Immune Globulin
Choosing the Product, Delivery Method and Site of Infusion

How Aging Affects Chronic Illness

Resources and Support for the Caregiver

Purchasing Life Insurance with a Chronic Illness

Understanding Small Fiber Neuropathy
with IG Living!

On Facebook
Find timely and relevant information posted daily, providing a venue for connecting with others in the IG community.

On the Go
The IG Living App allows you to connect Anytime, Anywhere! And it is FREE!
Up Front

5 Editorial
Considerations in IG Therapy
By Ronale Tucker Rhodes, MS

6 Abbie’s Corner
Regaining Control Over Healthcare and Improving Outcomes
By Abbie Cornett

7 Faces of IG
From our Facebook page
By Abbie Cornett

Departments

8 Ask the Experts
Healthcare professionals’ responses to patient questions

9 Immunology 101
DiGeorge Syndrome: Series Summary
By Terry O. Harville, MD, PhD

10 Therapeutic Helpline
How to Get Rid of Distressing Thoughts
By Erika Lawrence, PhD, LCP

12 Clinical Brief
Vamorolone: A Corticosteroid Replacement Under Study
By Michelle Greer, RN

14 In the News
Research, science, product and insurance updates

Columns

40 Let’s Talk!—
Christina Mangurian, MD, MAS
By Trudie Mitschang

42 Patient Perspective —
Well, I Wasn’t Expecting That to Happen
By Stacy Oliver

43 Life as a 20-Something —
Developing New or Unexpected Symptoms
By Ilana Jacqueline

44 Parenting — Helping a Child Control Anger and Aggressive Behavior
By Jessica Leigh Johnson

Features

16 Immune Globulin: Each Product Is Unique
By Ronale Tucker Rhodes, MS

18 Immune Globulin: Choosing a Delivery Method and Site Location
By Abbie Cornett

24 How Aging Affects Chronic Illness
By Jim Trageser

28 Caring for the Caregiver
By Trudie Mitschang

32 Purchasing Life Insurance with a Chronic Illness
By Ronale Tucker Rhodes, MS

36 Understanding Small Fiber Neuropathy
By Jim Trageser

Sources

46 Product Guide
Just Breathe: Asthma Relief Is Possible
By Heather Bremner Claverie

48 Book Corner
New and useful reading

50 Resource Center
Community foundations, associations, forums and other resources

Advertising in IG Living
IG Living Magazine is read by 30,000 subscribers who are patients that depend upon immune globulin products and their healthcare providers. For information about advertising in IG Living, download a media kit at igliving.com/Advertise.aspx. Or contact advertising@igliving.com.

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

IG Living, (ISSN 1949-4548), published bimonthly, is a community service provided by FFF Enterprises, 44000 Winchester Road, Temecula, CA 92590, (800) 843-7477 x1362, fax (951) 699-9655.

Subscriptions to IG Living are free, and readers may subscribe at IGLiving.com or by calling (800) 843-7477 x1351.

The opinions expressed in IG Living are those of the authors alone and do not represent the opinions, policies or positions of FFF Enterprises, the Board of Directors, the IG Living Advisory Board or editorial staff. This material is provided for general information only. FFF Enterprises does not give medical advice or engage in the practice of medicine. FFF Enterprises under no circumstances recommends any particular treatment for any individual and in all cases recommends individuals consult with a physician before pursuing any course of treatment.

IG Living accepts article submissions. Submit manuscripts in MS Word format, or submit a query letter that covers the idea in a brief paragraph and how it will be presented, to editor@igliving.com. IG Living retains the right to edit submissions. The contents of each submission and their accuracy are the responsibility of the author(s) and must be original work that has not been, nor will be, published elsewhere, without the written permission of IG Living. A copyright agreement attesting to this, and transferring copyright to FFF Enterprises will be required. Acceptance of advertising for products and services in IG Living in no way constitutes endorsement by FFF Enterprises. ©2019 FFF Enterprises Inc.
SINCE THE first use of immune globulin (IG) therapy more than 70 years ago, the number of IG products, their delivery methods and the choice of infusion sites has continued to evolve. Whether patients are new to IG therapy or have been treated for years, it is extremely important they arm themselves with the knowledge to make decisions concerning these components of their care.

We begin our feature section with a discussion of IG products in our article “Immune Globulin: Each Product Is Unique” (p.16). As the title implies, IG products are not pharmaceutically equivalent. Differences in a number of elements making up these products, including stabilizers, osmolality, IgA content and concentration, will factor into whether patients will tolerate one product better than another. In addition, patients’ clinical condition will dictate dosing and premedication. The key is to find the product that each patient tolerates best, and when a product is not tolerated, switching to another should be tried.

