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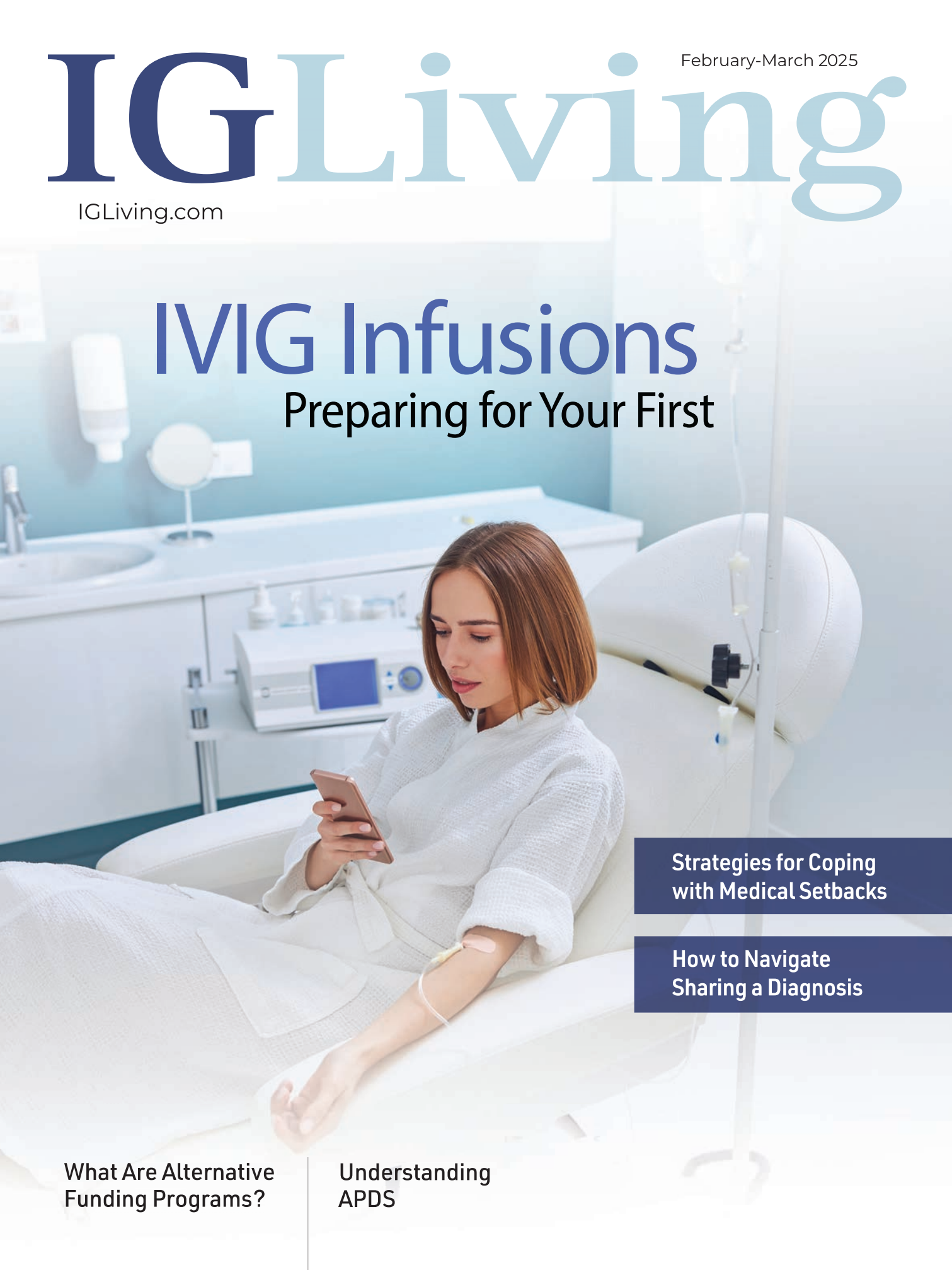

IVIG Infusions

Preparing for Your First

Strategies for Coping with Medical Setbacks

How to Navigate Sharing a Diagnosis

What Are Alternative Funding Programs?

Understanding APDS

FOR PATIENTS WITH PRIMARY HUMORAL IMMUNODEFICIENCY (PI)

IT'S WHAT'S INSIDE THAT COUNTS

ASCENIV™

IMMUNE GLOBULIN INTRAVENOUS (HUMAN) – slra 10% LIQUID

DESIGNED TO DELIVER

Talk to your doctor about whether ASCENIV™ is right for you asceniv.com

Important Safety Information for ASCENIV™

WARNING: RISK OF BLOOD CLOTS (THROMBOSIS), POOR KIDNEY FUNCTION, AND INABILITY TO FILTER WASTE FROM KIDNEYS. BLOOD CLOTS MAY OCCUR WITH INTRAVENOUS IMMUNE GLOBULIN PRODUCTS, INCLUDING ASCENIV.

Before taking ASCENIV, talk to your doctor if you:

- Are of advanced age
- Are unusually sedentary (long periods of sitting down or inactive)
- Are taking estrogen-containing medicines (birth control pills, hormone replacement therapy)
- Have a permanent intravenous (IV) catheter
- Have hyperviscosity of the blood (diseases such as multiple myeloma or other causes of elevated proteins in the blood)
- Have cardiovascular (heart) problems or previous history of stroke

Thrombosis may occur even if you do not have any risk factors.

Serious kidney problems and death can also happen in certain patients who receive such products.

If you are at high risk of thrombosis or kidney problems, your doctor should adjust the dose of ASCENIV and will monitor you for signs and symptoms of thrombosis and viscosity, as well as kidney function.

What is ASCENIV (immune globulin intravenous, human)?
ASCENIV (immune globulin intravenous, human) is a prescription medicine to help adults and adolescents (12 to 17 years old) with primary immunodeficiency fight and prevent infections.

ASCENIV is for intravenous administration only. ASCENIV is made from healthy human blood/plasma.

Who should not use ASCENIV?
ASCENIV should not be used if you had a severe allergic reaction to human immune globulin or if you have been told by a doctor that you are immunoglobulin A (IgA)-deficient and have developed antibodies to IgA and hypersensitivity after exposure to a previous plasma product.

What are possible warnings and precautions with taking ASCENIV?
Hypersensitivity. Severe allergic reactions may occur with immune globulin products, including ASCENIV. If you have a severe allergic reaction, stop the infusion immediately and get medical attention. ASCENIV contains IgA. If you have known antibodies to IgA, you may have a greater risk of developing potentially severe allergic reactions.

If you take ASCENIV or a similar immune globulin product, you could experience a serious and life-threatening blood clot (thromboembolism). This may include pain and/or swelling of an arm or leg with warmth over the affected area, discoloration of an arm or leg, unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness, or weakness on one side of the body. If you are at risk, your doctor may decide to adjust the dose of ASCENIV. Your doctor will monitor you for any signs or symptoms of blood clots or poor blood flow in your arteries.

Always tell your doctor immediately if your medical history is similar to what is described here, and especially if you experience any of these symptoms while taking ASCENIV.

Kidney problems or failure. Kidney problems, kidney failure, and death may occur with use of human immune globulin products, especially those containing sucrose (sugar). ASCENIV does not contain sucrose.

If you have kidney disease or diseases with kidney involvement, your doctor should perform a blood test to assess your hydration level and kidney function before beginning immune globulin treatment and at appropriate intervals thereafter. If your doctor determines that kidney function is worsening, they may discontinue treatment. If your doctor determines you to be at risk, they may start your dose of ASCENIV at a safe level.

People taking human immune globulin products, including ASCENIV, may experience hyperproteinemia (high levels of protein in the blood), hyponatremia (low levels of sodium in the blood), and hyperviscosity (poor blood flow). Your doctor may perform certain blood tests and monitor you to minimize any of the above risks.

Aseptic meningitis syndrome (AMS). Aseptic meningitis is a non-infectious inflammation of the membranes that cover the brain. It causes a severe headache, which may occur with human immune globulin treatment, including ASCENIV. AMS usually happens within a few hours to 2 days after treatment. AMS is more commonly associated with higher doses of treatment and/or after rapid infusion. Your doctor may perform a neurological exam, including spinal tap (sampling fluid which surrounds the spinal cord) to evaluate your condition and to rule out other causes of meningitis.

Hemolysis. Hemolysis refers to the destruction of red blood cells. Immune globulin products, including ASCENIV, may contain certain antibodies that can result in the rupturing of red blood cells. Your doctor should monitor you for signs and symptoms of hemolysis, which may include additional confirmation tests.

Taking intravenous human immune globulin products may cause a build up of fluid in the lungs (pulmonary edema) that is unrelated to heart problems. Your doctor should monitor you for lung-related side effects and may conduct appropriate tests that can detect the presence of certain white blood cells (anti-neutrophils) in the drug or your blood. If needed, your doctor may decide to use oxygen or other respiratory methods to help your breathing.

Transmissible infectious agents. Because ASCENIV is made from human blood, it may carry a risk of transmitting infectious agents such as viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent, and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent. Your doctor will report to the manufacturer any cases of suspected infections spread by the product.

Interference with lab tests. Because ASCENIV contains a variety of antibodies that are infused into your body, blood tests to determine antibody levels may provide misleading interpretations. Be sure to always tell your doctor, nurse, or lab technician of any medicines you are taking and that you are using ASCENIV.

Interactions with medicines. ASCENIV can make vaccines (like measles, mumps, rubella, and chicken pox vaccines) less effective in your body. Before you get any vaccines, tell your healthcare provider that you take ASCENIV.

What are other possible side effects of ASCENIV?
In clinical studies of ASCENIV, some patients experienced the following:

- Headache
- Sinus inflammation (sinusitis)
- Diarrhea
- Intestinal lining inflammation caused by virus (gastroenteritis)
- Common cold (nasopharyngitis)
- Upper respiratory tract infection
- Bronchitis
- Nausea

These are not all the possible side effects of ASCENIV. Talk to your healthcare provider about any side effect that bothers you or that does not go away.

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.

For additional safety information about ASCENIV, please see full Prescribing Information at www.asceniv.com

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Features

Up Front

Departments

Columns

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About IG Living
IG Living magazine brings together patients, advocates and caregivers in the immune globulin (IG) community.

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Subscriptions to IG Living are free, and readers may subscribe at IGLiving.com or by calling (800) 843-7477 x1461.

The Realities of Chronic Illness: Are You Prepared?

PREPARING FOR the realities of living with a chronic illness can help you maintain a sense of control over your health — whether you're undergoing your first intravenous immune globulin (IVIG) therapy, dealing with health setbacks or resolving how to explain your illness to others.

While many of our readers have been receiving IG infusions for several if not many years, we want to help those brand new to IG treatment prepare for your first infusion. IVIG versus subcutaneous IG infusions are standard when first beginning IG therapy. And, understandably, this type of therapy is likely to cause anxiety since it's much different than just being prescribed an oral or injectable medicine; it's a therapy that usually requires going to an outpatient infusion setting or even a hospital, and it takes several hours. As such, we explain in our article "Navigating Your First Infusion: Preparation, Support and What to Expect" (p.24) what steps you can take to prepare for your first infusion, from understanding your treatment plan and reviewing your insurance to financial responsibility and ensuring you are comfortable during treatment. We also detail what happens during the infusion, what side effects may or may not occur and how to manage your care after the infusion. Importantly, we strongly suggest establishing a support network to help you get connected and take an active role in your care.

Whether you are newly diagnosed and first starting treatment or have been dealing with your illness for some time, learning how to share your diagnosis with others can be difficult. That's the case when deciding with whom to tell, as well as how and when. Our article "Managing Difficult Conversations: Telling Others About Your Diagnosis" (p.28) provides some guidelines for whom to confide in about your illness and why. We also outline some specific nuances when sharing with family, friends and children, as well as specific strategies for how to share.

No matter what stage you are in managing your condition, there will always be medical setbacks, and we recognize these setbacks can affect you physically, emotionally and mentally. That's why in our article "How to Handle Medical Setbacks" (p.34), we encourage you to build resilience to help you adapt when the unexpected happens. We also provide some healthy ways for you to cope, including taking a breath to help you regroup, looking at the situation from different angles, strategizing ways to move forward, leaning on others and avoiding negative thinking.

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

Ronale Tucker Rhodes

Ronale Tucker Rhodes, MS

IGLiving

Our mission is to support the IG community through education, communication and advocacy

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At **FFF Enterprises**, we understand the critical nature of your work. Every transaction you make provides essential plasma products for patients in need. That's why we are dedicated to being your reliable supplier of safe and effective plasma products, including immune globulin (IG), hyperimmune globulin, coagulation, and albumin therapies.

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Keeping Health Goals Alive Beyond January: Strategies for Patients with Chronic Illnesses

By Abbie Cornett, MBA

EVERY DEC. 31, I find myself scrambling for New Year's resolutions. Like many people, mine often revolve around getting into better shape, eating healthier and improving myself overall. Typically, these resolutions carry me through to about mid-February — especially those related to health. Unfortunately, my New Year's goals are mostly ridiculously overambitious! This inevitably leads to disappointment and frustration with myself when I fall short. For people with chronic illness, maintaining New Year's resolutions can be even more difficult!

Like everyone else, people with chronic illness need to set realistic goals that consider their unique challenges such as limited energy, unpredictable setbacks and the need for taking extra precautions to avoid germs and manage complex treatment schedules. Additionally, they need to consider adjustments for changing medications or side effects that can make consistency difficult.

These health challenges are why a flexible approach to your goals is essential. You must allow yourself to adjust your expectations on low-energy days or during illness flare-ups. Embracing flexibility and self-forgiveness allows you to adapt your goals to the realities of your illness and can help foster a path to success without the pressure of trying to be perfect.

Redefining your idea of progress is crucial to your success. Instead of aiming for major changes, try cultivating sustainable habits. Focus on small, consistent actions that contribute to your overall well-being. Progress can look different daily. Some days will be more challenging, but that's OK. Make sure your health goals are achievable even on low-energy days. For instance, rather than setting a goal of a daily 30-minute walk, aim for five minutes of movement as a baseline. You can always increase the time on days you feel good.

Stay focused on your own journey, and avoid comparing yourself to others. Everyone's path is unique. What matters most is that you're moving forward in your own way. If you can't meet a goal, be kind to yourself — no one else will mind if you miss a target you set. Treat each day as a fresh chance to try again.[1] Whether it's a New Year's resolution or a goal set anytime during the year, it's important to make sure your goals are practical and allow for flexibility to keep them sustainable:

- *Make them realistic*: Choose goals that acknowledge physical limitations and the unpredictability of health.
- *Focus on what's possible*: Prioritize daily habits, like hydration, mindful breathing or gentle stretching, that can be done consistently even if you don't feel well.
- *Celebrate wins*: Every small step counts and deserves recognition.
- *Create a backup plan*: Prepare for low-energy days by having backup goals. An example would be practicing mindfulness instead of yoga so forward progress feels achievable even on days when you are short on spoons.

Further, building a support system is vital. Involving family and friends can provide the motivation and encouragement you need to stay on track, especially during setbacks. However, support doesn't have to come only from friends and family; online support groups with people with whom you share similar experiences are also valuable. A strong support network reinforces commitment to goals and helps reduce feelings of isolation, making the journey easier and more manageable.

Don't fear using technology. Technology can be a powerful ally in managing health goals. Apps, calendar reminders and health-tracking devices can help keep you on track and make monitoring your progress easier. Gentle daily or weekly reminders can provide a sense of accomplishment and keep motivation high, even on difficult days.

The most important thing to remember when setting goals is that they are for you. It's OK to reassess and adjust them as your health or circumstances change. Be flexible, and don't feel guilty if you need to adjust. Health goals are a journey, not a race; focusing on well-being over perfection makes it easier to stay on track. Celebrate each small step forward, knowing these steady efforts can bring meaningful, lasting change. IG

References

1. 5 Tips for Ringing in the New Year with Health and Wellness. Prime Women, Jan. 1, 2024. Accessed at primewomen.com/wellness/health/tips-for-ringing-in-the-new-year-with-health-and-wellness.
2. Support Groups: Make Connections, Get Help. Mayo Clinic, March 16, 2023. Accessed at www.mayoclinic.org/healthy-lifestyle/stress-management/in-depth/support-groups/art-20044655.

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

Are Your Infusion Nurses and Pharmacists IG-Certified?

I don't have a nurse; I do subcutaneous immune globulin (SCIG) infusions, and I've been doing them since 2014. I have no idea if the nurses who were sent to train me over a four-week period were or not [certified] because I hadn't heard of this organization before.

The nurse sent to "train" me to do SCIG infusions seven years ago was not IG-certified, and it made a world of difference. Due to her lack of familiarity with SCIG infusions, she caused me to have a severe cellulitis reaction. IG certification should be the minimal standard for doing trainings in the future.

What's Your Worst Experience with a Doctor?

When I was first diagnosed with an incurable form of blood cancer, I went through three oncologists before finding the one who knew what to do to save my life. I was in remission for quite a long time thanks to [that doctor]; unfortunately, it's back with a vengeance and I will be going through treatment again. This time, I knew exactly who I needed to seek help from.

I went to an allergist because of bad reactions to a new inhaler. [After listening] to a 10-minute speech about all the new options available for asthma, the doctor handed me the same inhaler I just had the reactions to.

I was told for years that all my health issues would go away if I exercised more and lost weight. Finally, I had IgG testing by an allergist who discovered I have common variable immune deficiency. When I told my family doctor, he said, "Huh, who diagnosed you with that?" The doctors I have now are wonderful, and I appreciate them.

I was seeing an immunologist for seven years, and despite signs of having a primary immunodeficiency, she did not diagnose me until another patient was sent to her by an ENT for common variable immune deficiency (CVID). She suddenly realized that I had CVID, too. The other patient is a good friend now, and we put the timeline together since we both saw this doctor. I am sad that I could have been receiving treatment even years earlier. Instead, I had more infections and more antibiotics in those seven years. She should have known! My IgM was very low, and my IgG was borderline from the start.

Did You Know Leafy Greens Can Affect How Blood Thinners Work?

My mom could only have one-quarter cup a week of the vitamin K veggies.

I am on warfarin, and there are a lot of diet restrictions.

Yes, my sister's late father-in-law had to avoid tons of veggies.

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.

How Can I Receive Notifications About the IG Living Advocate Podcast Episodes?

I am subscribed to *IG Living* magazine, and was told I would be notified about upcoming podcast episodes, but I have not received any notifications. Could you let me know how I can confirm my subscription or troubleshoot this issue? Additionally, is there another way to stay updated about new episodes such as through email or social media?

Abbie: You can access the IG Living Advocate Podcast in several ways:

1) It is available on our website at www.igliving.com/life-with-ig/ig-living-advocate-podcast.html.
2) You can listen to the podcast on your preferred platform, including Apple Podcasts, Google Podcasts, Spotify, Castbox and Buzzsprout.
3) You can watch episodes on YouTube.

To receive updates about upcoming podcast episodes, please subscribe to *IG Living* magazine. If you're currently subscribed and do not receive notifications, I recommend checking your email's spam or promotions folder. You can also contact our support team to confirm your subscription status. We also regularly post updates about new episodes on our Facebook page at www.facebook.com/IGLivingMagazine. Follow us! It is a great way to stay in the loop.

Has the Deadline Been Extended for the IVIG Demonstration Project, and What Are the Eligibility Requirements?

My wife has been participating in the IVIG Demonstration Project. Could you provide an update on whether the project has been extended and share any additional information we should know about it? Also, what are the eligibility requirements?

Abbie: The Medicare Intravenous Immune Globulin (IVIG) Demonstration Project was started to assess the benefits of providing payment for items and services necessary for in-home administration of IVIG to treat primary immune deficiency diseases (PI). The project began in October 2014, was extended twice by Congress and was set to end on Dec. 31, 2023. However, as of Jan. 1, 2024, the services and supplies necessary for in-home administration of IVIG became a permanent Medicare benefit for patients with PI. This change means patients can continue to receive their IVIG treatments at home without the need to enroll in the now-concluded demonstration project. The permanent benefit includes coverage for the IVIG medication, supplies and nursing services required for home administration.[1]

Previously, to participate in the IVIG Demonstration Project, beneficiaries needed to meet the following criteria:[2]

- Be enrolled in Medicare Part B and covered under the traditional Medicare fee-for-service program (i.e., not enrolled in a Medicare Advantage plan).
- Have a diagnosis of PI.
- Not be covered under a home health episode of care at the time of service, since those services are already covered under the Medicare home health benefit.

For more detailed information about this permanent benefit and how it may affect your wife's care, you can visit the Centers for Medicare and Medicaid Services' Medicare IVIG Demonstration page.

1. Centers for Medicare and Medicaid Services. Medicare Intravenous Immune Globulin (IVIG) Demonstration. Accessed at www.cms.gov/priorities/innovation/innovation-models/ivig.
2. Centers for Medicare and Medicaid Services. Medicare Intravenous Immune Globulin (IVIG) Demonstration: Beneficiary Frequently Asked Questions. Accessed at www.cms.gov/priorities/innovation/innovation-models/ivig/faq.

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

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

Learning to Lean Into Joy

By Mairead McConnell, PhD

LET'S FACE IT: Life can be stressful, especially for those living with or caring for someone with chronic health conditions. We live in a world that is full of heartache and, at any given moment, there may be a long list of things that are going or can go wrong. Learning to make space for difficult emotions such as sadness, anger, fear and disappointment is an important part of healing and living well. However, we sometimes overlook the crucial and powerful role of positive emotions, like joy. Even if it doesn't come naturally, or it feels like there is little joy to be found, allowing and embracing joy is something you can learn to do.

Foreboding Joy

I often hear people say, "I don't want to get my hopes up," or "It's too good to be true; something bad is going to happen." These phrases are examples of what Brené Brown calls "foreboding joy."[1] When we have trained our brains to be continually vigilant for what can go wrong, it can feel uncomfortable, and even unsafe, to allow things to be good. In that sense, joy is a vulnerable experience; we wish it would last forever, but it rarely does. Even so, preventing ourselves from feeling genuine joy doesn't actually prevent future disappointment; it simply robs us of the pleasant experience in the moment. If you find yourself approaching joy with a sense of foreboding or "waiting for the other shoe to drop," consider taking simple steps to allow more joy in your life.

Let the Light In

As human beings, we are wired for survival, but not necessarily to be joyful. Therefore, leaning into joy is something that takes practice and discipline.

1) *Notice*. Every day presents opportunities to appreciate beauty and goodness, but we must pay attention. Take time each day to put down distractions like smartphones, screens or to-do lists. Look up and look around. Find one thing that you like or appreciate. It could be as simple as the light coming through the window on a spring morning, the sound of birds chirping, the scent of coffee brewing or your favorite pair of shoes.

Preventing ourselves from feeling genuine joy doesn't actually prevent future disappointment; it simply robs us of the pleasant experience in the moment.

2) *Set worry aside*. If you find that worry and foreboding interfere with your ability to experience joy, consider creating a "worry box." Write down each worry on a small piece of paper. Fold up the paper and place it into the box. As you close the lid, trust that the box is holding your worries so you don't have to. Anytime a worry arises, you can add it to the box and then continue on your way.

3) *Make time for what you love*. You probably make time to attend your doctor appointments and to brush your teeth twice daily, but how much time do you make for the activities you most enjoy? We often forget that wellness is about more than taking the right medications or eating well. Connecting with others, participating in your favorite hobby and laughing with friends are just as important for health and well-being.[2]

4) *Share it*. Joy, like love, gets bigger when we share it with others. Consider sharing with someone the thing you are excited or joyful about; it could be a family member, a friend or a stranger. Invite them to share theirs with you, and notice how the joy multiplies.

5) *Carry on*. If this doesn't come easy at first, know that you're not alone, and carry on. Sprinkling bits of joy into your life won't mean that every moment will be great, but it will increase your capacity to embrace good moments when they arrive. IG

Reference

1. Brown, B. *Daring Greatly: How the Courage to Be Vulnerable Transforms the Way We Live, Love, Parent, and Lead*. New York, NY: Gotham Books, 2012.
2. McQuoid, J. Finding Joy in Poor Health: The Leisure-Scapes of Chronic Illness. *Social Science & Medicine*, 2017;(183)88–96. Accessed at www.sciencedirect.com/science/article/abs/pii/S0277953617302745.

