grocery delivery to a sophisticated voice-activated computer system or robotic caregiver — individuals can remain self-sufficient.

**A Growing Group**

Chronic diseases affect at least 133 million Americans or 40 percent of the population, according to the National Health Council. By 2020, that number is expected to balloon to about 157 million, with 81 million people suffering from multiple conditions. Of these Americans, about 40 million are limited in their usual activities, which is why devices that help improve quality of life and enable independence are becoming increasingly popular.

**Learning to Adapt**

Individuals with chronic illnesses are regularly plagued by fatigue and recurrent infections, making it difficult to complete daily tasks. For some, weekly or monthly infusions take time and are often physically and mentally draining. Sometimes these treatments lead to an array of side effects, including headaches, fatigue, insomnia, muscle pain and nausea. In addition, because these conditions leave individuals vulnerable to viruses or other infections, limited public exposure is often a necessity — particularly during this current pandemic.

Yet, if daily activities are hindered by any of these issues, that doesn’t necessarily mean independence is unattainable. The solution may lie in adaptive equipment — technologies that can improve both a person’s mobility and level of function.

Simple items such as dressing sticks, button hooks, sock aids, long-handled shoehorns and elastic shoelaces are easy to incorporate into daily life. If standing and lathering up is an issue, a tub/shower chair or bench may do the trick. Installing some grab bars or a handheld shower nozzle could be the solution to staying fresh and squeaky clean. For food preparation and dining, there are now a variety of food and grocery delivery services available.

**Keep It Simple**

Medication management technology is just another example of taking the high-tech or low-tech road. Regardless of the choice, the goal is the same: to ensure patients are safe, happy and healthy. For example, some simple gadgets such as a days-of-the-week pillbox or an automated dispenser that beeps when it’s time to take meds can make life easier.

Even in this awesome world of innovation, the simple option may be the best choice. Not only does simplicity usually translate to less expensive, these devices are often easier to use, less complex and require less maintenance.

So shelf the idea of a home care robot for the time being and just order a pill box, schedule a weekly grocery delivery and know you’ll be able to live independently and happily.

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**HEATHER BREMNER CLAVERIE** is a contributing writer for IG Living magazine.
**Scrub Up Safely**

If standing in the shower is difficult and stepping over the bathtub wall is dangerous, it may be time for the Safe Step Walk-In Tub. Walk straight into this tub via the easy-to-open door, grab a loofah, take a seat and start scrubbing. The dual hydrotherapy system helps soothe aches and pains, while the grab bars, anti-slip flooring and anti-scald technology ensure a safe bathing experience.

*Pricing varies; www.safestebath.com/walk-in-tub*

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**Easy Does It**

Pop those pills in this simple and affordable box, and it’s easy to remember what to take and when. The LazyMe Weekly Pill Box Meds Case holds a week’s work of pills for morning, noon and night. And, there’s even a slot for “backup” pills just in case. $5.98; [www.amazon.com/LazyMe-Weekly-Large-Compartments-Times/dp/B01H0HQ51Q](www.amazon.com/LazyMe-Weekly-Large-Compartments-Times/dp/B01H0HQ51Q)

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**Apples at Your Door**

Upload the app, fill the virtual basket and wait for groceries to appear on your doorstep. It’s that simple with Instacart. Order from participating retailers, which usually include most local markets, for same-day delivery or pickup. And once the first order is scheduled, the app conveniently saves your list, making future orders as simple as a swipe. First delivery is free.

*$99/year or $9.99/month; instacart.com*

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**You Need a Hero**

Set up your daily regimen with the Hero automatic pill dispenser that will pop out meds at the right time and on the right day. This automated device can dispense up to 10 different kinds of pills a day and store up to a 90-day supply. In addition, an app will send alerts regarding dose amounts, when and if medicines were taken and overall medication adherence.

*$29.99/month; herohealth.com/our-product*

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**Sock It to You**

Huffing and puffing when pulling on those socks? The Norco Easy-Pull Sock Aids can help. Simply stretch the sock over the flexible plastic trough, slip your foot into the terrycloth-lined device, grab the two large loop handles and pull. This device is available in a variety of models to suit various needs and preferences.

*$20.35; www.rehabmart.com/product/easypull-sock-aid-78.html*
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- Overcoming Multiple Sclerosis: overcomingms.org/community

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