MAIREAD MCCONNELL, PhD, is a clinical psychologist and assistant professor at Banner University Medical Center in Tucson, Ariz. She specializes in health psychology and is passionate about helping patients live well while navigating the challenges of chronic illness.

What is HyQvia®?

HyQvia [Immune Globulin Infusion 10% (Human) with Recombinant Human Hyaluronidase] is a liquid medicine that is given under the skin (subcutaneously) to treat primary immunodeficiency (PI) in people 2 years and older.

IMPORTANT SAFETY INFORMATION

What is the most important information that I should know about HyQvia?

- HyQvia can cause blood clots.
- Call your healthcare professional (HCP) if you have pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s), unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness or weakness on one side of the body.
- Your HCP may perform blood tests regularly to check your IgG level.
- Do not infuse HyQvia into or around an infected or red swollen area because it can cause infection to spread.

Who should not take HyQvia?

Do not take HyQvia if you:

- Are allergic to IgG, hyaluronidase, other blood products, or any ingredient in HyQvia.

What should I avoid while taking HyQvia?

- HyQvia can make vaccines (like measles/mumps/rubella or chickenpox vaccines) not work as well for you. Before you get any vaccines, tell your HCP that you take HyQvia.

What should I tell my HCP before I start using or while using HyQvia?

Tell your HCP if you:

- Have or had any kidney, liver, or heart problems or history of blood clots because HyQvia can make these problems worse.
- Have IgA deficiency or a history of severe allergic reactions to IgG or other blood products.
- Are pregnant, trying to become pregnant or are breast feeding. It is not known whether HyQvia can harm the unborn baby or breastfed infant.

What are the possible or reasonably likely side effects of HyQvia?

HyQvia can cause serious side effects. If any of the following problems occur after starting HyQvia, stop the infusion immediately and contact your HCP or call emergency services:

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of irritation and swelling of the lining around your brain.
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s). These could be signs of a blood clot.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.

Meet the only monthly* subQ IG treatment and say hy to more time doing what you love.

HyQvia also offers protection from infection backed by data:

0.025 infections per year

This is equivalent to 25 acute serious bacterial infections (ASBIs) out of 1,000 patients over the course of the 12-month study period.

The FDA standard for efficacy—that is, if an immunoglobulin works—is less than 1 ASBI per year. In the clinical trial, people taking HyQvia experienced significantly less than that.

0 days in the hospital per year

There was a mean of 0.037 days spent in the hospital due to infection during the study.

<4 days off work or school per year

On average, patients taking HyQvia missed 3.31 days of work or school due to an infection.

- HyQvia was studied in a clinical trial of 83 people with PI, with the main goal of measuring how many acute serious bacterial infections (ASBIs) they experienced over the course of 1 year
- ASBIs are short-term but serious infections caused by bacteria that require immediate medical care
- ASBIs included 2 episodes of pneumonia, both treated as outpatients with oral antibiotics. An additional episode of pneumonia requiring hospitalization occurred during the ramp-up
- The most common general (systemic) side effects were headache, antibody formation against hyaluronidase (Hy), fatigue, nausea, fever, and vomiting. The most common side effects at the infusion site (local) were pain, redness, swelling, and itching

subQ IG=subcutaneous immune globulin.

IMPORTANT SAFETY INFORMATION (continued)

- Chest pain or trouble breathing, blue lips or extremities. These could be signs of a serious heart or lung problem.
- Fever over 100°F. This could be a sign of an infection.

After HyQvia infusion a temporary, soft swelling may occur around the infusion site, which may last 1 to 3 days, due to the volume of fluid infused. The following possible side effects may occur at the site of infusion and generally go away within a few hours, and are less likely after the first few infusions.

- Mild or moderate pain
- Redness
- Swelling
- Itching

The most common side effects of HyQvia are:

- Headache
- Fatigue
- Nausea
- Fever
- Vomiting

Antibodies to the hyaluronidase component of HyQvia were formed in some patients taking HyQvia. It is not known if there is any long-term effect. In theory, these antibodies could react with your body's own hyaluronidase (PH20). PH20 is present in the male reproductive tract. So far, these antibodies have not been associated with increased or new side-effects.

These are not all the possible side effects. Talk to your HCP about any side effect that bothers you or that does not go away.

Please see Important Facts about HyQvia on the following page.

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.

Register for a community educational program created for people living with PI—*My Life, My Story.*

Learn more about what it's like to infuse HyQvia from clinical nurse educators and other patients like you.

IMPORTANT FACTS about HYQVIA (Hi-Q-via) [Immune Globulin Infusion 10% (Human) with Recombinant Human Hyaluronidase] Solution, for subcutaneous administration

What is the most important information I should know about HYQVIA?

- HYQVIA can cause blood clots.
- Call your healthcare provider (HCP) if you have pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s), unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness or weakness on one side of the body.
- Your HCP may perform blood tests regularly to check your IgG level.
- Do not infuse HYQVIA into or around an infected or red swollen area because it can cause infection to spread.

What is HYQVIA?

HYQVIA is a liquid medicine containing immune globulin and Recombinant Human Hyaluronidase. HYQVIA is given under the skin (subcutaneously) to treat primary immunodeficiency (PI) in people 2 years of age and older. HYQVIA contains IgG antibodies, collected from human plasma donated by healthy people.

- The antibodies help your body to fight off bacterial and viral infections.
- The hyaluronidase is found in your body naturally. It's the first part of your two-part infusion. It temporarily opens the space under your skin (the subcutaneous space), allowing a larger amount of IgG to reach your subcutaneous tissue and be absorbed into your bloodstream.

What should I tell my HCP before I start using or while using HYQVIA?

Tell your HCP if you:

- Have or had any kidney, liver, or heart problems or history of blood clots because HYQVIA can make these problems worse.
- Have IgA deficiency or a history of severe allergic reactions to IgG or other blood products.
- Are pregnant, trying to become pregnant, or are breastfeeding. It is not known whether HYQVIA can harm the unborn baby or breastfed infant.

Who should not take HYQVIA?

- Do not take HYQVIA if you are allergic to IgG, hyaluronidase, other blood products, or any ingredient in HYQVIA.

How should I take HYQVIA?

- HYQVIA is infused under the skin (subcutaneously) up to once every 4 weeks.
- You can get HYQVIA at your HCP's office, clinic, or hospital.
- You can use HYQVIA at home. You and your HCP will decide if home self-infusion is right for you.

What are the possible or reasonably likely side effects of HYQVIA?

After HYQVIA infusion a temporary, soft swelling may occur around the infusion site, which may last 1 to 3 days, due to the volume of fluid infused.

The following local reactions may occur at the site of infusion and generally go away in a few hours. Local reactions are less likely after the first few infusions.

- Mild or moderate pain
- Redness
- Swelling
- Itching

The most common side effects of HYQVIA are: headache, fatigue, nausea, fever, and vomiting.

Antibodies to the hyaluronidase component of HYQVIA were formed in some patients taking HYQVIA. It is not known if there is any long-term effect. In theory, these antibodies could react with your body's own PH20. PH20 is present in the male reproductive tract. So far, these antibodies have not been associated with increased or new side effects.

Call your HCP or go to your emergency department right away if you get:

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of irritation and swelling of the lining around your brain.
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s). These could be signs of a blood clot.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.
- Chest pain or trouble breathing, blue lips or extremities. These could be signs of a serious heart or lung problem.

These are not all of the possible side effects for HYQVIA. You can ask your HCP for information that is provided to HCPs. Talk to your HCP about any side effects that bother you or that don't go away.

How do I store HYQVIA?

Store HYQVIA refrigerated or at room temperature.

- You can store HYQVIA in the refrigerator (36°F to 46°F [2°C to 8°C]) for up to 36 months.
- You can store HYQVIA at room temperature (up to 77°F [25°C]) for up to 3 months during the first 24 months from the date of manufacturing (Mfg Date) printed on the carton.
- Do not return HYQVIA to the refrigerator if you take it out to room temperature.

Check the expiration date on the carton and vial label. Do not use HYQVIA after the expiration date.

Do not freeze.

Protect from light. You can use the original HYQVIA containers to protect it from light.

How do I get more information about HYQVIA?

The risk information provided here is not comprehensive. To learn more, talk about HYQVIA with your HCP or pharmacist. The FDA-approved Full Prescribing Information, including Information for Patients, can be found at www.HYQVIA.com or by calling 1-877-TAKEDA7 (1-877-825-3327).

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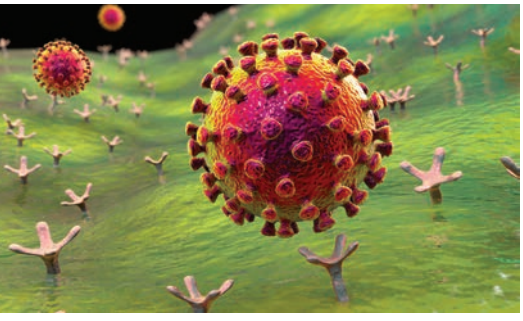
US-HYQ-0299v5.0 02/24

SARS-CoV-2 and COVID-19: Consequences of Autoimmunity, Specifically Regarding ACE2

By Terry O. Harville, MD, PhD



IN THE last issue, we discussed the abnormal immune response to SARS-CoV-2 (SARS2) and how this results in autoimmunity, with ACE2 a major target for this. We will now discuss the consequences of this.

Angiotensin-converting enzyme (ACE) and ACE2 are important members of the renin-angiotensin system (RAS), also termed the renin-angiotensin-aldosterone system (RAAS), whose role is to regulate blood pressure. When blood pressure is low, cells in the kidney detect this and release renin into the blood. Renin then cleaves angiotensinogen (which is made in and released from the liver into the blood) into angiotensin I (which does not have activity on its own). Angiotensin I circulates with the blood to the lungs, where ACE cleaves angiotensin I into angiotensin II. Subsequently, angiotensin II is converted to angiotensin III, which results in the production and release of aldosterone from the adrenal cortex, exerting a sodium retention effect on the kidneys for increasing blood pressure. Angiotensin III also has a direct "pressor" effect on the vasculature for increasing blood pressure. This is the manner for which normal blood pressure is maintained.

Unfortunately, angiotensin II is highly pro-inflammatory, affecting lymphocytes, endothelial cells and smooth muscle cells. Thus, angiotensin II can activate lymphocytes toward increased inflammation and possible autoimmunity. The endothelial cells line the blood vessels and, when inflamed, can affect proper blood flow to the tissues, as well as be at increased risk for blood clot formation. The smooth muscle cells have an important role in controlling the size of the arterial blood vessels. As such, angiotensin II can result in constriction, which can impair blood flow to vital organs, including the heart, kidneys, lungs and brain.

ACE2's main role is to counteract the effects of angiotensin II. ACE2 cleaves angiotensin II into angiotensin. In contrast to angiotensin II, angiotensin has anti-inflammatory and vasodilatory effects, making it important and beneficial.

Infection with SARS2 results in the formation of anti-ACE2 antibodies, which interfere with ACE2 function.[1] As a consequence, blood pressure regulation is disrupted and vascular inflammation is increased. This may be an important reason for the increased symptoms of postural orthostatic tachycardia syndrome (POTS) in patients with COVID and long COVID. This can also be part of dysautonomia occurring.

Myoung Hwa Lee and his group studied the brains from people who had COVID infections and passed away.[2] Regardless of the severity of COVID infection, microclotting was found, especially in the brain stem. (Mild infections had the same level of microclotting as severe infections.) They note that the anti-ACE2 antibodies were likely playing an important role in the clotting process due to complement activation, and the disruption of blood flow in the brain can have adverse consequences such as brain fog. Dysfunction of the brain stem can also result in dysautonomia.

Therefore, the increased presence of angiotensin II and the microclotting due to anti-ACE2 activating complement in the blood vessels appear to be a major cause of the dysautonomia and brain fog in long COVID.

This has been further corroborated by Margherita Bellucci and coworkers: They also report anti-ACE2 antibodies having a deleterious effect on the brain. Post-SARS2 infection and post-vaccine-related neurological complications share clinical features and the same positivity to anti-ACE2 antibodies.[3]

We will discuss more about SARS2 and COVID-19 in the next issue.

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TERRY O. HARVILLE, MD, PhD, is medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences and a consultant for immunodeficiencies, autoimmunities and transplantation.

Biologics and Biosimilars: What's the Difference?

By Michelle Greer, RN, IgCN

MANY TYPES of diseases and conditions are treated with biologics and, increasingly, biosimilars. Here's what patients need to know about the similarities and differences between the two.

Biologics vs. Biosimilars

Biologics are medicines made from living organisms or grown in a lab from living organisms; they are given as injections or infusions. While conventional drugs such as aspirin are made from small molecules (chemicals), biologics are made from large molecules (animal cells, microorganisms or plant cells) and are, therefore, more complicated and expensive to produce.

Biosimilars are biologic medicines that are made to be *highly similar* to other biologic medicines that have already been approved by the U.S. Food and Drug Administration (FDA). Biosimilars are made from the same source as the original biologic (or "reference product"). A biosimilar is very similar — but not identical — to its reference product.

Are Biosimilars Generic Biologics?

Not exactly. Brand name conventional medications have generic versions, which are exact replications of the branded drug. Biologics do not have generic versions because it is not possible to manufacture identical molecules for biologic agents due to the complexity of their molecular structure and manufacture.[1]

Conventional small molecule drugs have a low molecular weight (mainly chemicals), so they can be precisely replicated from one batch to another, but biologics are made from large molecules (living organisms), so they have a more complicated structure and cannot be precisely replicated and thus can vary from batch to batch. This makes the manufacturing process of biosimilars more complex, time-consuming and expensive than the manufacturing process of conventional drugs. However, like generic drugs, biosimilars were created to offer less expensive treatment options for conditions traditionally treated with biologics.[2]

How Are Biosimilars Approved?

Both biologics and biosimilars are regulated by FDA. For biosimilars to be approved by FDA, studies must show there are no differences in the safety or effectiveness between them and their reference products.[3] They must be made from the same living organisms; administered in the same manner, strength and dose; provide the same benefits for treating condition(s); and have the same side effect profiles, with no expectation of new or worsening reactions.

FDA approves biosimilar products through a shortened process. Per the Biologics Price Competition and Innovation Act of 2010, clinical trials proving safety and efficacy are not required for biosimilars; instead, only their similarity to the reference products must be proved (which results in lower clinical spend and a less expensive medicine).

What Do Biologics and Biosimilars Treat?

Biologics are safe and effective treatment options for many illnesses such as autoimmune diseases, cancer, chronic skin and bowel diseases, arthritis and kidney conditions. There are 61 biosimilar medications approved today available to treat many conditions, including rheumatoid arthritis, inflammatory bowel disease, psoriasis and other skin conditions, diabetes, some types of cancer, certain kidney conditions and macular degeneration.

Interchangeability and Substitutions

Some biosimilars are interchangeable, which means products may be substituted by a pharmacist without consulting the prescriber (depending on state pharmacy laws). This is similar to substituting a generic drug for a brand name drug. Substitution can improve access to biologics, which can make them more affordable for patients.

It's a common misconception that because interchangeable biosimilars meet additional statutory requirements, they are safer or more effective than non-interchangeable biosimilars. However, both biosimilars

and interchangeable biosimilars can be used in place of a reference product and are considered as safe and effective as the reference product.[5] *The Purple Book* is an FDA database that contains extensive information on biologics and their biosimilar and interchangeable biosimilar products. The database is available for free at purplebooksearch.fda.gov.

What This Means for Patients

So how can patients become comfortable and confident if prescribed a biosimilar?

First, remember that FDA ensures biosimilars meet the same requirements as their reference products: They must be made from the same living sources; provide the same benefits, including efficacy in the treatment of conditions; be administered in the same manner at the same dose; and have no new or worsened side effects.

Second, biosimilars increase access to much-needed biologic medications. They create more treatment options in conditions in which options might be limited, expensive or challenging to get approval. Due to the expense of biologics and the relative cost-savings of biosimilars, some health plans will require biosimilar use prior to the original biologic for this reason. This will vary between health plans.

Patients may still be nervous, however, and can be strongly influenced by their physician's opinion or attitude regarding biosimilar use. Providers (i.e., physicians, nurses and pharmacists) should provide understandable and up-to-date information tailored to the individual patients' needs. They should reassure patients with an open, empathetic and positive attitude.[6]

FDA ensures biosimilars meet the same requirements as the reference products.

Better Access to Lifesaving Care

According to the 2023 U.S. Generic and Biosimilar Medicines Savings Report, the average sales price for biosimilars is on average 50 percent less than the price of the branded biologic reference products at the time of biosimilar launch, and competition from biosimilars has reduced the average sales price of their corresponding branded biologic by an average of 25 percent.[7] However, another article published in the *Journal of the American Medical Association* in 2024 posed the following question among commercially insured patients in the U.S. who are using biologic drugs: Is competition by biosimilars associated with lower out-of-pocket (OOP) spending? Findings from this study suggest that the introduction of biosimilar competition did not systematically lower patient OOP spending on biologics, highlighting the need for targeted policy interventions to ensure savings from biosimilar competition improves affordability for patients.[8]

Overall, biosimilars are a positive movement toward increasing access to highly specialized biologic medications at potentially lower costs for patients, and more broadly, they may also decrease healthcare spending without impacting effective treatments. FDA-approved biosimilars are considered safe, effective and offer another option for accessible treatment. And with a shortened window toward approval, we are likely to see a continued rollout of these lifesaving medications.

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MICHELLE GREER, RN, IgCN, is senior vice president of sales at Nufactor, a specialty infusion company.

EDUCATION

Webinar Series Developed to Educate Physicians About PI

To help educate physicians about primary immunodeficiency diseases, the Immune Deficiency Foundation collaborated with Project ECHO at Penn State College of Medicine to produce a 10-episode webinar series that is available to view for free online. Titled "Understanding Primary Immunodeficiency," topics include immune system basics, signs and symptoms, steps in diagnosis, antibody deficiencies, treatment options, newborn screening, comorbidities, teamwork and social determinants of health.

The webinar series can be viewed at www.youtube.com/watch?v=l0kh3ACMcoc&list=PL8WZIZu0uFGDGENafTjiDjHR0i3EfCgS.

Gawel, R, Brock, C, and Felix, L. Q&A: Video Series Educates Physicians About Primary Immunodeficiencies. Healio, Oct. 30, 2024. Accessed at www.healio.com/news/allergy-asthma/20241030/qa-video-series-educates-physicians-about-primary-immunodeficiencies.

RESEARCH

Study Shows Stem Cell Editing Repairs Severe Immunodeficiency

In a study published in *Science Translational Medicine*, researchers developed an efficient method to repair recombination activating gene 1 (RAG1) genes in immune cell progenitors called hematopoietic stem cells (HSC) taken from severe combined immunodeficiency (SCID) patients, and revealed that they could restore immune function in mice.

RAG1 is central to this shapeshifting behavior of using B-cell and T-cell receptors, which take on various shapes to bind to different antigens on foreign invaders. It shuffles the order of DNA sequences in the genes for these receptors, producing multiple versions of the immune receptors that can bind staggering combinations of antigens. However, some people carry mutations in RAG1 that prevent the enzyme from recombining the DNA sequences that code for these receptors. Without properly functioning receptors, B and T cells fail to develop, leading to SCID, a condition in which even the mildest of infections can prove lethal.

In their study, the researchers set out to overcome some of the challenges with editing the RAG1 gene that researchers previously faced. In the past, scientists have taken healthy, functional HSC and inserted them into SCID-model mice, but they often get destroyed by other types of immune cells that recognize the transplants as foreign. Normally, doctors use immunosuppressants like chemotherapy before transplantation to deplete immune cells, but this isn't an option for SCID patients.

So, they modified a SCID patient's own stem cells to express a functional RAG1 gene. While other research groups had successfully added RAG1 to patient HSC, they were unable to properly regulate expression of the gene, and therefore couldn't ensure that the stem cells were safe or would effectively replenish B and T cells. Introducing the gene into the wrong site in the genome may have partly caused this shortcoming.

Rather than adding a functional copy of RAG1, the researchers decided to modify the existing copy, ensuring that the regulatory networks remained intact. They used the clustered regularly interspaced short palindromic repeats (CRISPR-Cas9) system to cut out a large section of the mutant gene, and then provided cells with the correct DNA sequence using a lentiviral delivery system. Since the correct sequence was nearly identical to the original gene, the cell could swap the sequences unassisted using homology-directed repair (HDR), a built-in DNA repair pathway that fixes double-strand DNA breaks using complementary DNA as a template.

They then inserted a backwards green fluorescent protein (gfp) gene flanked by sequences that RAG1 recognizes. They found the edited RAG1 inverted gfp comparably to RAG1 in HSC from healthy donors, thereby switching it to an "on" state, resulting in a functional gfp gene.

Next, they checked that these edited cells could restore immune function in the body. They transplanted these edited human cells into SCID-model mice and found that B and T cells spiked to levels similar to those seen in mice that received HSC from healthy donors. "What's intriguing from the study is that we don't need to correct all the stem cells. If we manage to correct at least 10 percent of the stem cells, this is going to give us a therapeutic benefit," said Saravanabhavan Thangavel, MD, a geneticist at the Institute of Stem Cell Research and Regenerative Medicine who was not involved with the work. However, he also mentioned, "We need to track the HDR-edited cells long term."

Nahas, K. Stem Cell Editing Repairs Severe Immunodeficiency. The Scientist, March 20, 2024. Accessed at www.the-scientist.com/stem-cell-editing-repairs-severe-immunodeficiency-71733.

RESEARCH

Study Finds Patients Prefer SCIG to IVIG

A recent review of 14 studies published in the *Journal of Comparative Effectiveness Research* has found a majority of patients in 13 of the 14 studies prefer subcutaneous administration of biologics, often administered at home with prefilled syringes, to intravenous administration at a hospital. Specifically, the meta-analysis found 83 percent of patients with autoimmune disease and 80 percent of those with a primary immunodeficiency (PI) preferred subcutaneous administration.

According to the researchers, these results correspond with current thinking about subcutaneous administration, which can be performed at home and therefore has the benefit of convenience. However, Ann Gardulf, RN, PhD, of the Karolinska Institutet in Stockholm, Sweden, and her colleagues note that reviews of studies of patients with specific conditions — PI and rheumatoid arthritis — have arrived at similar conclusions, but when considering treatment effectiveness and general health as part of the measurement, there is little difference between subcutaneous and intravenous administration.

"Patient preferences and circumstances can be very individual, and self-administration may not be suitable for all patients," the researchers wrote. In addition, they note that subcutaneous

administration is most likely to be most suitable for patients who are confident about the effectiveness of the treatment and wish to have greater autonomy. IG

Wherwein, P. Patients Prefer Sub-Q to IV, Review Finds. Managed Healthcare, Oct. 9, 2024. Accessed at www.managedhealthcareexecutive.com/view/patients-prefer-sub-q-to-iv-review-finds.

MEDICINES

FDA Grants Breakthrough Designation to Nipocalimab for Sjögren's Disease

The U.S. Food and Drug Administration (FDA) granted a breakthrough therapy designation to nipocalimab for the treatment of Sjögren's disease (SjD), a chronic autoimmune disease that causes symptoms such as mucosal dryness, joint pain and fatigue, and that may also impact multiple organ systems, including the joints, lungs, kidneys and nervous system. Nipocalimab is designed to block FcRn and reduce levels of circulating immunoglobulin G (IgG) antibodies, potentially without impact on other immune functions.

FDA's decision was supported by positive results from the mid-stage DAHLIAS study evaluating its effects in more than 160 adults with moderately-to-severely active primary SjD who were seropositive for anti-Ro60 and/or anti-Ro52 IgG antibodies. Results of the study demonstrated a statistically significant and clinically meaningful improvement in ClinESSDAI score, which measures disease activity across 11 organ systems, in nipocalimab-treated SjD patients versus placebo at 24 weeks compared to baseline.

"[This] announcement marks an important step forward in the continued research and development of nipocalimab, the first investigational FcRn blocker to demonstrate positive results in a Phase II study in adult patients with moderate-to-severe SjD," said Terence Rooney, vice president of rheumatology and immunology disease area leader at J&J Innovative Medicine. "With no treatments currently approved that may directly address the underlying cause(s) of the disease, innovation is critically needed to improve patient outcomes in SjD."

A Phase III trial of the candidate in SjD is currently underway.

J&J Nipocalimab Granted FDA Breakthrough Designation for Sjögren's Disease. PM Live, Nov. 13, 2024. Accessed at pmlive.com/pharma_news/jjs-nipocalimab-granted-fda-breakthrough-designation-for-sjogrens-disease.

RESEARCH

Vitamin D Deficiency May Trigger Autoimmune Conditions

Researchers from McGill University in Canada have made progress toward understanding how vitamin D deficiency early in life can trigger problems with the body's immune system. By studying mice genetically engineered not to produce vitamin D naturally, they found the thymus — a small organ that trains the immune system — ages faster in these mice, allowing self-attacking immune cells to run rampant. Specifically, the mice unable to produce vitamin D ended up with a thymus that was smaller and contained fewer cells, and showed biological signs of premature aging in the organ and lower levels of a key autoimmune regulator.

The thymus helps educate the T cell defense force in the body not to attack healthy cells, and vitamin D is closely linked to this process. The findings show how vitamin D deficiency can result in less protection against autoimmune diseases. "An aging thymus leads to a leaky immune system," said John White, PhD, a physiologist at McGill University. "This means the thymus becomes less effective at filtering out immune cells that could mistakenly attack healthy tissues, increasing the risk of autoimmune diseases like type 1 diabetes."

This has only been shown in mice so far, but the human thymus functions in a similar way, so there are good reasons to believe the same biological processes are at play. The researchers already have plans to investigate how vitamin D affects the human thymus, which has not been studied before.

"Our findings bring new clarity to this connection and could lead to new strategies for preventing autoimmune diseases," said Dr. White.

Nield, D. Vitamin D Deficiency Can Trigger Autoimmune Conditions. Here's Why. Science Alert, Nov. 3, 2024. Accessed at www.sciencealert.com/vitamin-d-deficiency-can-trigger-autoimmune-conditions-heres-why.

MEDICINES

FDA Approves Dupixent for COPD

The U.S. Food and Drug Administration (FDA) has granted approval for a new indication for Sanofi and Regeneron Pharmaceuticals' drug Dupixent. Already approved for use in treating many skin and lung disorders, the new FDA approval permits Dupixent's use as an add-on maintenance treatment for adults with chronic obstructive pulmonary disease (COPD), a progressive lung condition that is not adequately controlled by other available therapies. The every-other-week injectable drug is now the first biologic therapy approved by FDA for treating COPD.

Dupixent is an antibody designed to block IL-13 and IL-4, two signaling pathways that drive inflammation. In COPD, the drug is intended to specifically address type 2 inflammation, an excessive immune response characterized by accumulation of certain immune cells in tissue. Dupixent's FDA approval in COPD covers the drug's use to treat patients whose disease is driven by immune cells called eosinophils.

FDA approval of Dupixent in COPD is based on results from two Phase III studies that compared the drug to a placebo in adults who were currently receiving standard-of-care inhaled therapy. Results for both studies showed statistically significant reductions in the annualized rate of moderate or severe COPD exacerbations measured over one year.

Dupixent was first approved in 2017 for treating atopic dermatitis, followed by asthma in 2018. With the latest FDA approval, it can now treat six respiratory and dermatological conditions. According to Paul Rowe, MD, ATSF, Sanofi's head of medical affairs specialty care North America, "Given success we've had in these other disorders, we think there's a strong rationale for COPD as well." IG

Vinluan, F. Sanofi and Regeneron Biologic Drug Dupixent Notches a New FDA Approval in COPD. MedCityNews, Sept. 29, 2024. Accessed at medcitynews.com/2024/09/sanofi-regeneron-copd-dupixent-fda-approval-biologic-drug.

FDA-approved for adult and pediatric patients aged 2 years and older with primary immunodeficiency (PI)

cutaquig®
Immune Globulin Subcutaneous (Human)-hipp,16.5% solution

Count the reasons

to ask your care team about cutaquig

1 hour or less to **complete infusion***

2 or fewer **infusion sites**†‡

3 **flexible dosing** schedule options§

Not an actual patient.

*The estimated infusion duration for a 13 g (78 mL) weekly dose is approximately 45 minutes in an adult patient using 2 infusion sites, if tolerated, not including setup time.
†Depending on your dose and dosing schedule selected.
‡Most infusions only need 2 or fewer infusion sites.
§Every-other-week, weekly, or frequent dosing (2-7 times a week).

Scan to visit **CutaquigInfo.com** to learn more.

INDICATIONS AND USAGE

CUTAQUIG (Immune Globulin Subcutaneous [Human] - hipp) is a 16.5% immune globulin solution for subcutaneous infusion indicated for treatment of primary humoral immunodeficiency (PI) in adults and pediatric patients 2 years of age and older.

There are many forms of PI. Certain types of PI are associated with low immunoglobulin G (IgG), which are proteins that help fight infection.

CUTAQUIG is a liquid medicine for infusion that contains immunoglobulin G (IgG), which are proteins that help fight infection. It is made from human plasma that is donated by healthy people and contains antibodies that replace the missing antibodies in patients with PI.

CUTAQUIG is given under the skin (subcutaneous). Most of the time, infusions under the skin are given at home by self-infusion or by a caregiver. Only use CUTAQUIG by yourself after you have been instructed on use by a healthcare provider (HCP).

IMPORTANT SAFETY INFORMATION

WARNING: THROMBOSIS

See full Prescribing Information for complete BOXED WARNING

- **Thrombosis may occur with immune globulin products, including CUTAQUIG. 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 CUTAQUIG 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 of hyperviscosity.**

What is the most important information I need to know about CUTAQUIG?

CUTAQUIG can cause the following serious reactions:

- Severe allergic reactions causing difficulty in breathing or skin rashes
- Blood clots in the heart, brain, lungs, or elsewhere in the body
- Severe headache, drowsiness, fever, painful eye movements, or nausea and vomiting
- Decreased kidney function or kidney failure
- Dark colored urine, swelling, fatigue, or difficulty breathing

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

What should I know while taking CUTAQUIG?

- CUTAQUIG can make vaccines (like measles/mumps/rubella or chickenpox vaccines) not work as well for you. Before you get any vaccines, tell your HCP that you take CUTAQUIG
- Tell your HCP if you are pregnant, or plan to become pregnant, or if you are nursing

CUTAQUIG can cause serious side effects. If any of the following problems occur after starting CUTAQUIG, contact your HCP or call emergency services. If any of the following problems occur during CUTAQUIG infusion, stop the infusion immediately and contact your HCP or call emergency services:

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting, or dizziness. These could be signs of a serious allergic reaction
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of irritation and swelling of the lining around your brain
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem
- Pain, swelling, warmth, redness, or a lump in your legs or arms. These could be signs of a blood clot
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem
- Chest pain or trouble breathing, or blue lips or extremities. These could be signs of a serious heart or lung problem
- Fever over 100°F. This could be a sign of an infection

Ask your HCP whether you should have rescue medications available, such as antihistamines or epinephrine.

What are the possible or reasonably likely side effects of CUTAQUIG?

The most common side effects of CUTAQUIG are:

- Infusion site reactions (including but not limited to redness, swelling, itching, fluid in tissue, pain, mass, bruising)
- Headache
- Elevated body temperature

One or more of the following possible side effects may occur at the site of infusion; these may go away within a few hours and are less likely after the first few infusions:

- Mild or moderate pain
- Redness
- Itching

These are not all the possible side effects. Talk to your HCP about any side effect that bothers you or that does not go away.

Patients should always ask their doctors for medical advice about adverse events.

You may report an adverse event related to Pfizer products by calling 1-800-438-1985 (US only). If you prefer, you may contact the US Food and Drug Administration (FDA) directly. The FDA has established a reporting service known as MedWatch where healthcare professionals and consumers can report problems they suspect may be associated with the drugs and medical devices they prescribe, dispense, or use. Visit www.fda.gov/MedWatch or call 1-800-FDA-1088.

CUTAQUIG® is a registered trademark of Octapharma AG.

Please see brief summary of Full Prescribing Information on following page and Full Prescribing Information, including complete BOXED WARNING and Patient Information and Instructions for Use, at CutaquigInfo.com.

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PP-CQG-USA-0561

March 2023

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CONSUMER BRIEF SUMMARY

(CUTAQUIG: *cute-ah-quig*)

This brief summary highlights the most important information about CUTAQUIG. Please read it carefully before using CUTAQUIG and each time you get a refill, as there may be new information. This Patient Information does not take the place of talking with your healthcare provider about your medical condition or your treatment. If you have any questions after reading this, ask your healthcare provider. For more information, go to www.CutaquigInfo.com.

What is CUTAQUIG?

CUTAQUIG is a ready-to-use liquid solution of immunoglobulin G (IgG), also called antibodies, which protects the body against infection. CUTAQUIG is used to treat adult patients and pediatric patients 2 years of age and older with primary humoral immunodeficiency (PI).

There are many forms of PI. The most common types of PI result in an inability to make a very important type of protein called antibodies, which help the body fight off infections from bacteria or viruses. Regular administration of CUTAQUIG has been demonstrated to help your body to fight bacteria and viruses that cause infections. CUTAQUIG is made from human plasma that is donated by healthy people. CUTAQUIG contains antibodies collected from these healthy people; these antibodies replace the missing antibodies in patients with PI.

WARNING: THROMBOSIS

See full Prescribing Information for complete BOXED WARNING

- **Thrombosis may occur with immune globulin products, including CUTAQUIG. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling central vascular catheters, hyperviscosity, and cardiovascular risk factors.**
- **For patients at risk of thrombosis, administer CUTAQUIG 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 of hyperviscosity.**

Who should NOT use CUTAQUIG?

Do not use CUTAQUIG if you have ever had a severe allergic reaction to immune globulin or other blood products.

Tell your healthcare provider if you:

- Ever had any severe reaction to other immune globulin medicines
- Were told that you have a condition called IgA deficiency
- Have a history of heart or blood vessel disease
- Have had blood clots or thick blood
- Have been immobile for some time

CUTAQUIG can cause serious side effects. If any of the following problems occur after starting CUTAQUIG, contact your HCP or call emergency services. If any of the following problems occur during CUTAQUIG infusion, stop the infusion immediately and contact your HCP or call emergency services:

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- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem
- Chest pain or trouble breathing, or blue lips or extremities. These could be signs of a serious heart or lung problem
- Fever over 100°F. This could be a sign of an infection

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

What should I tell my healthcare provider before using CUTAQUIG?

Talk to your healthcare provider about any medical conditions that you have or have had.

Tell your healthcare provider:

- That you are taking CUTAQUIG before you get a vaccination, as vaccines may not work while you are taking CUTAQUIG.
- About all of the prescription and non-prescription medicines you take, including over-the-counter medicines, dietary supplements, or herbal medicines.
- If you are pregnant, plan to get pregnant, or if you are nursing because CUTAQUIG might not be right for you.
- If you have diabetes. If you need to do glucose testing, your healthcare provider may tell you to use a different way to monitor your blood sugar levels on the day that you receive a CUTAQUIG infusion. Some types of blood glucose testing systems (glucometers) can falsely interpret the maltose contained in CUTAQUIG as glucose. If you are uncertain, ask your healthcare provider which glucose testing system you can use while using CUTAQUIG.

The most common side effects that may occur with CUTAQUIG are:

- Infusion site reactions (including but not limited to redness, swelling, itching, fluid in tissue, pain, mass, bruising)
- Headache
- Elevated body temperature

One or more of the following possible side effects may occur at the site of infusion; these may go away within a few hours and are less likely after the first few infusions:

- Mild or moderate pain
- Redness
- Itching

These are not all the possible side effects. Talk to your HCP about any side effect that bothers you or that does not go away. If you encounter any problems or experience side effects during or after the infusion, contact your healthcare provider. When doing so, keep your treatment diary or logbook with you to be able to give all necessary information.

Patients should always ask their doctors for medical advice about adverse events.

You may report an adverse event related to Pfizer products by calling 1-800-438-1985 (US only). If you prefer, you may contact the US Food and Drug Administration (FDA) directly. The FDA has established a reporting service known as MedWatch where healthcare professionals and consumers can report problems they suspect may be associated with the drugs and medical devices they prescribe, dispense, or use. Visit www.fda.gov/MedWatch or call 1-800-FDA-1088.

This brief summary is based on the CUTAQUIG Prescribing Information (October 2021).

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RESEARCH

Keto Diet May Calm Overactive Immune System

Scientists at the University of California, San Francisco, have discovered that a ketogenic diet (keto diet) may calm an overactive immune system and help people with diseases such as multiple sclerosis (MS). In the study, they found a keto diet makes the gut and its microbes produce two factors that reduced symptoms of MS in mice, which may point toward a new way of treating MS and other autoimmune disorders in humans with supplements.

The keto diet severely restricts carbohydrate-rich foods, but allows unlimited fat consumption. Without carbohydrates to use as fuel, the body breaks down fat instead, producing compounds called ketone bodies. Ketone bodies provide energy for cells to burn; they can also change the immune system.

In the mouse model, researchers found that mice that produced more β-hydroxybutyrate (βHB), a particular ketone body, had less severe disease. The additional βHB also prompted the gut bacterium Lactobacillus murinus to produce a metabolite called indole lactic acid (ILA), which blocked the activation of T helper 17 immune cells that are involved in MS and other autoimmune disorders.

In this study, the team looked at how the ketone body-rich diet affected mice that were unable to produce βHB in their intestines and found that their inflammation was more severe. But when the researchers supplemented their diets with βHB, the mice got better.

To find out how βHB affects the gut microbiome, the team isolated bacteria from the guts of three groups of mice that were fed either the keto diet, a high-fat diet or the βHB-supplemented high-fat diet. Then, they screened the metabolic products of each group's distinct microbes in an immune assay and determined the positive effects of the diet were coming from a member of the Lactobacillus genus: L. murinus.

Genome sequencing and mass spectrometry confirmed that the L. murinus they found produced ILA, which is known to affect the immune system. Finally, the researchers treated the MS mice with either ILA or L. murinus, and their symptoms improved.

"What was really exciting was finding that we could protect these mice from inflammatory disease just by putting them on a diet that we supplemented with these compounds," said Peter Turnbaugh, PhD, of the Benioff Center for Microbiome Medicine. "The big question now is how much of this will translate into actual patients. But I think these results provide hope for the development of a more tolerable alternative to helping those people than asking them to stick to a challenging restrictive diet." IG

University of California, San Francisco. How the Keto Diet Could One Day Treat Autoimmune Disorders. Science Daily, Nov. 4, 2024. Accessed at www.sciencedaily.com/releases/2024/11/241104112030.htm.

RESEARCH

Interleukin-2 Shows Promise for Targeted Treatment of Autoimmune Diseases

A team of researchers from the department of immunology at the University Hospital Zurich and the University of Zurich tested the effect of low-dose interleukin-2 in 12 patients with systemic lupus erythematosus. They found that the therapy led to improvement in patient symptoms and an increase in regulatory T cells. Researchers showed that interleukin-2 activated the regulatory T cells and that specific subtypes with migration properties into specific tissues increased as a result. Using skin biopsies from patients before and after interleukin-2 therapy, the researchers also demonstrated the proliferation of these regulatory T cells in tissue affected by the disease.

The study showed that low-dose interleukin-2 can be used to inhibit harmful immune cell activity without restricting the immune defense and demonstrates the potential of interleukin-2 immunotherapy. IG

Novel Treatment Approach for Autoimmune Diseases with Interleukin-2. Universitats Spital Zurich news release, Aug. 27, 2024. Accessed at www.usz.ch/en/novel-treatment-approach-for-autoimmune-diseases-with-interleukin-2.

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Navigating Your First IVIG Infusion: Preparation, Support and What to Expect

Being well-prepared for your first infusion can help to ensure a successful outcome. These tips can prepare you for every step of the journey.

By Abbie Cornett, MBA

STARTING A NEW treatment can be overwhelming, especially when it involves something unfamiliar such as an intravenous immune globulin (IVIG) infusion. For patients with immune deficiencies or autoimmune conditions, IVIG can be a lifesaving therapy.[1] However, if it's your first time, you may have questions or even feel nervous about what to expect. Knowing what IVIG is, how to prepare and what the infusion process entails can make a big difference in helping you feel confident and ready. Following is some valuable information you need to know before starting IVIG therapy, including preparation tips, what to expect during your infusion, how to manage potential side effects and navigating financial considerations.

Understanding IVIG and Its Purpose

Made from plasma collected from thousands of healthy donors, IVIG strengthens the immune system by providing a wide range of antibodies, making it an important therapy for individuals whose immune responses are weakened or compromised. These antibodies work together to boost the body's defenses, offering protection against harmful bacteria, viruses and other invaders. The treatment is prescribed to manage primary immunodeficiencies, such as common variable immunodeficiency (CVID), to help patients combat infections their immune systems cannot handle on their own. Additionally, IVIG is prescribed for autoimmune diseases, such as such as myositis, polymyositis and chronic inflammatory demyelinating polyneuropathy (CIDP), to reduce the damaging effects of the immune system attacking the body's tissues, as well as to alleviate symptoms and prevent further complications.[2,3]

While IVIG therapy enhances the immune system and can decrease the frequency and severity of infections or autoimmune responses, it is important to remember that it does not cure the underlying condition. Instead, it helps

manage symptoms and improves the immune system's ability to respond to threats. For most patients, IVIG becomes part of a long-term treatment plan, requiring ongoing infusions to maintain its benefits.[4]

Preparing for Your First IVIG Infusion

If you are preparing for your first IVIG infusion, chances are you have already had many discussions with your doctors. However, there are a few steps *you* can take to ensure your first infusion goes as smoothly as possible:

• *Review your treatment plan.* Start by reviewing the details of your treatment plan with your healthcare provider. Usually, no special preparation is needed for IVIG therapy; you can eat, drink non-alcoholic and decaffeinated beverages and continue taking your regular medications. Still, it is a good idea to confirm with your provider if any changes to your routine are necessary before your infusion appointment. Your doctor may recommend some pre-medications to help with any potential side effects. In addition, your doctor may recommend blood tests or other evaluations to check your overall health before starting IVIG. These tests can establish a baseline and allow your medical team to monitor any changes during treatment.[2]

• *Understand insurance and financial considerations.* One of the first things you need to understand about IVIG is that it is expensive! This makes understanding your insurance policy an essential step before your first infusion. Start by confirming pre-approval or pre-authorization with your insurance provider to ensure the therapy is covered. If you skip this step, it could lead to unexpected expenses or delays in starting your treatment. Review your insurance plan carefully to determine what is covered and whether you have co-pays, deductibles or out-of-pocket costs. If the financial burden is overwhelming, investigate assistance programs offered by pharmaceutical companies or nonprofits that specialize in supporting IVIG patients.

If you are receiving your medication via a specialty pharmacy, it is important to work with that company to arrange delivery and handle billing details.[5] Additionally, the pharmacy can be a valuable resource to help you figure out the details of your treatment. By taking care of these steps ahead of time, you can focus on your treatment and feel less stressed about the costs and logistics.

• *Create a comfort plan.* When preparing for your IVIG infusion, bringing a few comfort items with you can make the experience much easier, since sessions often last several hours. Consider packing a blanket to stay cozy and snacks and water to stay nourished and hydrated during the treatment. Entertainment such as books, headphones for music or podcasts, or a tablet can help pass the time. Wear loose, comfortable clothing to allow easy access to your arm for the IV and to ensure you remain comfortable throughout the session. Staying well-hydrated before and during the appointment is especially important! It can reduce the risk of side effects such as headaches and make it easier for the nurse to start the IV without multiple sticks.[6] After the infusion, rest. You need to give your body time to recover and adjust to the treatment.

Proper post-infusion care can play a big role in reducing side effects and helping your body recover more quickly.

What to Expect During Your Infusion

Understanding the IVIG infusion process can help ease any uncertainty you might feel before your first treatment. A nurse will begin by reviewing your medical history, checking your vital signs and making sure you're comfortable. IVIG is administered in multiple settings, including supervised outpatient settings, such as clinics or infusion suites, and sometimes in hospitals if necessary. Frequently, infusions can be administered at home under the supervision of a healthcare professional such as a nurse. In some cases, you may start your therapy at home. Depending on your prescriber and your specific clinical situation, an outpatient setting may be preferred initially with transition to home infusion after you've had successful treatments in a supervised setting without any complications.[4]

The infusion starts with the nurse inserting a needle into a vein, typically in your arm. This is connected to an IV line that delivers the IVIG solution directly into your bloodstream. The nurse will begin the infusion at a slow rate to monitor your body's initial response and ensure you

Tips for Preparing for an IVIG Infusion

Hydrate: Drink water or other non-alcoholic or decaffeinated fluids on the days before, during and after your infusion to reduce the risk of headaches and other side effects. Your doctor may also pre-hydrate you with normal saline. Talk with your prescriber, pharmacist or nurse to determine what the right amount of fluid is for you.

Take medication: Ask your doctor or nurse if you should take your usual medications on the day of your infusion. You may also receive pre-medications such as acetaminophen, diphenhydramine or corticosteroids to prevent side effects.

Bring supplies: Bring a sweater, blanket or socks to keep you warm, and something to keep you occupied. The infusion can take many hours depending on your dose and how you tolerate the infusion.

Wear comfortable clothing: Wear clothing that's comfortable and makes it easy to access the IV site.

Plan for time off: You may need to take a day off work or school, and you may feel tired after the infusion.

Check your insurance: Make sure your infusion is covered by your insurance plan.

tolerate the treatment well. Gradually, the infusion rate may be increased, depending on your reaction and the protocol set by your healthcare provider. Infusions may be administered by gravity flow or with the assistance of an infusion pump.

The initial infusion is usually administered at the lowest concentration to assess your tolerance. If it is your first infusion, you can expect the process to take several hours,[2] depending on your required dose and how your body responds. The slow infusion rate reduces the risk of side effects, such as headaches, nausea or chills. During this time, the nurse will closely monitor your vital signs and watch for any signs of discomfort or adverse reactions. If you experience anything unusual, such as a headache or feeling unwell, let the nurse know immediately so adjustments can be made. Once the infusion is complete, the nurse will remove the IV (unless there is more than one day of therapy) and recheck your vital signs.

Possible Side Effects of IVIG

IVIG is widely used and generally well-tolerated, with a low risk of complications. In fact, experts estimate that fewer than five percent of people experience moderate or severe side effects.[2] Most reactions are mild and occur within the first 30 minutes to an hour of the infusion. These immediate side effects often subside when the infusion is stopped or slowed down and may include flushing, headache, malaise, fever, chills, fatigue and lethargy. While side effects are more common during your first infusion, they can also happen with subsequent treatments. This is why infusions are closely monitored by a healthcare professional who can adjust the treatment as needed to ensure your safety.

Serious side effects from IVIG are rare but can occur and require immediate medical attention. In some cases, an allergic reaction, such as anaphylaxis, may develop during or shortly after the infusion. Symptoms can include chest tightness, difficulty breathing, swelling of the tongue or face, a sudden drop in blood pressure or the appearance of a rash. If a reaction occurs during your IVIG infusion, the nurse will take immediate steps to manage it. This may include slowing down or stopping the infusion and administering medications such as Benadryl (diphenhydramine) or an EpiPen (epinephrine) to help control the reaction. The nurse will also notify your doctor about the situation and, if the symptoms do not improve, call for emergency medical assistance to ensure you receive appropriate care.[4]

Regardless of the severity, all side effects should be reported to your doctor. Serious symptoms such as chest tightness, difficulty breathing or swelling need immediate medical attention. However, even mild side effects are important to share with your healthcare provider. Reporting any reactions allows your doctor to adjust your treatment plan, such as changing the infusion rate or adding pre-medications, to make future infusions safer and more comfortable for you. Open communication with your doctor is key to ensuring the best possible treatment experience.[7]

Post-Infusion Care and Managing Side Effects

Getting ready for your first IVIG infusion doesn't end when the treatment is over. Taking care of yourself after the infusion is just as important. Proper post-infusion care can play a big role in reducing side effects and helping your body recover more quickly. Rest and hydration are key to your success! Staying adequately hydrated before, during and after your infusion can significantly reduce common side effects such as headaches and fatigue. The standard suggestion for hydration is eight eight-ounce glasses of water per day; however, additional clinical conditions must be considered to

avoid too much fluid. The suggested amount of water to drink per day is less in pediatric patients. The amount of hydrating fluids you consume should be discussed with your prescriber, nurse or pharmacist. If you experience mild symptoms like a headache or nausea, over-the-counter pain relievers such as acetaminophen or ibuprofen may help, but always check with your healthcare provider before taking any new medication. Resting in a quiet, comfortable space is another way to help your body recover effectively and reduce discomfort.

Building Support and Advocating for Yourself

Starting IVIG therapy can feel overwhelming, but it's important to remember that you're not alone. Building a strong support system and learning how to advocate for yourself can make a big difference. Connecting with others who understand your experience and ensuring your voice is heard can help you feel more empowered and supported throughout your treatment journey. Following are suggestions on how to get connected and take an active role in your care.

• *Self-advocacy for new patients.* The first step is taking an active role in your healthcare. Begin by educating yourself about your condition. This means remembering two things: First, knowledge is power, and second, there are no dumb questions. Learn everything you can about IVIG therapy, and never hesitate to ask your healthcare provider detailed questions about the infusion process, potential side effects and any other concerns you may have. Knowledge will boost your confidence and help you make informed decisions about your care.

Additionally, open communication with your healthcare team is a must! If you are uncomfortable with aspects of your treatment or they are not meeting your needs, such as infusion schedules, settings or how your body responds, voice your concerns. Sharing your preferences allows your providers to tailor the treatment to better suit you, ensuring a more comfortable and effective therapy experience.

• *Support networks and patient groups.* Connecting with others undergoing IVIG therapy can provide invaluable emotional support and practical insights. Patient support groups, available both in-person and online, offer platforms to share experiences, seek advice and find encouragement. Social media platforms also play a significant role in connecting patients, with many dedicated groups and pages where people can share their stories, ask questions and receive support. Additionally, there are resources specifically for IVIG patients, including educational materials, personal stories and forums where you can connect with others navigating similar challenges. These communities provide a safe space to learn, share and feel less isolated during your treatment journey.

Support networks can do more than help you feel less alone; they can provide helpful advice for managing IVIG therapy. They can provide tips on dealing with side effects and guidance on the infusion process. Connecting with others who understand what you're going through can make a big difference. It doesn't matter whether you join a support group, attend local meetups or connect with others online; these relationships can give you the confidence and support you need. Building these connections ensures you have people to turn to, so you don't have to face this journey alone.

Finding Strength Through IVIG: Preparing for a Healthier Journey

Starting IVIG therapy is a significant step, and being well-prepared can make a world of difference. Understanding the infusion process, building a support system and maintaining open communication with your healthcare team is key to better outcomes! With proper preparation, IVIG can be a powerful tool for managing health conditions and improving your quality of life.

The benefits of IVIG extend far beyond the treatment itself. By using available resources, connecting with support networks and advocating for your needs, you can take control of your care and navigate your healthcare journey. Remember, you're not alone. There are tools, communities and healthcare professionals ready to support you every step of the way. With the right approach, IVIG therapy can be a manageable and beneficial part of your health journey.

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

Managing Difficult Conversations: *Telling Others About Your Diagnosis*

Choosing who and how to inform others about your health is a deeply personal decision, but planning ahead can help you handle it with grace.

By Amy Scanlin, MS

KNOWING WHO to share a devastating health diagnosis with can be as challenging as knowing when to share it. For some patients, the retelling of painful information can reignite the trauma felt when they first heard their diagnosis. Though deciding who to share with, as well as how and when to share, is as personal as your own health journey, here are some thoughts that may make the process a little bit easier.

Deciding Who to Tell

Receiving a diagnosis brings feelings of fear, confusion and often a sense of urgency to reach out to others for emotional support. Deciding who to confide in is often among a patient's first worries.

"There comes a time when people ask themselves, 'Whom should I tell?'" says Troy Fontenot, MSW, a social worker based in the Washington, D.C., area. "But, I think an additional question should be, 'Why am I telling this person? What am I hoping to receive in doing so?'" Answering those questions can help to manage expectations on both sides.

According to Fontenot, sharing provides an opportunity to connect and understand, to speak and be heard, and it can help bring a universal commonality of shared experiences. Sharing can also open the door to more intimate conversations about a diagnosis, about what it means and about how each party is feeling. "Sharing can let people know that it's OK to talk about it, to move past stigma," he says.

Choosing who to share with (and *how* to share) is likely to be a very different decision depending on the nature of your relationship. Life partners and others in your inner circle may be more trusted entities than those on the periphery such as co-workers or connections on social media. A feeling of trust will dictate how much information you are comfortable sharing. Emotional maturity is also an

important consideration. Young children, for instance, will comprehend differently than teens, and teens will internalize differently than adults.

Creating a safe space in which to engage in these important conversations is about knowing your audience so that expectations can be managed. "Ask yourself, 'Will you feel safe sharing with this person? Do you have an expectation as to what reaction you are hoping to receive based on your relationship?'" says Fontenot.

There is no need to rush to snap decisions when deciding to share a diagnosis. The most important thing in the early post-diagnosis days is for you to sit with the news and come to terms with what you are facing so that when and if the conversations come, you are ready.

A feeling of trust will dictate how much information you are comfortable sharing.

Close and Connected: Life Partners, Family and Friends

In most cases, patients will share their diagnosis with their life partner and those they are closest to early on. And, for most, that's a good thing because it will enable much-needed emotional and logistical support. A diagnosis doesn't just affect the patient, it will affect many others, and will affect some people profoundly.

No person knows at the outset of a diagnosis what the future holds. But, by allowing trusted others in, the shared experience of what comes next can be faced together.

Comfort and Reassurance: Children

How information should be shared with children will differ given their age and maturity level. Regardless, focus on comfort and reassurance when speaking with children. They are attuned to those close to them and will notice when things are different. Neurologic and physical changes may be scary without context. Some kids may wonder what they have done wrong if someone they trust and spend time with is suddenly less available. In the absence of information, they may blame themselves when they don't understand what's going on.

Sharing a diagnosis with young children should be as simple, straightforward and positive as possible. It is important to be honest but also to not overshare before the child is mature enough to understand. Regardless of how much is shared with young children, Fontenot encourages creating a supportive environment where no topic is off limits. "It is important for kids to know that there is nothing we can't talk about," explains Fontenot. "As adults, we can answer as much or as little as we want, but we should always create a safe space where kids feel comfortable speaking."

Another consideration when sharing with children is the possibility that the diagnosis has a genetic or behavioral component that can directly affect them both now and with future decisions. Whether the child is young and the parent chooses to seek genetic testing on the child's behalf, or the child is older and can make decisions of his or her own, sharing the right information at the right time can help navigate the repercussions of the diagnosis in the child's life.

Broader Outreach: Social Platforms

Though many find invaluable support through social media platforms, think before broadcasting personal information widely, particularly because of the potential for losing control over how far that information will travel. Also, be aware that well-meaning connections may choose to share your personal information that you intended to be private. Make your expectations for further sharing clear, and be aware that unsolicited comments, advice and even scams or other kinds of exploitation are possible as the information travels in the digital space.

Strategies for How to Share

The amount of information you choose to share is deeply personal and will be different than other individuals. Before sharing, decide how much detail to reveal and how you will communicate the message. "Facial and body language cues are an important part of communication," says Fontenot. He says whenever possible, particularly with close relationships, sharing information in person will help each party to communicate most effectively. When in-person conversations are not possible, video communications are the next best thing.

But, Fontenot also notes that when communicating to your extended network, a phone call or text message may also be

Reconnect with game night

People with primary immunodeficiency (PI) who infuse CUVITRU weekly or every other week may be able to experience more of these moments.

What is CUVITRU®?

CUVITRU [Immune Globulin Subcutaneous (Human)] 20% Solution is a ready-to-use liquid medicine that is given under the skin (subcutaneously) to treat primary immunodeficiency (PI) in people 2 years and older.

IMPORTANT SAFETY INFORMATION

What is the most important information I need to know about CUVITRU?

CUVITRU can cause the following serious reactions:

- Severe allergic reactions causing difficulty in breathing or skin rashes
- Decreased kidney function or kidney failure
- Blood clots in the heart, brain, lungs, or elsewhere in the body
- Severe headache, drowsiness, fever, painful eye movements, or nausea and vomiting
- Dark colored urine, swelling, fatigue, or difficulty breathing

Who should not use CUVITRU?

Do not use CUVITRU if you:

- Have had a severe allergic reaction to immune globulin or other blood products.
- Have a condition called selective (or severe) immunoglobulin A (IgA) deficiency.

What should I avoid while taking CUVITRU?

- CUVITRU can make vaccines (like measles/mumps/rubella or chickenpox vaccines) not work as well for you. Before you get any vaccines, tell your healthcare provider (HCP) that you take CUVITRU.
- Tell your HCP if you are pregnant, or plan to become pregnant, or if you are nursing.

What are the possible or reasonably likely side effects of CUVITRU?

CUVITRU can cause serious side effects. If any of the following problems occur after starting CUVITRU, stop the infusion immediately and contact your HCP or call emergency services:

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of irritation and swelling of the lining around your brain.
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain, swelling, warmth, redness, or a lump in your legs or arms. These could be signs of a blood clot.

Cuvitru
[Immune Globulin Subcutaneous (Human)] 20%

Proven protection from infection

In the North American (NA) study, there were 0.012 acute serious bacterial infections (ASBIs) per patient-year.*† This exceeds the FDA standard for effectiveness, which is one serious ASBI per year.

Nearly all infusions (99.8%) were completed without reduction, interruption or discontinuation due to tolerability

No patients discontinued due to local adverse reactions (ARs) and 0 serious ARs related to CUVITRU were reported.

The most common adverse reactions observed in clinical trials in ≥5% of patients were: local adverse reactions including mild or moderate pain, erythema, and pruritus, and systemic adverse reactions including headache, nausea, fatigue, diarrhea, and vomiting.

Flexible administration that can be tailored to fit your lifestyle‡§

CUVITRU can be infused at the fastest rates and highest volumes with the fewest infusion sites of any subQ IG.§

In the NA clinical study, CUVITRU was studied in 77 people with PI ≥2 years of age. The main goal of the study was to measure how many acute serious bacterial infections (ASBIs) were experienced over the course of 1 year. ASBIs are short-term but serious infections that require immediate medical care. ASBIs were evaluated in 74 people taking CUVITRU for an average of 380.5 days (range, 30-629 days).

*One ASBI that occurred during the study was a case of pneumonia in a 78-year-old person.

†A patient-year is a patient experience in a clinical trial over the course of 1 year. One patient-year is equal to, for example, the experience of 2 patients for 6 months, or 12 patients for 1 month each.

‡In the NA study, the average infusion time was 0.95 hours (range 0.2-6.4 hours) and most (84.9%) used 1 to 2 needlesticks.

§You'll infuse your first 2 infusions at 10 to 20 mL/hr/site. After that, you'll be able to increase your rate up to 60 mL/hr/site as tolerated. Infuse at up to 4 sites simultaneously.

SubQ IG=subcutaneous immune globulin.

IMPORTANT SAFETY INFORMATION (continued)

- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.
- Chest pain or trouble breathing, or blue lips or extremities. These could be signs of a serious heart or lung problem.
- Fever over 100°F. This could be sign of an infection.

The following one or more possible side effects may occur at the site of infusion. These generally go away within a few hours, and are less likely after the first few infusions.

- Mild or moderate pain
- Redness
- Itching

The most common side effects that may occur are:

- Headache
- Nausea
- Fatigue
- Diarrhea
- Vomiting

These are not all the possible side effects. Talk to your HCP about any side effect that bothers you or that does not go away.

Please see Important Facts about CUVITRU on the following page.

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.

Register for a community educational program created for people living with PI—*My Life, My Story.*

Learn more about what it's like to infuse CUVITRU from clinical nurse educators and other patients like you.

Takeda

IMPORTANT FACTS about CUVITRU (CUE-vih-troo) [Immune Globulin Subcutaneous (Human)] 20% Solution

What is the most important information I need to know about CUVITRU?

CUVITRU can cause the following serious reactions:

- Severe allergic reactions causing difficulty in breathing or skin rashes
- Decreased kidney function or kidney failure
- Blood clots in the heart, brain, lungs, or elsewhere in the body
- Severe headache, drowsiness, fever, painful eye movements, or nausea and vomiting
- Dark colored urine, swelling, fatigue, or difficulty breathing

What is CUVITRU?

CUVITRU is a ready-to-use liquid medicine that contains immunoglobulin G (IgG) antibodies, which protect the body against infection. CUVITRU is used to treat patients with primary immunodeficiency diseases (PI).

There are many forms of PI. The most common types of PI result in an inability to make a very important type of protein called antibodies, which help the body fight off infections from bacteria or viruses. CUVITRU is made from human plasma that is donated by healthy people. CUVITRU contains antibodies collected from these healthy people that replace the missing antibodies in PI patients.

Who should not use CUVITRU?

Do not use CUVITRU if you have a known history of a severe allergic reaction to immune globulin or other blood products. If you have such a history, discuss this with your healthcare provider (HCP) to determine if CUVITRU can be given to you. Tell your HCP if you have a condition called selective (or severe) immunoglobulin A (IgA) deficiency.

How should I use CUVITRU?

CUVITRU is given under the skin (subcutaneously). Most of the time, infusions under the skin are given at home by self-infusion or by caregivers. Instructions for giving CUVITRU under the skin (subcutaneously) are provided in the FDA-approved patient labeling (Information for Patients and Instructions for Use). Only use CUVITRU by yourself after you have been instructed by your HCP.

What should I avoid while taking CUVITRU?

CUVITRU can make vaccines (like measles/mumps/rubella or chickenpox vaccines) not work as well for you. Before you get any vaccines, tell your HCP that you take CUVITRU.

Tell your HCP if you are pregnant, or plan to become pregnant, or if you are nursing.

What are the possible or reasonably likely side effects of CUVITRU?

The following are one or more possible reactions that may occur at the site of infusion. These generally go away within a few hours, and are less likely after the first few infusions.

- Mild or moderate pain
- Redness
- Itching

The most common side effects of CUVITRU are headache, nausea, fatigue, diarrhea, and vomiting.

If any of the following problems occur after starting treatment with CUVITRU, stop the infusion immediately and contact your HCP or call emergency services. These could be signs of a serious problem.

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of irritation of the lining around your brain.
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain, swelling, warmth, redness, or a lump in your legs or arms. These could be signs of a blood clot.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.
- Chest pain or trouble breathing, or blue lips or extremities. These could be signs of a serious heart or lung problem.
- Fever over 100°F. This could be a sign of an infection.

These are not all the possible side effects. You can ask your HCP for a physician's information leaflet. Tell your HCP about any side effect that bothers you or that does not go away.

Whenever giving yourself treatments at home, you should have another responsible person present to help treat side effects or get help if you have a serious adverse reaction occur. Ask your HCP whether you should have rescue medications, such as antihistamines or epinephrine.

How do I store CUVITRU?

Store CUVITRU refrigerated or at room temperature.

- You can store CUVITRU in the refrigerator (36°F to 46°F [2°C to 8°C]) for up to 36 months or
- You can store CUVITRU at room temperature (up to 77°F [25°C]) for up to 24 months.
- Do not return CUVITRU to the refrigerator if you take it out to room temperature.
- Do not freeze.
- Do not shake.
- Check the expiration date on the carton and vial label. Do not use CUVITRU after the expiration date.
- Protect from light. You can use the original CUVITRU containers to protect it from light.

How do I get more information about CUVITRU?

The risk information provided here is not comprehensive. To learn more, talk about CUVITRU with your HCP or pharmacist. The FDA-approved Full Prescribing Information, including Information for Patients, can be found at www.CUVITRU.com or by calling 1-877-TAKEDA7 (1-877-825-3327).

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perfectly appropriate, allowing you to share any information you wish without directly engaging. There is no one right answer.

Prior to having the conversation, think through what the messaging will be. For example, if the aim of your message is just to make someone aware, sometimes saying "I have some information that is difficult for me to share. I don't need any advice, I just need you to listen," is direct and tells the recipient what you need at that time.

However, adds Fontenot, we live in a culturally independent world, and asking for help is often as therapeutic for the patient as it is for those being asked, particularly when the patient's energies should be elsewhere. Have some things in mind for when the "What can I do?" questions come so that there is a ready list of ideas that can ease unnecessary burdens, things such as walking the dog, bringing a weekly meal, organizing friends for a visit or taking the kids to school occasionally. "Think of things that will make part of your life easier, a little less worrisome," he says. People want to help and often just don't know what kind of help is most needed.

Timing Your Message and Invaluable Support

Along with the decision of who is the question of when. Though the decision to share doesn't have to be made right away, for many, sharing with close family and friends can be incredibly therapeutic.

Allowing trusted people in relatively early can be a good thing. Despite the worry that the news of illness will be a burden or make you feel stigmatized or treated differently, the opposite is almost always the case. Even so, the timing of sharing is personal, and again, there is no single best answer.

You should also be aware that in some cases, the people with whom you choose *not* to share may feel hurt or excluded, and you may find yourself feeling guilty for not being forthcoming. If you are a patient and that is the case: *Stop*. No one should feel emotionally burdened by carrying other people through their own diagnosis and treatment.

When Their Reaction Isn't What You Need

Sharing is tricky, particularly when patients need to share with people whose reactions may be more challenging. Whether overly attentive, pestering with questions or offering unsolicited advice, well-meaning people may unintentionally intrude. In other cases, the opposite is true. The people closest to you may back away, unsure of what to say or afraid of saying the wrong thing.

While avoidance of another person's reaction should not be a primary driver for deciding whether or not to share your diagnosis, preparation is helpful so his or her reaction doesn't catch you off guard. Having responses ready for any hurtful comments is helpful, too. Your diagnosis is not about that person.

Prepare yourself for the conversation by remembering why you made the decision to share in the first place and what your expectations are. Some people have less empathy, others more. Hopefully, says Fontenot, everyone has people in their lives who will react in a supportive way. But, when reactions fall short, instead of responding with anger, try forgiveness and understanding.

Everyone is coping with their own life, reminds Fontenot, and every action is a coping strategy. For instance, hearing about your diagnosis may bring up unresolved trauma in other people, and perhaps "their only way of coping, at least in the moment, may be to back away." That can be very difficult for you because you may feel abandoned. But remember, just because someone doesn't respond in the way you had hoped, that doesn't mean they don't greatly care about you. "We can't force people to show up in the way we need them to," says Fontenot. "Have grace and patience, with yourself and with those around you."

And, if someone's reactions seem completely out of character, it may be an opportunity to find out what is going on with that person instead of feeling hurt or judged. When each party understands where the other is coming from they can possibly find common ground for support. Certainly, most people want to be helpful, not hurtful. But, they may not realize how what they are saying, or not saying, is being received.

Managing Expectations

It is important for you to be prepared to have conversations with anyone with whom you've shared your diagnosis. "Ask yourself if you are ready to talk about this with this person," says Fontenot. If the answer is no, it may not be the right time or you may need to establish clear communication boundaries before doing so.

Sharing a diagnosis isn't easy and many will find the information hard to hear and discuss. But, a little consideration and empathy on both sides can be helpful in managing expectations. "Sometimes all we need to hear is 'I care about you. Thank you for telling me,'" says Fontenot.

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

How to Handle Medical Setbacks

Many strategies can help you cope when your health takes a downturn, but the key is reacting to it in a proactive manner to help you get through it.

By Rachel Maier, MS

ONE STEP forward, two steps back. You make progress toward better health and — bam! — something pops up and you've got to readjust. Sometimes medical setbacks are completely unexpected, making an already hard health journey even more difficult. Other times, medical setbacks aren't surprising so much as they are discouraging: You knew this day would come, but you were hoping you still had more time before it did. Your health goes up and down and round and round, following the rhythm of life circumstances. It's mentally, physically and emotionally exhausting.

When you live with a chronic illness, your feet start to expect to walk this familiar, frustrating pace. You probably know what steps to take to address practical problems and logistical ramifications, such as slowing down, giving your body what it needs and asking for help while you wait to bounce back. But medical setbacks can take a mental toll, too, and learning how to prioritize your mental health is just as important as dealing with the physical aspects of your illness. Learning how to handle all of it well is a dance many of us struggle with, but there are strategies we can learn that can help us do better.

Resilience

When things go wrong, do you typically feel like a victim (e.g., Why is this happening to *me*?) or turn to coping mechanisms that do more harm than good (such as binge eating, drug or alcohol use or other risky behaviors)? It's natural to feel singled out when bad things happen to you, and it's common to want to self-soothe in ways that make you feel better, even if it's temporary. But these behaviors are not healthy, and neither

strategy will help you deal with your setback in any meaningful way. That's why developing resilience — the ability to adapt to difficult or challenging life experiences, especially through mental, emotional and behavioral flexibility and adjust to external and internal demands — is so important.[1] When you're resilient, you can see past your problems and adapt your life accordingly. Resilience helps you deal.

Resilience isn't something you're born with; it's something you build over time. It does not involve getting rid of the stress in your life; instead, it is about identifying and using your strengths as you navigate through stressful situations. Lisa Wexler, a professor of social work and research professor in the Research Center for Group Dynamics at the Institute of Social Research at the University of Michigan, explains that your strengths are protective factors that defend against stress and/or directly promote well-being.[2] Examples of these strengths are your community and the ceremonies and cultural practices you engage in that help you cope. Other protective factors include a healthy body, expressing your feelings, looking at problems from different angles and leaning on a support system. The key, according to Cindy Bergemen, PhD, a psychology professor at the University of Notre Dame, is asking yourself how your needs match with the tools you have available.[2]

Healthy Ways to Cope

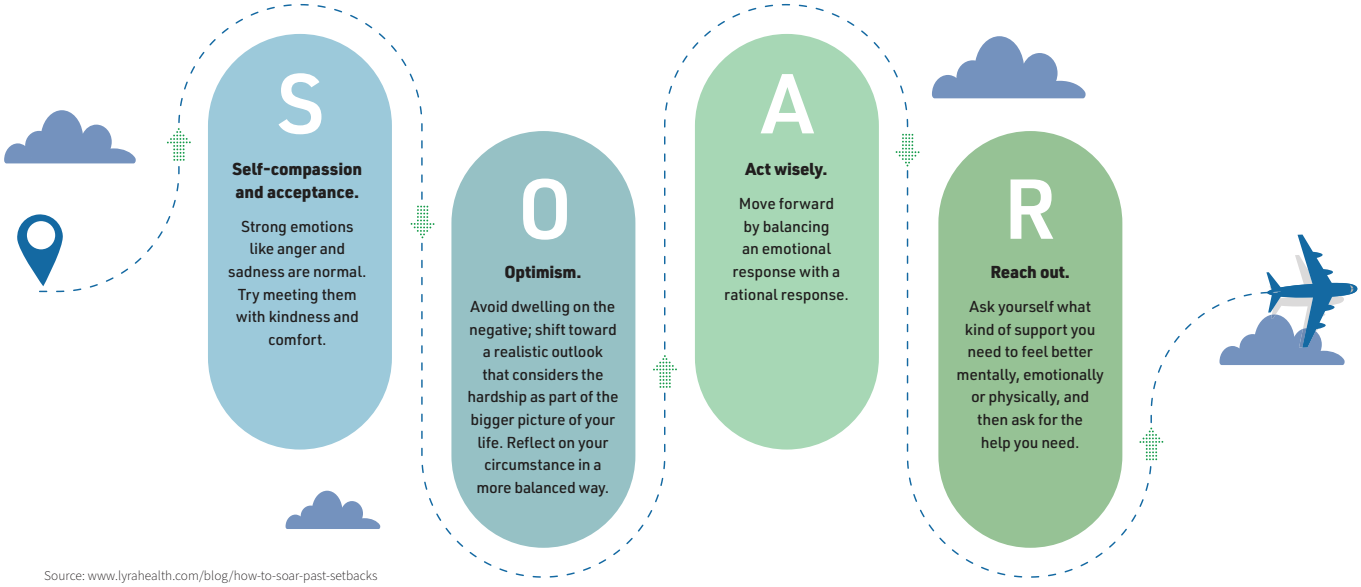
To proactively build healthy coping mechanisms, betterhelp.com, the world's largest online provider of professional and personalized therapy, suggests doing the following when an unforeseen situation sets you back:[3]

Take a breather. You might be tempted to problem-solve and fix the issue immediately, but you might benefit from setting the problem aside to take a deep breath first. In fact, this is one of the best things you can do for yourself when you experience a setback of any kind. According to betterhelp, "Taking a break can be one of the best coping skills at times. The willingness to take a break doesn't necessarily mean giving up [...] It may simply mean clearing your head, regrouping and later returning to the situation with a fresh mindset and potentially new ideas."[3]

Look for a way to learn. Every setback brings an opportunity to learn. Look at problems from different angles, and think of challenging situations as opportunities to grow. Analyzing what went wrong and asking yourself what lesson you can take away from the situation can help prepare you for future setbacks. It will shift you from a reactive response to a proactive response, meaning you'll start anticipating problems before they happen and come up with potential

How to "SOAR" Past Setbacks

Building resilience boosts your ability to adapt. Use this four-step process to rise above challenges.



Source: www.lyrahealth.com/blog/how-to-soar-past-setbacks

solutions ahead of time instead of responding to adversity with raw, rash emotions.

Find a different path forward. There isn't always one "right" way to move ahead. Often, there is more than one treatment plan, medicine or even medical provider to consider. If the one you were using isn't working anymore, it's time to think about a new strategy. Talk to trusted doctors and patients who have walked the same road. Discuss your options with them and with loved ones. Seek advice and consider sound wisdom, and then try a new approach.

Lean on loved ones. Surround yourself with people who care about you. Having someone to talk to goes a long way when processing the emotional ups and downs of medical setbacks. Plus, they love you, so they will help you carry the weight of your hardship. When others offer to help, accept their offer. Arm yourself with a list of practical things you can ask for before a medical setback happens so you know what to say when they ask you how they can help. Some examples are mowing the lawn, running an errand, shuttling kids to and from school, picking up some soup from your favorite deli or a weekly phone conversation where you can vent your feelings and frustrations without judgment.

Avoid negative thinking. When you're caught in a negative thought, it's really hard to get out of it. Avoiding negative thinking doesn't mean pretending everything is OK when it's not; it means being honest about the bad stuff while also acknowledging the good stuff. Choosing to shift your mindset away from negativity and toward the positive things in your life promotes positive thinking instead.

Practice resilience and perseverance. It's one thing to know about resilience and another to act on what you know. Maybe you understand that resilience means being able to adapt to hardships life throws at you, but you have a hard time actually doing that. Developing resilience is a process, much like building up muscle. It takes time and intentional effort. According to the American Association of Psychology, resilience can be built by focusing on four core components: connection, wellness, healthy thinking and meaning. Together, they can empower you to learn from difficult experiences.[4] Connect with others — don't withdraw. Tend to your personal needs: Eat healthy food, move your body, rest, pray. Process your feelings and do things that bring you joy. Practice positive self-talk: Be gentle and encouraging; evaluate negative thoughts and respond in a positive way. Focus on the bigger picture: You still have purpose even though you are in pain. Remember the things that make you who you are and bring meaning to your life.

Your Reaction Is Key

Everyone experiences setbacks, but a medical setback feels more personal and painful than other kinds of setbacks, especially when you have a chronic illness. You may wonder what you did wrong, how you could have prevented it, what other people think about you, whether you "deserve" it or if you'll ever be yourself again. Common responses to serious medical setbacks include anger or frustration, fear of death, worry about the future, grief over the loss of health, feeling powerless or hopeless, regret or guilt, denial, isolation and loss of self.[5]

What separates people who handle medical setbacks well from people who handle them poorly is their response. "It's usually not the setback itself, but instead your reaction to it, that can negatively impact your mental health."[3] When you spend mental energy wondering what others think, you end up being your very own roadblock. You cannot control what other people think about you, your situation or the way you choose to move forward. You *can* control what *you* think about it.

A Setback, Not a Stalemate

Medical setbacks aren't easy. They are discouraging on so many levels, affecting physical, emotional and mental health. Even the most resilient people experience a wide range of emotions and burnout when setbacks happen; it's completely normal. But what keeps resilient people going anyway is simple: They remember a setback isn't a stalemate and they approach their circumstance with an open mind and a positive outlook.

You can do that too. Plan ahead for a setback before it happens. Then, when it does, slow down, treat your body kindly, lean on loved ones, accept help and be willing to pivot your plans. It will help guard against bitterness and resentment and will give you the tools you need to get through the hard season. IG

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RACHEL MAIER, MS, is the associate editor of *IG Living* magazine.

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What Are Alternative Funding Programs?

The director of advocacy at the Infusion Access Foundation breaks down why these alleged cost-cutting programs are deceptive and harmful to patients.

By Kindyl Boyer, MPP

AT THE INFUSION Access Foundation, we often relate our fight for patient access to the game of Whack-A-Mole — an arcade game where the player repeatedly "whacks" a mole that pops up randomly through one of five holes. But instead of walking away from the game with mild amusement or annoyance, our patient community is left exhausted and on edge when waiting for the next insurance or pharmacy benefit manager "mole" to pop out.

One example happened in November of 2022, when a pharmaceutical wholesaler approached our advocacy department with a curious issue: An insurance company in Alabama had begun sending vaguely worded letters to infusion patients. The recipients of these letters were mostly beneficiaries of a group health plan that served current and retired public education employees. The letters stated that all recipients were to register with a third-party company — we'll call it Alt-X — to receive coverage for their infusion medications. The letter explained that the health plan was implementing a new program run by Alt-X that would provide 100 percent coverage for the infusion medications the plan had covered in years' past.

After receiving the letter, most patients went to their infusion centers and physician offices, letter in hand, asking for clarity and registration assistance. Providers attempted to help their confused patients by contacting Alt-X but were unsuccessful. Alt-X insisted that the patients themselves must contact them to complete registration. Most infusion centers and physician offices found this bizarre, since they had previously worked with co-pay assistance programs that allowed them to facilitate registration on patients' behalf.

Once the initial confusion subsided, patients were able to enroll in Alt-X's program, and the insurance company began processing claims, seemingly like normal. But what patients didn't realize was that their insurer had taken their infusion medications completely off of their list of covered drugs, and the patient was now at the mercy of Alt-X's ability to source manufacturer patient assistance to pay for the medications. Alt-X was using financial assistance meant for patients who are actually uninsured to pay for the medications of patients who were beneficiaries of this health plan.

But from the patient perspective, it didn't seem like anything was amiss. For these patients and the many others

who have enrolled in programs similar to Alt-X's, things seem to run smoothly for a while — that is, until one of two things happens: 1) the manufacturer patient assistance runs out, and the is patient no closer to his or her deductible or out-of-pocket maximum, or 2) the manufacturer figures out that this particular individual *is* insured and is *not* the proper beneficiary of patient assistance funds, thus requiring the patient's prescription to be sent back to the patient's health plan for coverage.[1]

After talking with the wholesaler about this problem back in 2022 and then talking to others in the patient advocacy space, we realized a new "mole" had popped up — alternative funding programs.

A Deceptive Cost-Cutting Practice

Alternative funding programs market themselves as healthcare cost management companies or even patient advocacy organizations. However, these programs are not affiliated with insurance companies, employers or pharmaceutical companies. Rather, they are third-party vendors who have been hired by an employer-sponsored health plan under the guise of cutting costs. The company managing the alternative funding program works with the employer-sponsored health plan to exclude specialty medications from coverage by defining them as "non-essential health benefits" (non-EHBs) in an attempt to make their beneficiaries appear uninsured or underinsured. [Notably, EHBs are the 10 categories of services health insurers must cover under the Affordable Care Act.[2]] The alternative funding program then sources funds from patient assistance programs (PAPs) to cover the cost of care for their beneficiaries.[3]

Alternative funding programs take advantage of PAP funds to cover the cost of specialty drugs.

However, PAPs are typically run by charitable foundations, often funded by pharmaceutical companies, and serve as financial assistance programs for low-income individuals.[4] Typically, pharmaceutical companies will designate these programs for uninsured or underinsured individuals. Underinsured refers to individuals who have insurance, but their healthcare costs make up a substantial percentage of their household income.[5] Alternative funding programs take advantage of PAP funds to cover the cost of specialty drugs that an employer-sponsored health plan has intentionally removed from coverage. Because the specialty medications are now defined as non-EHB, patients must enroll in the alternative funding program or be responsible for 100 percent of their medication's cost. Furthermore, any amount paid for the medication by or on behalf of beneficiaries will not count toward their deductible or out-of-pocket cost responsibility due to the medication's non-EHB status.

If patients need one of these uncovered drugs, their request is first denied because their plan technically does not cover the drug; then, the third-party vendor will source financial coverage from a PAP, which in turn allows the patients' employer health plan to avoid paying for the medication the patients need.

With this process, patients experience denial of treatment, share their medical situation and personal information with an external third-party, and wait to see if an alternative funding source is available. If alternative funding isn't available, or a PAP deems a patient ineligible for its program, the patient's prescription is sent back to the health plan for coverage under the pharmacy benefit. In some cases, third-party vendors have been known to import drugs from outside the United States. However, the U.S. Food and Drug Administration (FDA) prohibits the importation of fraudulent prescription and nonprescription drugs and medical devices to protect patient safety.[6] At the beginning of 2023, the nonprofit health policy organization Aimed Alliance sent a letter to the FDA Office of Regulatory Affairs and Office of Importation Operations alerting the agency that alternative funding programs are illegally importing prescription drugs.[7] FDA indicated it will look into the matter and take action against those that import illegal products.

Misuse of PAP's Purpose

The sentiment from drug companies we have spoken with is that PAPs were not designed to help employer plans save a few bucks; they were created to help individuals without

insurance access the medications they need. In a STAT News article, journalist Ed Silverman puts it plainly: "Plan sponsors — usually employers that fund their own health coverage — no longer have to pay for medicines. Instead, the pharmaceutical company bears the cost."[8] In May of 2023, AbbVie, the pharmaceutical company responsible for SKYRIZI, RINVOQ and HUMIRA, sued a company known as Payer Matrix. In the lawsuit, AbbVie alleges that Payer Matrix "operates a fraudulent and deceptive scheme to enrich itself by exploiting AbbVie's PAP through the enrollment of insured patients into a charitable program not intended for them."[9] The complaint goes on to say that AbbVie's PAP, myAbbVie Assist Assistance Program, is only for uninsured and underinsured patients, and that Payer Matrix's practices are intended to exploit this eligibility requirement. Specifically, it states, "Payer Matrix knowingly maneuvers ineligible patients into AbbVie's PAP — specifically, insured patients who should be receiving their medicine through their employers' health insurance plan […] then charges the patients' employers a substantial fee for reducing the employers' health insurance costs through its scam."[10]

Since its initial filing, AbbVie has updated its complaint to include four new claims under federal law, including a Racketeering Influenced and Corrupt Organization Act (RICO) claim, two claims under the Lanham Act and a common law fraud claim. The Infusion Access Foundation, along with 23 other patient advocacy groups, signed on to Aimed Alliance's amicus brief in support of AbbVie's motion and described additional harmful impacts these programs have on patients.[10]

Why It Matters

The Infusion Access Foundation's concern is two-fold. First, denying coverage to vital drugs for premium-paying beneficiaries who deal with chronic conditions is unconscionable. Not only is the plan revoking coverage and placing the care of their beneficiaries in the hands of an alternative funding program, but the inevitable delays associated with enrollment is unacceptable for individuals battling a chronic illness.[11]

Second, the pool of alternative funding will inevitably grow smaller as insured patients enter into competition with uninsured patients for already scarce funding. Not to mention, pharmaceutical companies may even consider ending any and all PAPs if the trend continues. A 2023 report from Pharmaceutical Strategies Group titled "Trends in Specialty Benefit Design" demonstrates the growing prevalence of alternative funding programs by analyzing responses from 104 employers and 45 health plans. The previous year, the report looked at 114 employers and 30 health plans. From 2021 to 2022, the share of employers who use an alternative funding program increased from six percent to 14 percent, with large employers (those with greater than 10,000 beneficiaries) more likely to utilize an alternative funding program.[12]

An Uncertain Future

It's difficult to predict whether this latest "mole" is here to stay or if the outcome of a certain lawsuit will extinguish the industry altogether. For chronic illness patients who need access to specialty medications, it will be close to impossible to avoid engaging with one of these companies should their health plan choose to work with them. However, awareness of how the system works and who the players are can, at the very least, provide beneficiaries, consumers, patients and the public with the knowledge they need to ask questions and make informed decisions.

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KINDYL BOYER, MPP, is director of advocacy for the Infusion Access Foundation, where she enhances the foundation's advocacy efforts at both the state and federal levels. She actively participates in various coalitions, representing the organization's interests and amplifying its impact in the healthcare community. Kindyl is dedicated to creating and expanding educational materials for patients receiving provider-administered medications and empowering patients in their navigation of the healthcare system.

If you're an adult living with primary immunodeficiency (PI), ALYGLO™ can reduce the risk of infection from PI and its impact on your daily life.[1]
Based on a clinical study of 33 adults ages 17-70 in North America.[1]

0.03 SERIOUS INFECTIONS per patient year[1]

0.2 DAYS OF HOSPITALIZATION per patient year[1]

6 DAYS MISSED OF WORK OR SCHOOL per year[1]

INDICATION

ALYGLO™ is indicated for the treatment of primary humoral immunodeficiency (PI) in adults aged 17 years and older. This includes, but is not limited to, congenital agammaglobulinemia, common variable immunodeficiency (CVID), Wiskott-Aldrich syndrome, and severe combined immunodeficiencies.

IMPORTANT SAFETY INFORMATION

- **Thrombosis (blood clot formation) can happen with ALYGLO. Factors that increase this risk include advanced age, prolonged immobility, certain medical conditions, and cardiovascular risk factors.**
- **ALYGLO may affect the kidneys. In some cases, it can lead to acute renal failure or death.**
- **If you're at risk for blood clots or kidney problems, your doctor should give you ALYGLO at the lowest effective dose and infusion rate. Staying well-hydrated before treatment is essential.**
- ALYGLO is not suitable for people who have had severe allergic reactions to immune globulin or those with IgA deficiency and a history of hypersensitivity.
- If you experience any signs of hypersensitivity during the infusion, treatment should be stopped and epinephrine (an emergency medication) should be administered immediately.
- ALYGLO may cause hyperproteinemia, increased serum viscosity, and hyponatremia (low sodium levels).
- Aseptic Meningitis Syndrome (AMS) is a rare condition that can occur after receiving ALYGLO, especially with high doses or rapid infusion. Symptoms usually start within a few hours to 2 days after treatment. If AMS occurs, stopping ALYGLO usually leads to improvement within several days without lasting effects.
- Hemolysis, a breakdown of red blood cells, may occur. Some patients may experience delayed hemolytic anemia due to increased sequestration of red blood cells. Severe hemolysis-related kidney dysfunction or disseminated intravascular coagulation has been reported.
- Transfusion-Related Acute Lung Injury (TRALI) is a rare complication characterized by severe respiratory distress, pulmonary edema, and fever. Patients with TRALI may need oxygen therapy and ventilator support.
- ALYGLO is made from human blood, which may carry a risk of transmitting infectious agents (such as viruses).
- After receiving ALYGLO, some antibodies from the treatment may temporarily show up in blood tests. This could lead to misleading results, so your healthcare provider will consider this when interpreting lab results.
- Common side effects include headache, nausea/vomiting, fatigue, nasal/sinus congestion, rash, arthralgia, diarrhea, muscle pain/aches, infusion site pain/swelling, abdominal pain/discomfort, cough, and dizziness.

Reference: 1. ALYGLO Prescribing Information. GC Biopharma; 2023.

For more information about ALYGLO, talk to your doctor and see Brief Summary of Prescribing Information on next page.

GC Biopharma

Alyglo™
immune globulin intravenous, human-stwk 10% liquid

BRIEF SUMMARY OF PRESCRIBING INFORMATION
Please see full Prescribing Information at ALYGLO.com.

WARNING: THROMBOSIS, RENAL DYSFUNCTION and ACUTE RENAL FAILURE

See full prescribing information for complete boxed warning.

- **Thrombosis may occur with immune globulin intravenous (IGIV) products, including ALYGLO. 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.**
- **Renal dysfunction, acute renal failure, osmotic nephropathy, and death may occur with the administration of IGIV products in predisposed patients.**
- **Renal dysfunction and acute renal failure occur more commonly in patients receiving IGIV products containing sucrose. ALYGLO does not contain sucrose.**
- **For patients at risk of thrombosis, renal dysfunction or renal failure, administer ALYGLO 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

ALYGLO is a 10% immune globulin liquid for intravenous injection, indicated for the treatment of primary humoral immunodeficiency (PI) in adults. This includes, but is not limited to, the humoral immune defect in congenital agammaglobulinemia, common variable immunodeficiency (CVID), X-linked agammaglobulinemia, Wiskot-Aldrich syndrome, and severe combined immunodeficiency (SCID).

DOSAGE AND ADMINISTRATION

For intravenous use only.

Dose

Table 1 Recommended Dose

Dose	Infusion Number	Initial Infusion Rate	Maintenance Infusion Rate
300 - 800 mg/kg body weight every 21 or 28 days	For the 1st Infusion	1 mg/kg/min (0.01 mL/kg/min)	Double the infusion rate every 30 minutes (if tolerated) up to 8 mg/kg/min (0.08 mL/kg/min)
300 - 800 mg/kg body weight every 21 or 28 days	From the 2nd Infusion	2 mg/kg/min (0.02 mL/kg/min)	Double the infusion rate every 15 minutes (if tolerated) up to 8 mg/kg/min (0.08 mL/kg/min)

Significant differences in the half-life of IgG among patients with PI may necessitate the dose and frequency of immunoglobulin therapy to vary from patient to patient. Determine the proper dose and frequency by monitoring clinical response.

Measles Exposure

If a patient has been exposed to measles, consult with physician to administer an extra dose of IGIV as soon as possible and within 6 days of exposure. A dose of 400 mg/kg should provide a serum level > 240 mIU/mL of measles antibodies for at least two weeks.

If a patient is at risk of future measles exposure and receives a dose of less than 530 mg/kg every 3 - 4 weeks, then the dose should be increased to at least 530 mg/kg. This should provide a serum level of 240 mIU/mL of measles antibodies for at least 22 days after infusion.

Administration

- Monitor vital signs throughout the infusion. Slow or stop the infusion if adverse reactions occur. If symptoms subside, the infusion may be resumed at a lower rate that is comfortable for the patient.
- Ensure that patients with pre-existing renal insufficiency are not volume depleted. For patients at increased risk of renal dysfunction or thrombotic events, administer ALYGLO at the minimum infusion rate practicable, and consider discontinuation of administration if renal function deteriorates [*see Boxed Warning, Warnings and Precautions*].
- After administration, the infusion line may be flushed with either normal saline or 5% dextrose in water.

CONTRAINDICATIONS

ALYGLO is contraindicated in:

- Patients who have a history of anaphylactic or severe system reaction to the administration of human immune globulin.
- IgA-deficient patients with antibodies to IgA and a history of hypersensitivity [*see Warnings and Precautions*].

WARNINGS AND PRECAUTIONS

Hypersensitivity: Severe hypersensitivity reactions may occur[1]. In case of hypersensitivity, discontinue ALYGLO infusion immediately and institute appropriate treatment. Have epinephrine available for immediate treatment of severe acute hypersensitivity reactions.

ALYGLO contains trace amounts of IgA (≤ 100 mcg/mL). Patients with known antibodies to IgA may have a greater risk of developing potentially severe hypersensitivity and anaphylactic reactions. ALYGLO is contraindicated in IgA-deficient patients with antibodies against IgA or a history of hypersensitivity reaction [*see Contraindications*].

Thrombotic Events: Thrombosis may occur following treatment with ALYGLO[1]. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling central vascular catheters, hyperviscosity and cardiovascular risk factors. Thrombosis may occur in the absence of known risk factors.

Consider baseline assessment of blood viscosity in patients at risk for hyperviscosity, including patients with cryoglobulins, fasting chylomicronemia/markedly high triacylglycerols (triglycerides), or monoclonal gammopathies. For patients at risk of thrombosis, administer ALYGLO 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 [*see Boxed Warning, Dosage and Administration*].

Renal Failure: Renal dysfunction, acute renal failure, osmotic nephropathy, and death[1] may occur upon use of ALYGLO. Ensure that patients are not volume-depleted before administering ALYGLO. Monitor renal function and urine output periodically, especially in patients who are at higher risk of renal failure. Assess renal function, including measurement of blood urea nitrogen (BUN) and serum creatinine before the initial infusion of ALYGLO and at appropriate intervals thereafter. If renal function deteriorates, consider discontinuing ALYGLO. In patients who are at risk of developing renal dysfunction, because of pre-existing renal insufficiency or predisposition to acute renal failure (such as diabetes mellitus, hypovolemia, overweight, use of concomitant nephrotoxic medicinal products or age > 65 years), administer ALYGLO at the minimum infusion rate practicable [*see Boxed Warning, Dosage and Administration*].

Hyperproteinemia, Increased Serum Viscosity, and Hyponatremia: Hyperproteinemia, increased serum viscosity, and hyponatremia may occur in patients receiving ALYGLO. It is critical to clinically distinguish true hyponatremia from a pseudohyponatremia that is associated with or causally related to hyperproteinemia with concomitant decreased calculated serum osmolality or elevated osmolar gap. Such treatment aimed at decreasing serum free water in patients with pseudohyponatremia may lead to volume depletion, a further increase in serum viscosity, and a possible predisposition to thrombotic events[1].

Aseptic Meningitis Syndrome (AMS): AMS may occur with ALYGLO. AMS usually begins within several hours to 2 days following ALYGLO treatment. Discontinuation of treatment has resulted in remission of AMS within several days without sequelae[1].

AMS may occur more frequently with high doses (2 g/kg) and/or rapid infusion of ALYGLO. AMS is characterized by the following signs and symptoms: Severe headache, nuchal rigidity, drowsiness, fever, photophobia, painful eye movements, nausea, and vomiting. Cerebrospinal fluid (CSF) studies frequently reveal pleocytosis up to several thousand cells per cubic millimeter, predominantly from the granulocytic series, and elevated protein levels up to several hundred mg/dL, but negative culture results. Conduct a thorough neurological examination on patients exhibiting such signs and symptoms, including CSF studies, to rule out other causes of meningitis.

Hemolysis: ALYGLO may contain blood group antibodies that can act as hemolysins and induce *in vivo* coating of red blood cells (RBCs) with immunoglobulin, causing a positive direct antiglobulin test (DAT) (Coombs test) result and hemolysis[1]. Delayed hemolytic anemia due to enhanced RBC sequestration, and acute hemolysis, consistent with intravascular hemolysis, have been reported. Cases of severe hemolysis-related renal dysfunction/failure or disseminated intravascular coagulation have occurred following infusion of IGIV.

Hemolysis (cont.):

The following risk factors may be associated with the development of hemolysis following IGIV administration: High doses (e.g., 2 g/kg or more), given either as a single administration or divided over several days, and non-O blood group. Other individual patient factors, such as an underlying inflammatory state (as may be reflected by, for example, elevated C-reactive protein or erythrocyte sedimentation rate), have been hypothesized to increase the risk of hemolysis following administration of IGIV[1], but their role is uncertain.

Closely monitor patients for clinical signs and symptoms of hemolysis, particularly patients with risk factors noted above. Consider appropriate laboratory testing in higher risk patients, including measurement of hemoglobin or hematocrit.

If clinical signs and symptoms of hemolysis or a significant drop in hemoglobin or hematocrit have been observed, perform confirmatory laboratory testing, including direct antiglobulin test. If transfusion is indicated for patients who develop hemolysis with clinically compromising anemia after receiving ALYGLO (immune globulin intravenous, human-stwk), perform adequate cross-matching to avoid exacerbating ongoing hemolysis.

Transfusion-Related Acute Lung Injury (TRALI): Noncardiogenic pulmonary edema [Transfusion-Related Acute Lung Injury (TRALI)] may occur in patients administered ALYGLO[1]. TRALI is characterized by severe respiratory distress, pulmonary edema, hypoxemia, normal left ventricular function, and fever. Signs and symptoms typically appear within 1 to 6 hours following treatment. Patients with TRALI may be managed using oxygen therapy with adequate ventilator support.

Monitor patients for pulmonary adverse reactions. If TRALI is suspected, perform appropriate tests for the presence of antineutrophil antibodies and anti-human leukocyte antigen (HLA) antibodies in both the product and the patient's serum.

Transmissible Infectious Agents: Because ALYGLO is made from human blood, it may carry a risk of transmitting infectious agents, e.g., viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent. The risk of infectious agent transmission has been reduced by screening plasma donors and by including virus inactivation/removal steps in the manufacturing process of ALYGLO.

Report all infections thought by a physician possibly transmitted by ALYGLO to GC Biopharma USA, Inc. at 1-833-426-6426. Discuss the risks and benefits of its use with the patient before prescribing or administering this product.

Monitoring Laboratory Tests

- Periodic monitoring of renal function and urine output is particularly important in patients at increased risk of developing acute renal failure. Assess renal function, including measurement of blood urea nitrogen (BUN) and serum creatinine before the initial infusion of ALYGLO and at appropriate intervals thereafter.
- Because of the potential for increased risk of thrombosis with ALYGLO, consider baseline assessment of blood viscosity in patients at risk for hyperviscosity, including those with cryoglobulins, fasting chylomicronemia/markedly high triacylglycerols (triglycerides), or monoclonal gammopathies.
- If signs and/or symptoms of hemolysis are present after an infusion of ALYGLO, perform appropriate laboratory testing for confirmation.
- If TRALI is suspected, perform appropriate tests for the presence of anti-neutrophil antibodies in both the product and patient's serum.

Interference with Laboratory Tests: After infusion of immunoglobulin, the transitory rise of the various passively transferred antibodies in the patient's blood may yield positive serological testing results, with the potential for misleading interpretation. Passive transmission of antibodies to erythrocyte antigens (e.g., A, B, and D) may cause a positive direct or indirect antiglobulin (Coombs) test.

ADVERSE REACTIONS

The most common adverse reactions, observed in ≥ 5% of study subjects, were headache, nausea/vomiting, fatigue, nasal/sinus congestion, rash, arthralgia, diarrhea, muscle pain/aches, infusion site pain/swelling, abdominal pain/discomfort, cough, and dizziness.

Clinical Trials Experience: Because clinical trials are conducted under widely varying conditions, adverse reaction rates observed in the clinical trials of a drug cannot be directly compared to rates in the clinical trials of another drug and may not reflect the rates observed in practice.

In an open-label, single-arm, multicenter, non-randomized clinical trial, 33 subjects with primary humoral immunodeficiency received doses of ALYGLO ranging from 319 mg/kg to 817 mg/kg every 21 days or 28 days for up to 12 months.

The passive transfer of antibodies with IGIV administration may interfere with the response to live virus vaccines such as measles, mumps, rubella, and varicella. Immunizing physicians should be informed of recent IGIV therapy so that appropriate measures may be taken.

Twenty-eight subjects (85%) experienced a total of 145 temporally associated adverse reactions (adverse events that occurred during or within 72 hours after the end of an infusion) during the study. The temporally associated ARs were headache (13 subjects, 39%), nausea/vomiting (11 subjects, 33%), fatigue (6 subjects, 18%), nasal/sinus congestion (5 subjects, 15%) rash (4 subjects, (12%), arthralgia, diarrhea (3 subjects, 9% each), muscle pain/aches, infusion site pain/swelling, abdominal pain/discomfort, cough, dizziness (2 subjects, 6% each).

These are presented in Table 2. There were no deaths and no adverse reactions leading to withdrawal from the study.

0008-01-2024 3/2024

Table 2 Adverse Reactions* (ARs) (within 72 hours after the end of an ALYGLO infusion) in ≥ 5% of Subjects

Adverse Reactions (ARs)	No. of Subjects Reporting ARs (Percentage of Subjects) [N[1]=33]	No. of Infusions with ARs (Percentage of Infusions) [N[2]=427]
Headache	13 (39)	32 (7.5)
Nausea/vomiting	11 (33)	20 (4.7)
Fatigue	6 (18)	18 (4.2)
Nasal/sinus congestion	5(15)	5 (1.2)
Rash	4 (12)	4 (0.9)
Arthralgia	3 (9)	4 (0.9)
Diarrhea	3 (9)	3 (0.7)
Muscle pain/aches	2 (6)	7 (1.6)
Infusion site pain/swelling	2 (6)	6 (1.4)
Abdominal pain/discomfort	2 (6)	3 (0.7)
Cough	2 (6)	2 (0.5)
Dizziness	2 (6)	2 (0.5)

*Adverse events that occurred during or within 72 hours after the end of an infusion
[1]Total number of subjects
[2]Total number of infusions

Postmarketing Experience: Because postmarketing reporting of adverse reactions is voluntary and from a population of uncertain size, it is not always possible to reliably estimate the frequency of these reactions or establish a causal relationship to product exposure. The following adverse reactions have been identified and reported during the post-approval use of marketed IGIV products:

Blood and lymphatic system disorders: leukopenia, hemolysis, pancytopenia; **Immune system disorders:** hypersensitivity (e.g., anaphylaxis), anaphylactic shock, anaphylactic reaction, anaphylactoid reaction, allergic reaction, angioedema, face edema; **Metabolic and nutritional disorders:** fluid overload, (pseudo) hyponatremia; **Psychiatric disorders:** agitation, confusion, anxiety, nervousness; **Nervous system disorders:** coma, loss of consciousness, seizures, (acute) encephalopathy, cerebrovascular accident, stroke, aseptic meningitis, migraine, speech disorder, paresthesia, hypoesthesia, photophobia, tremor; **Cardiac disorders:** myocardial infarction, cardiac arrest, angina pectoris, tachycardia, bradycardia, palpitations, cyanosis; **Vascular disorders:** hypotension, (deep vein) thrombosis, peripheral circulatory failure/collapse, hypertension, phlebitis, pallor; **Respiratory, thoracic and mediastinal disorders:** apnea, Acute Respiratory Distress Syndrome (ARDS), TRALI, respiratory failure, pulmonary embolism, pulmonary edema, bronchospasm, dyspnea, hypoxia, wheezing, cough; **Gastrointestinal disorders:** diarrhea, hepatic dysfunction, abdominal discomfort; **Skin and subcutaneous tissue disorders:** eczema, urticaria, rash (erythematous), dermatitis, pruritus, alopecia, Stevens-Johnson syndrome epidermolysis, skin exfoliation, erythema (multiform), dermatitis (e.g., bullous dermatitis); **Musculoskeletal and connective tissue disorders:** back pain, arthralgia, myalgia, musculoskeletal pain, muscle stiffness, pain in extremity, neck pain, muscle spasm; **Renal and urinary disorders:** acute renal failure, osmotic nephropathy, renal pain; **General disorders and administration site conditions:** injection-site reaction, chills, chest pain or discomfort, hot flush, flushing, flu-like illness, feeling cold or hot, edema, hyperhidrosis, malaise, asthenia, lethargy, burning sensation; **Investigations:** hepatic enzymes increased, oxygen saturation decreased, falsely elevated erythrocyte sedimentation rate, positive direct antiglobulin (Coombs) test.

DRUG INTERACTIONS

Clinical studies have not evaluated mixture of ALYGLO with other drugs and intravenous solutions. It is recommended that ALYGLO is administered separately from other drugs or medications which the patient may be receiving. Do not mix the product.

Transitory rise of the various passively transferred antibodies in the patient's blood after infusion of immunoglobulin may yield positive serological testing results, with the potential for misleading interpretation.

USE IN SPECIFIC POPULATIONS

Geriatric use: In patients over age 65 or in any patient at risk of developing renal insufficiency, do not exceed the recommended dose, and infuse ALYGLO at the minimum infusion rate practicable.

Reference: 1. ALYGLO Prescribing Information. GC Biopharma USA, Inc.; 2023.

Manufactured by:
GC Biopharma Corp.
107 Ihyeon-ro 30-beongil
Giheung-gu, Yongin-si
Gyeonggi-do 16924
Republic of Korea
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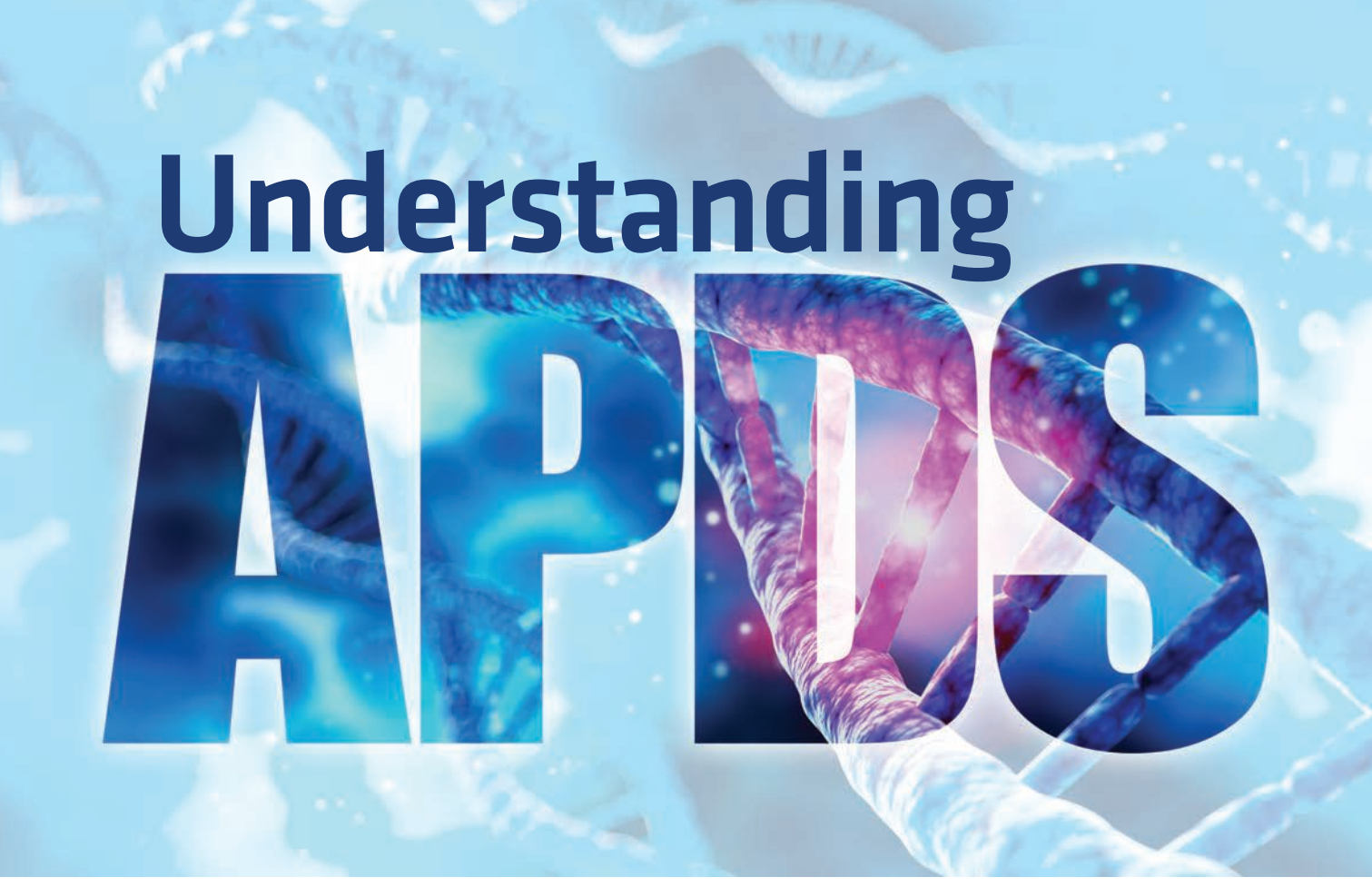
500 Frank W. Burr Boulevard
Teaneck, NJ 07666 USA

Understanding APDS



A lot has been learned about this primary immunodeficiency disorder despite its relatively recent discovery. While diagnosis is often delayed, there are effective treatments and a great deal of support for patients and their caregivers.

By Ronale Tucker Rhodes, MS

MORE THAN six million people are affected by primary immunodeficiency disorders (PI), also known as inborn errors of immunity, around the world today, and it is believed 70 to 90 percent of those are still undiagnosed. PIs are rare diseases caused by inherited defects in one or more component of the immune system, which leaves people living with these defects more prone than others to infections, as well as severe autoinflammation, autoimmunity, allergy and malignancy.[1] Today, close to 500 different PIs have been identified, with agammaglobulinemia the first to be described in 1952.[2]

The discovery of new PIs has been expanding at a rapid pace. One of the more recent to be identified is activated PI3K delta syndrome (APDS), previously known as PASLI disease, with only 256 cases reported globally as of 2024.[3,4] However, the exact number of people with APDS is unknown because many patients are not diagnosed or registered.[5] The actual number may be closer to one to two million people.

What Is APDS?

APDS is a rare underdiagnosed PI that was first characterized in 2013.

There are two types: APDS1 is due to variants in the PIK3CD gene and APDS2 is due to variants in the PIK3R1 gene.[6] Both genes are vital to the development and function of immune cells in the body.[3] Individuals often have abnormally functioning numbers of white blood cells (WBCs) (lymphopenia), particularly B cells and T cells, which normally recognize and attack foreign invaders, such as viruses and bacteria, to prevent infection.[8]

APDS affects males and females equally.[6] The median age of people diagnosed with APDS1 is 13 years, and the median age of people with APDS2 is 14 years.[7]

Due to the rarity of the disease and the heterogeneous clinical picture, many patients are not diagnosed until

years after symptom onset. Another challenge is the large number of PIK3CD and PIK3R1 variants whose functional significance for developing APDS is inconclusive.[5]

Symptoms of APDS

APDS1 and APDS2 have similar symptoms and are inherited in an autosomal dominant pattern in families, which means one copy of the altered gene in each cell is sufficient to cause the disorder.[6]

Symptoms include recurrent infections, particularly in the lungs, sinuses and ears, beginning in childhood. Those who experience recurrent respiratory tract infections can lead to a condition called bronchiectasis, which damages the passages leading from the windpipe to the lungs (bronchi) and can cause breathing problems. Symptoms may also include chronic active viral infections such as Epstein-Barr virus (EBV), herpes simplex virus or cytomegalovirus (CMV).

A possible symptom of APDS is abnormal clumping of WBCs that can lead to enlarged lymph nodes (lymphadenopathy) or an enlarged spleen (splenomegaly). These WBCs can also build up to form solid masses (nodular lymphoid hyperplasia), usually in the moist lining of the airways or intestines.

Those with APDS also have an increased risk of developing forms of blood cancer called Hodgkin lymphoma and non-Hodgkin lymphoma. In addition, some may develop autoimmunity, which occurs when the body attacks its own tissues and organs by mistake.[8]

However, the severity of APDS varies widely. In some people, symptoms are very mild, while in others, they are much more severe. Typically, most people with APDS have symptoms beginning in childhood, but symptoms can begin at any age, with some people failing to develop symptoms until adulthood. What's more, there are some reports of people who have been diagnosed with APDS who do not have any symptoms.[6]

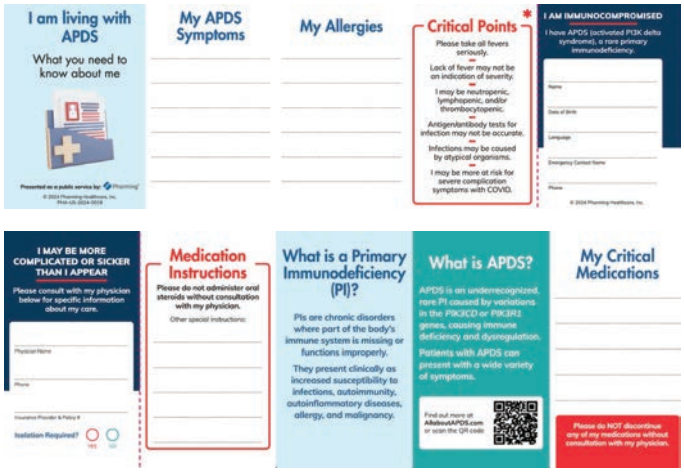
Diagnosing APDS

An APDS diagnosis is based on clinical manifestations (symptoms and clinical exam), a detailed family history and lab tests that look for abnormalities in the levels and function of immune cells.[6]

The website All About APDS in its "Steps to Diagnose APDS" details what is looked for in clinical manifestations, especially symptoms of immune deficiency (severe recurrent infections; persistent herpesvirus, especially EBV and/or CMV; recalcitrant or rare bacterial, viral and fungal infections, including abscesses, skin lesions/infections, eye infections and oral ulcerations; and low IgA or IgG levels), immune dysregulation (autoimmunity, particularly cytopenias, lymphoproliferation, lymphoma and enteropathy,

APDS Preparedness Wallet Card

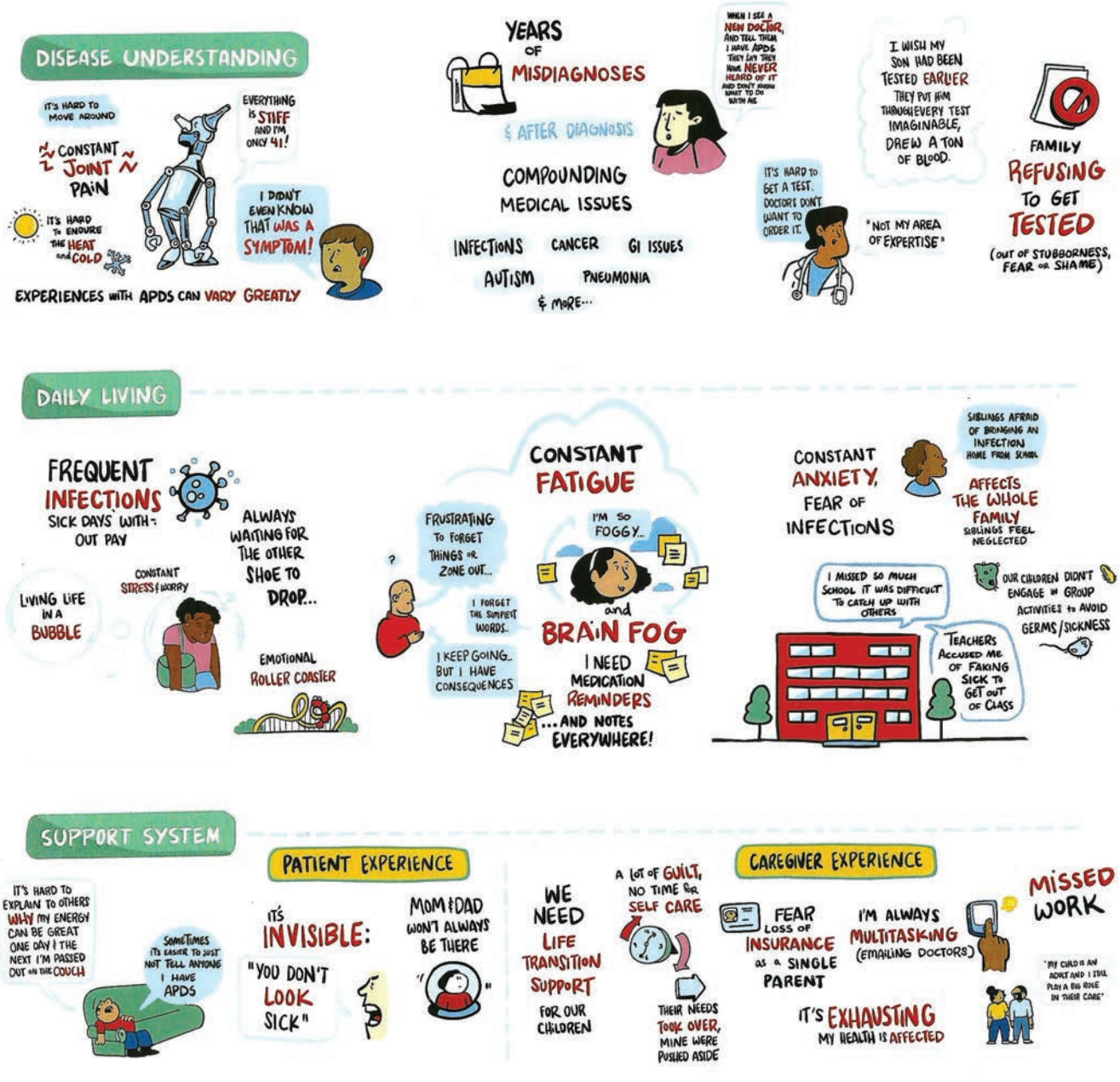
- **Dual sided information on PI and APDS for use when interacting with someone who may not be familiar with either.**
- **APDS is often unfamiliar to hospitals. This card communicates essential medical information for emergency personnel about how to handle APDS.**
- **The card gives patients and caregivers confidence to travel to unfamiliar areas.**



including failure to thrive), combined immune deficiency/immune dysregulation (end organ damage, for example, bronchiectasis); and neurodevelopmental delay or failure to thrive.[9]

However, a definitive diagnosis can only be made with genetic testing. This is because APDS can mimic signs of other PIs. Genetic testing via a blood or saliva sample is conducted using a gene panel that tests for pathogenic variants in a group of genes related to disorders that have similar symptoms.[6]

It should be noted that genetic testing can be expensive. However, Pharming Healthcare, a biopharmaceutical company developing and commercializing protein replacement therapies and precision medicines to treat rare disease patients, provides a no-charge genetic testing program through Navigate APDS (navigateapds.com)



Source: Pharming Healthcare. Insights Into APDS: Revealing the Patient Experience

for eligible patients. Pharming also provides information about the program to healthcare professionals and distributes requisition forms for the test and test kits to them. The actual tests and services are performed by PreventionGenetics.[9]

Treating APDS

Treatment for APDS is focused on managing symptoms, preventing infections and lowering inflammation. Treatments include antibiotics to treat infections, immunosuppressive medicines, long-term immune globulin (IG) replacement therapy and hematopoietic stem-cell transplant (HSCT).[6]

Ear tubes (tympanostomy tubes) may be needed for children with APDS who experience frequent ear infections. Some children may need to have their tonsils removed because of swelling and sinus infections. People with more severe APDS may eventually require respiratory support, including extra oxygen and chest physiotherapy, as well as physical, occupational and/or speech therapy for long-term complications.[9]

According to All About APDS, antibiotics are the most common antimicrobial prophylaxis and are prescribed for 61 to 79 percent of patients. Forty-six percent of patients receive corticosteroids, with 87 percent of those showing at least partial short-term benefit. Rituximab, which targets CD20+ B cells for destruction and therefore may reduce B cell overactivation, has been used to treat lymphoma and autoimmune diseases. Between 11 and 38 percent of patients have received mTOR inhibitors, one of the downstream targets of PI3K, signalling that it has a significant role in regulating immune responses.

All About APDS also reports that IG therapy is used to address sinopulmonary infections or autoimmune cytopenias in 63 to 89 percent of patients, starting at a reported median age of 5 years, with nearly half of patients receiving treatment by age 10 years. IG therapy can be administered either intravenously or subcutaneously and is often used in combination with antibiotics.

Only between nine and 17 percent of patients who do not respond to therapy have been treated with HSCT. This is because while HSCT has the potential to resolve the clinical symptoms of APDS, it is associated with a relatively high risk of post-transplant morbidity and mortality. In fact, 91 percent of patients with APDS experienced adverse events during and/or after HSCT.[10]

In 2023, Pharming's Joenja (leniolisib) was the first and only treatment approved by the U.S. Food and Drug Administration (FDA) for APDS in adults and children 12 years of age and older. Joenja works to control the hyperactive PI3K delta pathway, which helps to rebalance the immune system, thus working on the source of the disease itself, rather than just the symptoms.[11]

Approval of Joenja was based on a 12-week blinded, randomized, placebo-controlled study of 31 adult and pediatric patients 12 years of age and older with confirmed APDS-associated genetic PI3Kδ mutation, with a documented variant in either PIK3CD or PIK3R1. During the study, 21 patients received 70 mg of Joenja and 10 received placebo twice a day for 12 weeks. The co-primary efficacy endpoints were improvement in lymphoproliferation (reduction in the size of lymph nodes) and the normalization of immunophenotype as measured by the percentage of naïve B cells out of total B cells. By day 85 of the study, patients taking Joenja saw a reduction in lymph node size and a 37 percent improvement in naïve B cells counts compared to placebo, indicating a correction of the underlying immune defect.[12]

Burden of APDS

APDS can have a significant disease burden on patients and their families.

A study published in 2024 in *Orpharnet Journal of Rare Diseases* and funded by Pharming Healthcare that explored the disease burden of APDS described the day-to-day experience and health-related quality of life (HRQoL) impact of APDS from the perspective of individuals living with the condition, caregivers and treating physicians. In the study, qualitative interviews were conducted with healthcare providers (HCPs), individuals with APDS and caregivers. Some individuals and caregivers also completed a narrative account exercise. Six HCPs participated in an interview. Seven participants completed the narrative account exercise (N = 5 caregivers and N = 2 individuals with APDS) and 12 took part in an interview (N = 4 caregivers and N = 8 individuals with APDS).

"Themes identified from HCP interviews included symptoms, clinical manifestations, HRQoL impacts and treatments/management of APDS. The narrative account exercise identified similar themes, but with the addition to the journey to diagnosis. These themes were explored during the individual/caregiver interviews. Reported clinical manifestations and symptoms of APDS

included susceptibility to infections, lymphoproliferation, gastrointestinal (GI) disorders, fatigue, bodily pain and breathing difficulties. HRQoL impacts of living with APDS included negative impacts to daily activities, including work, education and social and leisure activities, physical functioning, as well as emotional well-being, such as concern for the future, and interpersonal relationships. Impacts to caregiver HRQoL included negative impacts to physical health, work, emotional well-being, interpersonal relationships and family life and holidays."[13]

Another study of survival rates in APDS patients published in 2024 found, "despite available treatments, survival for individuals with APDS appears to be shortened from the average lifespan. A Kaplan–Meier survival analysis for APDS showed the conditional survival rate at the age of 20 years was 87 percent, age of 30 years was 74 percent and ages of 40 and 50 years were 68 percent. Review of causes of death showed that the most common cause of death was lymphoma, followed by complications from HSCT. The overall mortality rate for HSCT in APDS1 and APDS2 cases was 15.6 percent, while the mortality rate for lymphoma was 47.6 percent."[14]

Support Resources

Despite how recently APDS was discovered, a surprising number of resources have been created by Pharming Healthcare for patients and their caregivers. The APDS Assist website provides patients' support through an APDS Clinical Educator, assistance with insurance coverage and authorization requirements for treatment with Joenja, and offers personal help through a care coordinator.

Pharming Healthcare also has a patient advocacy arm. According to Kristie Cline, head of patient advocacy at Pharming, the company's patient advocacy program supports both APDS and hereditary angioedema. For APDS, the program's staff conducted two separate focus groups in 2023, one for patients and one for caregivers. "I believe speaking to the groups separately allows each type of participant to share their true thoughts and feelings without holding back for fear of hurting someone else in the focus group," explains Cline. These groups are conducted virtually, which Cline says "is great for the APDS community because many of them are immunocompromised and can participate without the fear of being in a room with others and getting sick."

Pharming's Patient Advocacy Key Learning Points from Patient Focus Groups

- Fatigue and infections have the biggest impact on their life.
- Fatigue is the most insidious and challenging symptom.
- Brain fog induces forgetfulness and is linked to fatigue.
- Increased risk of infection impacts daily activities.
- Patients suffer from joint pain and temperature sensitivity.
- Patients don't connect APDS with symptoms and challenges.
- Frequent infections result in financial impact for patients.
- Misdiagnosis correlates with suboptimal care and mistrust among healthcare providers.
- Patients have varied genetic testing experience.
- Living with an invisible disease is far-reaching.
- Support systems vary; patients speak highly of APDS Assist.
- Patients desire support, connection and information.
- 100% of participants responded with a desire for more emotional/mental health support.
- Patients are unified in voicing the need for a dedicated patient organization.
- Patients recognized the need for more patient-friendly disease information and real patient stories.

Source: Pharming. Patient & Caregiver Focus Group Insights, September 2023.

Pharming's Patient Advocacy Key Learning Points from Caregiver Focus Groups

- The APDS caregiver role is multifaceted and demanding.
- The emotional impact on caregivers is intense.
- Caregiving continues with life transitions, and challenges evolve.
- Responsibilities of caregiving impact employment and financial strain.
- Most caregivers experience near constant stress.
- Caregivers feel guilty that their other children pay a steep price.
- There is considerable time consumed by medical appointments and hospitalizations.
- Caregivers neglect their own self care.
- Fatigue and infections limit daily life.
- Caregivers aren't aware of resources to help.
- APDS contributes to compounding medical issues and complications.
- Frequent illness takes a toll on their child's mental health.
- Caregivers worry most about life transitions.
- Caregivers are desperate for community and connection; they want a formal patient organization for APDS.
- Caregivers want financial and mental health support.

Source: Pharming. Patient & Caregiver Focus Group Insights, September 2023.

Information learned from these first two focus groups was also used to create an abstract published in *Allergy & Asthma Proceedings* and two posters that were presented at the Eastern Allergy Conference and the National Organization of Rare Disorders (NORD) Breakthrough Summit to raise awareness for the APDS patient and caregiver lived experience. These focus groups are conducted once a year, and Pharming is hoping to expand them to twice a year in 2025.

In addition to the focus groups, Pharming's All About APDS website provides educational webinars, downloads (such as the 10 Warning Signs of APDS), videos, podcasts, a list of frequently asked questions and links to organizations, genetic testing, an APDS preparedness wallet card and more.[3]

"One of the most well-received resources that we created in partnership with the Jeffrey Modell Foundation as a public service announcement is the '10 Warning Signs of APDS.' These have been translated to Spanish, French-Canadian, German and Italian, and we are in the process of translating them into Hebrew, and they are being used now globally to educate and raise awareness for APDS," says Cline.

"When you have a rare disorder that was just described 11 years ago, it's important to share information as broadly as possible to educate not only the patient community, but the professional community as well. The '10 Warning Signs' were developed with input from patients, caregivers and healthcare professionals and written in patient-friendly language to make it easier for patients and caregivers to have a discussion with their physician who may or may not be aware of APDS."

Ongoing Research

Currently, studies are being conducted to develop therapies targeted at the specific genes affected in APDS, as well as drugs known as mTOR inhibitors. mTOR inhibitors have been shown to help regulate the immune system and have been helpful in reducing swelling and inflammation symptoms in some patients with APDS.[6]

On the ClinicalTrials.gov website, there are currently two studies actively recruiting, both of which are investigating leniolisib (Joenja). One of these studies (NCT05693129) is a two-part, prospective, open-label, single arm, multicenter study to evaluate the safety, tolerability, pharmacokinetics, pharmacodynamics and efficacy of leniolisib granules in at least 15 pediatric patients (aged 1 to 6 years) with activated APDS.[15] The other study (NCT06249997) is an open-label, non-randomized study to assess the safety and efficacy of Joenja in Japanese patients with APDS followed by an open-label long-term extension study (NCT05438407) in pediatric patients age 4 through 11, which has completed enrollment and is ongoing.[16]

New Treatments Are Needed

APDS is a relatively newly identified and rare disease that is difficult to diagnose since its symptoms are similar to other PIs. Genetic testing is the only way to definitively diagnose the disease, but certain criteria must first be met to undergo genetic testing.

While there are many effective treatments for APDS, the estimated annual cost ranges from $83,057 to $793,620.[17] However, these treatments aren't effective for all APDS patients, making HSCT the only other option. Furthermore, survival and mortality data illustrate that new treatments are needed to prevent or delay disease progression and contribute to a better quality of life in individuals with APDS.

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RONALE TUCKER RHODES, MS, is the senior editor-in-chief of *IG Living* magazine.

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Profile: Maureen E. Basye, MS

By Trudie Mitschang

AFTER NUMEROUS diagnoses of celiac disease within her immediate family, Maureen Basye understood the travel industry limitations for patients living with severe dietary restrictions. As she reflected on how much she and her family enjoyed cruise ship vacations, in a flash of inspiration, she made the career transition from speech pathologist to celiac travel consultant. Today, as the founder of industry-changing Celiac Cruise, Maureen shares her passion for helping others enjoy safe, carefree adventures while making memories that last a lifetime.

Trudie: What inspired you to start Celiac Cruise?

Maureen: I started Celiac Cruise in 2018 with a dream to be able to provide families impacted by celiac disease the opportunity to sail the open seas in a safe, gluten-free environment. Celiac Cruise hosts various sailings around the world and has partnerships with Royal Caribbean International, Oceania Cruise Lines and AmaWaterways.

Trudie: How did celiac disease impact you personally?

Maureen: In September 2017, we — like so many others — got that phone call informing us that our son Peter's blood levels looked off. I was surprised, but then again with the last three years of recurrent pneumonias and awful constipation with *so* much pain, I wasn't. The momentary relief of hearing we possibly had an answer brought a combination of joy and fear. From there, the endoscopy and additional labs were scheduled, and approximately three months later, we had a diagnosis: celiac disease. Shortly thereafter, many additional familial diagnoses of celiac disease followed.

Trudie: What did you already know about the disease?

Maureen: Celiac disease is not something new to our family, as five of our loved ones have been diagnosed, including my husband Geoff and son Peter.

Trudie: What lifestyle adjustments did you have to make?

Maureen: The changes to our kitchen and home commenced, we had meetings with various teachers and school staff, and we soon recognized the approximate $500 a month added to our monthly grocery bill! *Ouch!* I found the first six months we were on autopilot, just go, go, go. A few steps forward and a few steps back. Peter started to feel better, but we had many hurdles to cross. Travel? How do we handle that? Can we even do that? How do I trust my child and family will be in safe hands in restaurants, let alone on a vacation?

Trudie: How did you navigate that challenge?

Maureen: Up to that point, traveling had always been a huge piece of our family memory making. This mama got to work and began exploring options. Since Peter's diagnosis, I've put my healthcare masters' degree job on the shelf and have become a passionate celiac/gluten-free travel consultant. There is so much out there, but also so many fears that many of us wrestle with daily. This insight into my background and value system helps explain my mission today: to connect exciting and safe travel opportunities with families and people who have the desire to travel — but aren't sure of the possibilities available to them.

Trudie: Why did you decide to pursue a celiac-focused cruise?

Maureen: Cruises have always been one of our family's crowd pleasers. It's a way to see the world where adults and children alike are able to celebrate together and there is so much to do. We had cruised gluten-free on a variety of cruise lines. Some do better than others, but efficiency continued to be an issue across all brands. I saw an opportunity.

Trudie: Tell us about the start of Celiac Cruise.

Maureen: In the fall of 2018, my business partner, Connie Saunders, and I reached out to Royal Caribbean and presented the idea of a gluten-free cruise to them. Not only were they open to it, they invited our family and team onboard to test drive and opened the kitchen galley doors for many meetings and a full gluten-free inspection. I was so impressed and have done my best to share that experience on my website. After many

meetings with executive chefs, food and beverage managers and Royal Caribbean executives, the inaugural celiac cruise was set. After months of planning, preparation, meetings and site visits, our very first cruise set sail in January 2020 and was a huge success. Our guests loved the freedom and the worry-free environment, and we can't even begin to thank all of our partners that helped us pull it off.

Trudie: How did the idea evolve after that initial cruise?

Maureen: We took a bit of a hiatus like the rest of the world due to COVID, and our next cruise wasn't until March 2022 when we had about 800 people join us. We obviously had ample time to make some minor adjustments from lessons learned the first time around, which included additional menu offerings, onboard events and increasing the variety of destinations and cruise line partnerships.

Trudie: How many destinations have you visited?

Maureen: Great question! So far, we've completed 15 cruises to the Bahamas, the Eastern and Western Caribbean, Paris on the Seine River, Germany, Austria, Budapest on the Danube, the Mediterranean and numerous trips to Alaska. We wrapped up 2024 with a cruise through Switzerland, France, Germany and the Netherlands. Looking ahead to 2025, we have nine exciting cruises planned, including Bordeaux, France, Norway and Denmark, and so many other destinations. Our goal remains to keep offering safe, gluten-free experiences that allow our guests to see the world without worry.

Trudie: How is your son's health today?

Maureen: Peter is doing well and has become a tremendous champion for Celiac Cruise. While he still faces the daily challenges of managing celiac disease, he approaches them with incredible resilience. His journey inspired me to start this mission, and now he plays a significant role in guiding our path in continuously advocating for safe, gluten-free travel.

Trudie: What has this experience taught you about yourself?

Maureen: Starting Celiac Cruise has taught me so much about resilience and the power of community. I learned that I have the determination to create something truly unique and meaningful, even when faced with challenges. Creating an environment people could truly trust wasn't easy, especially when our guests are literally placing their health in our hands. But it was a lot easier to continue to persevere knowing I was helping to create an environment where so many could vacation again without having to focus nonstop on every bite of food they put in their mouths.

Trudie: Planning a cruise is a lot of work. How do you stay motivated?

Maureen: Giving this community the chance to feel "normal," even if for just a few days, is truly the only motivation any person needs to create this kind of magic. Every time we see our guests relax and connect without the worry of their dietary restrictions, it reaffirms our dedication to Celiac Cruise. Overall, starting Celiac Cruise has shown me that with passion, perseverance and a supportive community, it's possible to create anything you want with the right team by your side.

Trudie: What advice do you have for other families diagnosed with celiac disease?

Maureen: Honestly, my biggest piece of advice is to take a deep breath and give yourself some time to adjust. It can feel *very* overwhelming at first, but there's a whole gluten-free world out there. Get to know your grocery stores and local restaurants, and don't hesitate to experiment in the kitchen — gluten-free cooking can be really fun! Also, don't be shy about reaching out to the celiac community. There are so many people who understand exactly what you're going through, and they can offer support, tips and even some delicious recipes (us/me included)! And remember, it's OK to have some tough days. Just focus on what works for your family, and soon enough, you'll find your way!

Trudie: What are you most proud of?

Maureen: I'm incredibly proud of how we've turned a challenging situation into something positive with Celiac Cruise. Watching my husband Geoff and our son Peter navigate life with celiac disease has been both inspiring and motivating. Instead of letting it hold us back, we channeled those challenges into creating a space where individuals can travel safely and enjoyably. What really touches my heart are the letters, emails and hugs we receive from our guests. Knowing that we've changed their lives for the better and given them a chance to feel normal, relax and truly unplug, even if just for a few days, is incredibly rewarding. It's amazing to see how we've created an environment where they can enjoy their vacations without worry. I love that my family is part of this journey with me, and it fills me with pride to know we're making a difference together.

Editor's note: To learn more about traveling with Celiac Cruise, visit CeliacCruise.com.

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

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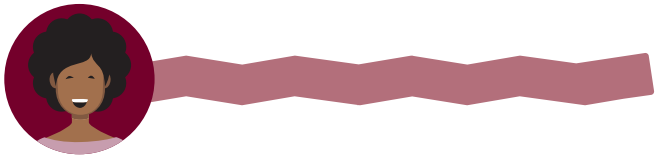

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Hizentra®, Immune Globulin Subcutaneous (Human), 20% Liquid, is a prescription medicine used to treat:

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- Chronic inflammatory demyelinating polyneuropathy (CIDP) in adults

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.

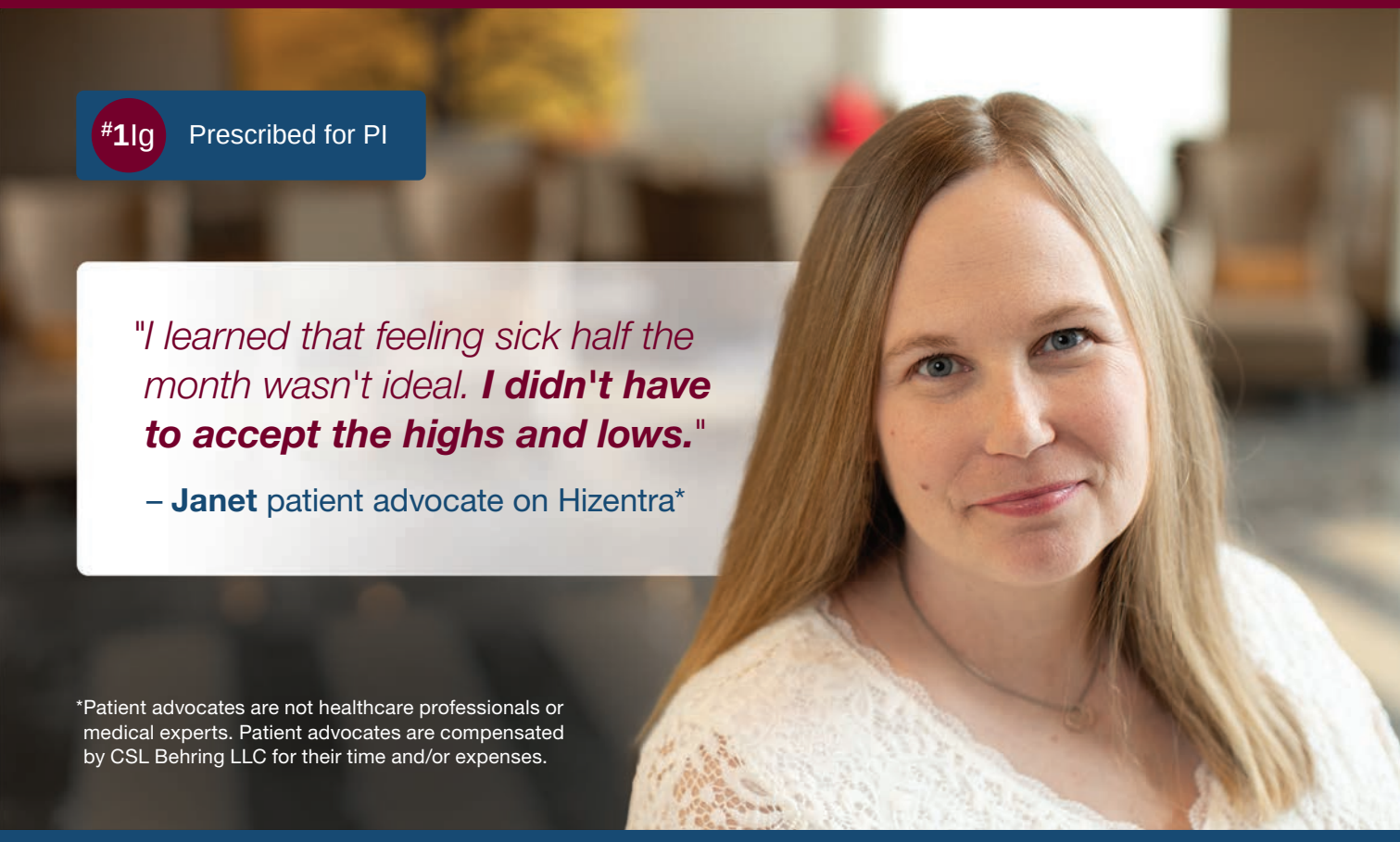
Treatment with Hizentra might not be possible if your doctor determines you have hyperprolinemia (too much proline in the blood), or are IgA-deficient with antibodies to IgA and a history of hypersensitivity. Tell your doctor if you have previously had a severe allergic reaction (including anaphylaxis)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

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

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

Immediately report to your physician any of the following

Please see Brief Summary of full Prescribing Information on reverse.



Janet, Melaine, and other PI patients are able to get back to daily living while on Hizentra

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 the patient product information.

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.

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BRIEF SUMMARY OF PRESCRIBING INFORMATION
These highlights do not include all the information needed to use HIZENTRA safely and effectively. Please see full prescribing information for HIZENTRA, which has a section with information directed specifically to patients.

What is HIZENTRA?
HIZENTRA is a prescription medicine used to treat primary immune deficiency (PI) and chronic inflammatory demyelinating polyneuropathy (CIDP). Infuse HIZENTRA only after you have been trained by your doctor or healthcare professional. HIZENTRA is to be infused under your skin only. DO NOT inject HIZENTRA into a blood vessel (vein or artery).

Who should NOT take HIZENTRA?
Do not take HIZENTRA if you have too much proline in your blood (called "hyperprolinemia") or if you have had reactions to polysorbate 80. Tell your doctor if you have had a serious reaction to other immune globulin medicines or have been told that you have a deficiency of the immunoglobulin called IgA.

Tell your doctor if you have a history of heart or blood vessel disease or blood clots, have thick blood, or have been immobile for some time. These things may increase your risk of having a blood clot after using HIZENTRA. Also tell your doctor what drugs you are using, as some drugs, such as those that contain the hormone estrogen (for example, birth control pills), may increase your risk of developing a blood clot.

What are possible side effects of HIZENTRA?
The most common side effects with HIZENTRA are:

- Redness, swelling, itching, and/or bruising at the infusion site
- Headache/migraine
- Nausea and/or vomiting
- Pain (including pain in the chest, back, joints, arms, legs)
- Fatigue
- Diarrhea
- Stomach ache/bloating
- Cough, cold or flu symptoms
- Rash (including hives)
- Itching
- Fever and/or chills
- Shortness of breath
- Dizziness
- Fall
- Runny or stuffy nose

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.

Tell your doctor right away if you have any of the following symptoms. They could be signs of a serious problem.

- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain and/or swelling of an arm or leg with warmth over the affected area, discoloration of an arm or leg, unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, or numbness or weakness on one side of the body. These could be signs of a blood clot.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of a brain swelling called meningitis.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a blood problem.
- Chest pains or trouble breathing.
- Fever over 100°F. This could be a sign of an infection.

Tell your doctor about any side effects that concern you. You can ask your doctor to give you more information that is available to healthcare professionals.

Please see full prescribing information, including full boxed warning and FDA-approved patient product information. For more information, visit Hizentra.com.

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

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

Based on April 2023 version.

CSL Behring

Granting Myself a Wish: Embracing Joy as an Adult with Chronic Illness

By Michelle Searle

MY HUSBAND and I recently participated in a Make-A-Wish walk in our community. As a child living with a chronic illness, I was fortunate enough to receive a wish from the Make-A-Wish Foundation myself. I remember the excitement of being asked what I wanted most. I chose a bedroom makeover. The wish not only provided me with a new and beautiful room, but it also shifted my focus. Instead of constantly thinking about doctor appointments, infusions, my medication schedule and feeling sick, I was able to think about what kind of bed I wanted and what color to paint my walls. I spent weeks looking through the Pottery Barn Teen catalog, imagining all the ways I could transform my room. This new focus brought me joy and distraction, allowing me to escape the challenges of my illness.

As I transitioned into adulthood, living with my chronic illness became a different journey still filled with the realities of medical appointments, infusions, medicine side effects and fatigue. But just because I'm an adult navigating the complexities of my illness, I realize now that the magic of wishes doesn't have to end when childhood does. Preparing for the Make-A-Wish walk made me realize the importance of treating myself and nurturing my own desires as an adult, too, and I asked myself: What would my wish be at this stage of my life?

The answer was easy: to attend the Taylor Swift Eras Tour concert in Miami. As a fan of hers for years, this event is more than just a concert; it represents a celebration of music, vulnerability, resilience and the powerful connection that songs can create. Also, since the concert took place in my hometown, it was a great chance to catch up with my family. With my dad working security for the event, I knew I'd also have the opportunity to see and hang out with him there. After spending weeks debating whether I could make this dream a reality, I finally decided to spend the money to go, and that's when the fun planning began.

After monitoring ticket prices for weeks, I secured my ticket and began exploring outfit and nail ideas. I ordered several outfit options and spent time trying them on to choose the best one. I also ordered a clear purse for the stadium and beads to make friendship bracelets. Of course, there were logistical challenges, such as ensuring I wouldn't be due for an infusion around the day of the concert so any side effects would prevent me from going. I also brought masks for the flights and one for the concert, which I wore while navigating through the crowds, not during the concert itself.

The day of the concert arrived, and I was filled with excitement as I made my way to the stadium. When I entered, I was enveloped in a sea of fellow Swifties, all buzzing with anticipation and trading friendship bracelets. The energy in the air was electric, and I felt waves of gratitude for being able to experience this. Fulfilling this wish was more than just attending a concert; it became a powerful reminder of the importance of prioritizing joy in my life. I left the concert with my heart filled with happiness. The experience reaffirmed that it's not just OK to grant yourself a wish — it's essential.

For those reading this, consider your own wishes. What brings you joy? What small or grand adventure can you embark on? Life with a chronic illness often demands that we put our needs on the back burner, but it doesn't have to be that way. Embrace the idea that you deserve moments of happiness and fulfillment. Whether it's attending a concert, pursuing a long-desired hobby or simply taking a day for self-care, allowing yourself to dream and pursue those dreams can be incredibly therapeutic. It's a testament to resilience and self-love. So go ahead, make that wish for yourself, and take the steps to turn it into reality. You deserve it.

Reflecting on my journey, I understand that life may present challenges, but it also offers countless opportunities for joy. By granting myself a wish as an adult, I've learned that the magic of dreaming is timeless. Celebrate your dreams, no matter your circumstances, and remember: It's never too late to make a wish. IG

MICHELLE SEARLE is a teacher from South Florida who was diagnosed with common variable immunodeficiency at 11 years old. She is currently living in New York where you will most likely find her eating pizza or trying to make friends with the local cats.

Putting One Foot in Front of the Other

By Megan Ryan

LET ME take you back to fourth grade PE class — the part of the school day I hated. I loved school but I was the least athletically inclined kid in the building. Give me problems to solve, books to read and academic projects any hour of the day, and I was in my element. But balls, nets and rackets were just not my thing. I was almost always picked last for a team. And I dreaded the weeks leading up to the annual Presidential Fitness Test, including the run and sit-ups. All in all, participation in sports was not for me.

But in high school every kid was required to pick a sport and play for two years because small Texas towns have to field a team in all the sports! I opted for golf because that's what a friend was doing and the uniform was reasonable — khaki shorts and a collared shirt. And I could attend a summer golf camp on a university campus for a week!

I completed my two years of required team sports but never did fully embrace the sport of golf. I did learn to love "walking the course," and that instilled in me a love of walking for health and fitness. I enjoyed being outside in nature and looking up at the trees, observing the different types of grasses growing and checking out the birds and other animals that called the golf course home.

Thirty years later, I'm still walking, just not a golf course. I love the simplicity of walking — no special equipment is required other than a good pair of sneakers and making sure I use sunscreen to protect my skin. Because I live on the Gulf Coast of Texas, I can walk outside all year. And just about anywhere I travel I can find a walking path or trail or just explore city streets. I've made walking for health and fitness a part of my daily routine. Every morning, I'm out of bed and lacing up my sneakers, getting out the door when the sun is coming up. On an average day, I walk four brisk miles, and on a good day, I take it up to six or more miles. Even when I'm not feeling that great, I get up and get out for a mile walk or so because I know it's going to make me feel better in the long run.

On my daily walks, I have at least an hour for daily prayer, time to listen to a podcast or two and a chance to plan out my day by prioritizing projects and tasks. I feel accomplished after a walk and am better prepared to take on the day. If you are trying to add more physical activity into your day, don't underestimate the power of walking to support your physical and mental health. Your body, mind and soul will thank you for the time you spend putting one foot in front of the other with a walking routine. IG

MEGAN RYAN is a native Texan, lover of flowers, plants and gardening and always planning for an upcoming travel adventure. For more than 22 years, Megan has lived with common variable immune deficiency. She's taken her weekly treatments on the road to more than 20 countries and four continents so far.

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Preparing Your Child for Infusions

By Jessica Leigh Johnson

IF YOU'RE RAISING a child with a primary immune deficiency (PI), it's very likely that immune globulin (IG) therapy is part of your child's treatment plan. Whether infusions are administered intravenously (IVIG) every month in a clinical setting, or through weekly subcutaneous infusions (SCIG) performed at home, IG therapy is the standard treatment for most PI diagnoses. For a parent of a newly diagnosed child, this can be a very overwhelming prospect. It's difficult enough to encourage a child to sit calmly and endure the sting of vaccinations that only need to be given once every few years, but a needle poke every month or even every week? If this is to become a regular occurrence, how do parents calm their children's nerves and dispel their fears in order to make the infusion experience more relaxing for them and less stressful for parents?[1]

My two youngest sons have had the benefit of being diagnosed with PI almost since birth. Because of that, they've been receiving SCIG infusions since they were 6 weeks old. I didn't have to do anything to prepare them other than applying numbing cream for a while, sticking the needle in and holding them until they stopped crying, which usually didn't take long. They were so young; they didn't know there was any reason to be scared of needles. The infusions quickly became part of their weekly routine, and it was never a problem.

For older children whose experience with needles and pokes has been limited, the idea of something sharp going into a vein, or even the idea of seeing blood, can be scary and unsettling.[1] SCIG does not involve veins, and it uses much smaller needles, but they still bleed from time to time, and no amount of numbing cream can eliminate 100 percent of the pain of the needle insertion or the feeling of tightness or burning from the medicine as it fills the nearby tissue.

How to Prepare Children for Infusions

Because IG infusions are necessary for those with PI to stay healthy, kids with PI have to accept them as part of life. It's not easy for parents if their children fight them before every infusion or struggle during them. The more comfortable children become with their regular infusions, the easier the treatments will become for everyone.[2] Here are a few things parents can do to help prepare their children for their infusion:

1) *Explain what to expect.* When it comes to younger children, parents should talk through what is about to happen in simple, age-appropriate language and answer any questions children may have. This can help ease their worries before the infusion.[2] To help children visualize what is about to happen, explain the process in a series of easy-to-understand steps. Say things like, "First we'll pull off the sticker, and next we'll wipe the numbing cream off with an alcohol pad."[2] Also, give an approximate length of time that the infusion will last. Knowing there is an endpoint in the not-so-distant future can be comforting to children.[2]

Another way to help kids understand what is about to happen is through play therapy, where a doll, stuffed animal or puppet is used to act out what will happen during an infusion session.[2] Providing children with a toy syringe from a play doctor kit (or even a real syringe with the needle removed) to use on the doll can help them become more familiar with the instruments and devices being used during their infusion.[2]

2) *Provide distractions.* Many hospitals employ child and family life specialists whose job is to make medical procedures easier for kids by providing distractions. They may hand children an iPad and get them involved in a game during a blood draw or provide them with a sensory toy to take their mind off of what the nurse is doing beside them. A distraction could be as simple as a TV show or movie that the children enjoy. This is especially easy to do when infusing at home, where children have access to everything they need to be comfortable — a favorite stuffed animal, blanket, book or even a comfort food. If the infusions are performed in a hospital setting, parents should consider bringing some of these comfort items along.[1] Even if children are not overly anxious, the infusion process can take time, and bored children may need these items in order to pass the time.[1]

3) *Practice relaxation techniques.* Parents can help their children stay calm before an infusion by incorporating relaxation techniques such as deep breathing or visualization.[1] Have children take a few deep breaths before the needle is inserted, or have them find a spot somewhere in the room to focus on rather than the needle site, especially if it makes them uncomfortable to watch.[1] One idea is to have children look into their parent's eyes while the parent holds their hand or strokes their head. This takes the attention off of the spot where the needle is going in. Using relaxation strategies can also help parents, who may sometimes find it difficult to watch their children being poked. Parents who are nervous about the infusions need to be careful not to project their own fears or anxieties onto their children.[2] Children are perceptive and can pick up on how their parents feel. Since our facial expressions can give a lot away, try smiling while the needle goes in rather than cringing or wincing.

4) *Reward children.* Kids with PI or other chronic illnesses face quite a bit of adversity. Getting regular infusions is a big deal, and the first one can be especially challenging. When children face their fears and do something they know might be painful, it is perfectly OK to reward them.[1] A reward doesn't have to be a big-ticket item. It could be a hug after their infusion, or some encouraging words. Or, parents could promise children a visit to a nearby park or a trip through the drive-through of their favorite restaurant.[1] At our local hospital, kids get a token whenever they go to the lab. After their blood is drawn, kids put their tokens into a vending machine and get a small toy. When administering subcutaneous infusions at home, sometimes the opportunity to sit and play video games or watch a movie is reward enough. It's important not to think of the reward as a bribe, but as an incentive for good behavior.[1]

Some children, especially those with PI, need infusions to stay healthy. It's simply a fact of life. They'll do it often enough that they will eventually get used to it, but the first few times can be scary and even painful, and it's understandable for them to be apprehensive. By adopting a few simple strategies, both parents and children will become comfortable and confident with the infusion process.[3] IG

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1. Done. Infusion Therapy: Parents, Prepare Your Child. Accessed at www.itsdone.marketing/article/infusion-therapy-parents-prepare-your-child.
2. Pediatric Rheumatology Consultants. First Time Infusion Tips, 2020. Accessed at www.pedirheumtx.com/2022/02/09/first-time-infusion-tips.
3. Thurrott, S. Subcutaneous Injections for Children: What Parents Should Know. Banner Health, May 21, 2024. Accessed at www.bannerhealth.com/healthcareblog/teach-me/subcutaneous-injections-for-children.

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

Gear for Parenting Medically Complex Kids

By Rachel Maier, MS

RAISING KIDS is hard. Raising medically complex kids is harder. Activities of daily life that are usually straightforward such as introducing solid foods, playing on the floor or even sending them to school become far more challenging the moment these kids receive a diagnosis. Whether it's a primary immune deficiency, neurological disorder, cancer or any number of rare diseases that require constant care at home, children with special health needs aren't one-size-fits-all, and as such, the gear they need isn't that way either. Knowing what you need and where to start looking can be daunting!

A Tricky Balance

Children with medical complexity are a subset of children and youth with special healthcare needs. They have multiple significant chronic health problems that often come with functional limitations, and many of them may need medical devices to help them function day-to-day. Constant caretaking puts even more demands on parents' time and energy because they find themselves playing a dual role of both nurse and parent.

A 2020 qualitative study exploring the challenges experienced by families caring for this subset of children found that "parents have substantial responsibilities, including performing medical procedures, managing emergencies (sometimes life-threatening), coordinating care and advocating for their child."[2] They balance the daily demands of providing routine at-home medical treatments, administering medicine and coordinating medical appointments with nurturing the parent/child relationship, letting their kids be kids and maintaining as normal a life as possible, and it's tough. "Their responsibilities have an enormous impact on the family: Going out of the home becomes a challenge, there are constant constraints on time, parents are sleep-deprived and there are wider impacts on siblings," the authors of the study wrote.[2]

Support for Parents

Gear is important, but sometimes support is more important than a gadget or gizmo that has functional value. After all, navigating a complex medical journey is a marathon, not a sprint, and the gear needed to get through changes with new legs of the journey. No matter the parenting season, finding encouragement and connection is an important part of staying the course and doing it well. Books such as *The Other Side of Special* and *Embracing This Special Life: Learning to Flourish as a Mother of a Child with Special Needs*; podcasts such as The Special Needs Mom and Take Heart Special Moms; and online communities such as Parent to Parent USA and Federation for Children with Special Needs are all great resources designed to help parents cope, connect and face each new day with determination and grace.

Innovative Gear Available Online

But of course, innovative ideas for products that solve a problem or simplify life are a godsend. There are several places to look online: Some of my favorite online sources for general adaptive and assistive gear include the Adaptive Mall, Spoonie Threads and Mighty Well. But there are other unique products out there, too, that are harder to find on general online retailers. Things like the Safety Sleeper and Cubby Beds (adaptive, enclosed bed solutions), TalkTools Sensi (a sensory oral therapy tool designed for speech and feeding therapy), Born Abel Books (books that represent medically complex kids in picture books) and StickyJ Medical (kids' engraved medical ID bracelets) are just a small sample of these creative solutions. Check out the Shopping Guide for more items designed to make life with complex medical kids a little bit easier for everyone. IG

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1. American Academy of Pediatrics. Children with Medical Complexity Overview. Accessed at www.aap.org/en/patient-care/children-with-medical-complexity.
2. Page, BF, Hinton, L, Harrop, E, et al. The Challenges of Caring for Children Who Require Complex Medical Care at Home: "The Go-Between for Everyone Is the Parent and as the Parent, That's an Awful Lot of Responsibility." *Health Expectations*, 2020 Oct;23(5):1144-1154. Accessed at pmc.ncbi.nlm.nih.gov/articles/PMC7696130.

RACHEL MAIER, MS, is the associate editor of *IG Living* magazine.

Pediatric Medical Planner

Medical planners are an absolute must when managing the care of a medically complex child. This pediatric medical planner is available as an instant download that can be stored on your favorite device or printed and stored in a 3-ring binder. From diagnosis summaries and medication lists and schedules to appointment trackers and IV care information, this template includes more than 50 customizable pages that make keeping your child's essential medical information and records up-to-date, easy and totally doable. *$7.50; (binder not included); www.etsy.com/listing/1728766676*

TalkTools' Itsy and Bitsy Feeding Collections

TalkTools' Itsy and Bitsy collections are designed to make mealtime much easier. Tested by speech-language pathologist and occupational therapist teams and their clients, the plates, lids, spoons and sporks are not only super cute, they're also ergonomic and appropriately sized to move your child along the developmental progression of feeding, helping them unlock their greatest potential in self-feeding. *Starts at $5.99; talktools.com/collections/feeding-tools*

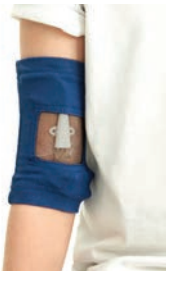
Shopping Guide for Gear for Medically Complex Kids

Puffaluffs

Puffaluffs are large, comforting stuffed animals (unicorns, giraffes, elephants, cows) designed to attach securely to the top and back of an IV pole. They feature an innovative center opening, allowing medical equipment to be discreetly integrated through the animal's body, reducing the visibility of wires and devices. This thoughtful design shifts focus from the intimidating equipment to the friendly presence of the Puffaluff, helping to ease anxiety and create a more soothing environment. *$119.24; puffaluffs.com/shop*

Adaptive Onsies

For babies with feeding tubes, PICC lines or ports, adaptive clothing prioritizes easy access (and saves parent sanity). Whereas these pieces used to be clunky and awkward, now they are intuitive and adorable. This bodysuit is designed for every parent who wants tummy access without having to remove their kiddo's clothes. The zipper makes accessing devices super easy while keeping your child dressed. Ideal for hospital and treatment visits, convenient for everyday use. *$20; spooniethreads.com/collections/kids-baby-all*



Care + Wear Kids Ultra Grip PICC Line Covers

Keep your child comfortable and secure without worrying about slippage at home or on the go with the Kid's Ultra Grip PICC Line Cover. The transparent mesh window provides breathability and visual access to monitor the insertion site for infection; a silicone band keeps cover in place for worry-free wear; and the antimicrobial fabric keeps the PICC comfortably in place. *$32; www.careandwear.com/products/kids-ultra-grip-picc-line-cover*

Medi Teddy IV Covers

When Ella, who has idiopathic thrombocytopenic purpura, was 13 years old, she created an invention that makes the hospital less scary for kids. Medi Teddy IV covers are made from BPA-free, latex-free, FDA grade silicone (front) and plastic (rear) that live on the IV pole and wait with an encouraging smile to greet each new patient. Each of the two cover versions is 11 inches tall, six inches wide, weighs .9 lbs and hides any size IV bag. *$39.99; ivcomfortsolutions.com*

Embracing This Special Life: Learning to Flourish as a Mother of a Child with Special Needs

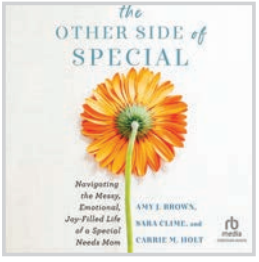
Author: Jenn Soehnlin
Publisher: Independently published

As the mother of two boys with special needs, Jenn Soehnlin knows the struggles of juggling the appointments, stress, grief and loneliness that accompany the special needs parenting journey. She seeks to encourage other mothers with scriptural truths to provide a biblical view on special needs parenting; free them from unhealthy expectations, worries and emotions; and enable them to embrace the life and blessings that God has for them. This book is intended to help guide parents toward hope, spiritual victory and renewed purpose in their own special needs parenting journey.

The Other Side of Special: Navigating the Messy, Emotional, Joy-Filled Life of a Special Needs Mom

Authors: Amy J. Brown, Sara Clime and Carrie M. Holt
Publisher: Revell

In *The Other Side of Special*, three mothers raising children with physical, medical, mental and emotional special needs have joined forces and pooled their experience and expertise to provide this resource. They take a deep dive into the most common emotions felt by special needs moms. They acknowledge the hard things as those who have been there. They celebrate the unique joys of being a special needs mom. And they offer encouragement for the journey, remaining realistic about the challenges special needs moms will continue to face.



New and Useful Reading

How I Became a Butterfly: Creative Tools for Coping with Life and Chronic Illness

Author: Amanda McKinlay
Publisher: Self-published

How I Became a Butterfly is a guidebook to the parts creativity and spirituality can play in managing the challenges of living with a chronic illness, or just the challenges of life in general. With a mix of personal journey and practical, holistic tools, including meditation, crystals and Angel work, this book is designed to help readers find and spread their wings so they can have happier journeys along their life paths.

Everything Is Not Peachey: One Woman's Pursuit of Happiness in the Face of Chronic Illness

Author: Lisa Peachey
Publisher: 5D Garden LLC

Everything Is Not Peachey takes readers on a profound journey through the author's battle with chronic illness, offering an unflinching glimpse into the realities of a condition that transforms every aspect of life. This book transcends mere personal narrative; it serves as a guiding light for anyone grappling with the complexities of chronic illness, whether personally affected or supporting a loved one.

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* Produced by IG Living magazine, written for patients treated with immune globulin therapy and their caregivers.

Abbie Cornett, MBA
IG Living Patient Advocate

Ataxia Telangiectasia (A-T)

Websites

- A-T Children's Project: www.atcp.org

Chronic Inflammatory Demyelinating-Polyneuropathy (CIDP)

Websites

- GBS/CIDP Foundation International: www.gbs-cidp.org

Evans Syndrome

Online Peer Support

- Rare Connect Evans Syndrome Community Group: www.rareconnect.org/en/community/evans-syndrome/faqs

Guillain-Barré Syndrome (GBS)

Websites

- GBS/CIDP Foundation International: www.gbs-cidp.org
- The Foundation for Peripheral Neuropathy: www.foundationforpn.com

Online Peer Support

- GBS Support Group: www.gaincharity.org.uk
- GBS/CIDP Foundation International Community Forums: forum.gbs-cidp.org

Immune Thrombocytopenia (ITP)

Websites

- ITP Support Association – UK: www.itpsupport.org.uk
- Platelet Disorder Support Association: www.pdsa.org

Kawasaki Disease

Websites

- American Heart Association: www.heart.org/en/health-topics/kawasaki-disease
- American Academy of Family Physicians: www.aafp.org/afp/2006/1001/p1141.html
- Kawasaki Disease Foundation: www.kdfoundation.org
- KidsHealth: www.kidshealth.org/parent/medical/heart/kawasaki.html

Mitochondrial Disease

Websites

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

Multifocal Motor Neuropathy (MMN)

Websites

- The Foundation for Peripheral Neuropathy: www.foundationforpn.com

Multiple Sclerosis (MS)

Websites

- Multiple Sclerosis Association of America: www.mymsaa.org
- Multiple Sclerosis Foundation: www.msfocus.org
- National Multiple Sclerosis Society: www.nationalmssociety.org

Online Peer Support

- Friends with MS: www.FriendsWithMS.com
- MSWorld's Chat and Message Board: www.msworld.org
- Overcoming Multiple Sclerosis: www.overcomingms.org/community

Myasthenia Gravis (MG)

Websites and Chat Rooms

- Myasthenia Gravis Foundation of America (MGFA): www.myasthenia.org
- Myasthenia Gravis Association: mgakc.org

Online Peer Support

- Genetic Alliance: www.geneticalliance.org

Myositis

Websites

- The Myositis Association: www.myositis.org
- International Myositis Assessment and Clinical Studies Group: www.niehs.nih.gov/research/resources/imacs/index.cfm

Online Peer Support

- Juvenile Myositis Family Support Network: www.curejm.org/fsn/index.php
- The Cure JM Foundation: www.curejm.org
- Myositis Association Support Group: www.myositis.org/patient-support/support-groups
- Myositis Support Group – UK: www.myositis.org.uk

Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)

Websites

- PANS/PANDAS UK: www.panspandasuk.org
- PANDAS Network: www.pandasnetwork.org
- PANDAS Physician Network Family Resources: www.pandasppn.org/parent-information
- National Institute of Mental Health: www.nimh.nih.gov/health/publications/pandas/index.shtml

Pemphigus and Pemphigoid

Websites

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

Peripheral Neuropathy (PN)

Websites

- Neuropathy Action Foundation: www.neuropathyaction.org
- Western Neuropathy Association: www.pnhelp.org
- Neuropathy Alliance of Texas: www.neuropathyalliancetx.org
- The Foundation for Peripheral Neuropathy: www.foundationforpn.com

Primary Immune Deficiency Disease (PI)

Websites

- Immune Deficiency Foundation: www.primaryimmune.org
- Jeffrey Modell Foundation: www.info4pi.org
- The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/Pages/index.aspx
- American Academy of Allergy, Asthma & Immunology: www.aaaai.org
- International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: www.ipopi.org
- Rainbow Allergy-Immunology: www.uhhospitals.org/rainbow/services/pediatric-allergy-and-immunology

Online Peer Support

- IDF Friends: www.idffriends.com
- Jeffrey Modell Foundation Facebook Page: www.facebook.com/JMFworld
- IDF Peer Support Program: www.primaryimmune.org/idf-peer-support-program

Scleroderma

Websites

- Scleroderma Foundation: www.scleroderma.org
- Scleroderma Research Foundation: www.srfcure.org
- Johns Hopkins Scleroderma Center: www.hopkinsscleroderma.org

Online Peer Support

- Scleroderma Support Forum: www.curezone.com/forums/f.asp?=404

Stiff Person Syndrome (SPS)

Websites

- American Autoimmune Related Diseases Association Inc.: www.aarda.org
- Genetic Alliance: www.geneticalliance.org
- Living with Stiff Person Syndrome (personal account): www.livingwithsps.com
- The Stiff Person Syndrome Research Foundation: stiffperson.org

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