As the number of IG products has grown, so too have the choices of delivery methods and sites of care for infusion. As we explain in our article “IG Treatment: Choosing a Delivery Method and Site Location” (p.18), patients can now elect to receive IG therapy either intravenously (IV) or subcutaneously (SC), and there are advantages and disadvantages to each. IVIG can be administered in a clinical setting or in the home, and in either setting, the benefit of additional medical supervision and more interaction with medical staff is an important consideration for those with comorbidities. On the other hand, SCIG is always infused in the home, and it provides more quality-of-life advantages. What is important is that patients choose the method and site that best suits their needs.

It’s inevitable that many patients treated with IG for their chronic illness may experience the effects of aging. In our article “Exploring the Effects of Aging on Chronic Illness” (p.24), we discuss a relatively new field of study known as geroscience. Not only is aging the leading risk factor for most chronic diseases, but nearly all primary immunodeficiency diseases (PIDs) are exacerbated by the aging process, which makes this area of research extremely important for our readers. As we discuss, advances are slowly being made in geroscience to improve the effectiveness of care for older adults suffering from chronic conditions such as PI and, thus, prolong their health span.

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.
Regaining Control Over Healthcare and Improving Outcomes

By Abbie Cornett

HEALTHCARE COSTS in the U.S. far surpass any other developed country, both in dollars and as a percentage of gross domestic product (GDP). In 2016, the Centers for Disease Control and Prevention reported Americans spent a staggering $10,348 per capita on healthcare expenditures, representing 17.9 percent of GDP.¹ But, increased spending doesn’t necessarily translate to better healthcare,² particularly for those with chronic diseases. In fact, Americans have less of a chance of living to 80 years old than people in many other developed countries. Further, Americans rank last compared with citizens of 10 other wealthy countries when it comes to managing emotional distress, struggling to pay for care and skipping doctor visits.³

One major reason healthcare in the U.S. is so expensive is because the system is fundamentally different compared with other competitive markets.⁴ In the U.S., healthcare sits outside of the normal marketplace, whereas in other countries, it’s at the center of it. This is unfortunate for U.S. patients, as consumers need to be involved in the choices made for their care, rather than treated as bystanders. For instance, when insurance providers or the government pay for medical procedures, patients usually have no idea how much their care costs or what they will owe until after they receive care. In other countries, consumers are aware of what they are paying for upfront. Further, in the U.S., patients regularly have to fight payers for medications that will best treat their illness. This lack of control over their healthcare is part of the reason many patients have poorer outcomes.

So, how can people regain control of their healthcare and improve their outcomes, while reducing costs?

The first step is for patients to increase their health literacy. Health literacy is defined as the capacity to seek, understand and act on health information.⁵ In other words, individuals need to learn everything they can about their disease so they can be their own best advocates. A great way to find up-to-date information and support is to become involved with an advocacy group specific to their disease.⁶ Many such chronic illness groups can be found on the Internet (see Advocacy Organizations). Also, understanding what is covered by an insurance policy can save patients a lot of stress and money. To get the most from a policy, individuals need to read their explanation of benefits (EOB), which will give them a good idea of what is covered. An EOB will include information about the deductible and what services to which it can apply, as well as what the copay is. It will also show what the out-of-pocket expenses will be. Other important information included in the EOB is whether a referral to see a specialist is required and if there is a covered visit limitation. Further, some plans limit individuals to a certain network of providers.⁷

Additionally, patients can improve their outcomes and save money by keeping copies of their medical records. These records will help patients better understand the specifics of their condition and more actively participate in the monitoring and managing of their healthcare such as by reducing redundant testing.⁸

Healthcare in the U.S. has many challenges, but patients can be part of the solution by being proactive and taking part in managing their care.

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

Advocacy Organizations:

A Starting Point

- Immune Deficiency Foundation: primaryimmune.org
- GBS/CIDP Foundation International: www.gbs-cidp.org
- The Myositis Association: www.myositis.org
- Neuropathy Action Foundation: neuropathyaction.org
- PANDAS Network: pandasnetwork.org

References

Join the conversation! Connect with other immune globulin patients through IG Living’s Facebook page at www.facebook.com/IGLivingMagazine. See our daily posts of interesting articles and facts, as well as thought-provoking questions that you can chime in on. Following are some snapshots of what’s being discussed.

Have you ever suffered from side effects?

[Intravenous immune globulin] and Haegarda caused me to have hyperprolinemia and glutamate toxicity. I seriously thought I was going to die. I could barely walk 10 feet without shortness of breath, utter exhaustion, muscle and joint aches and couldn’t remember stuff. A mere two weeks without [the medicines], I started feeling better, but I still don’t have muscle tone or strength, and fatigue and memory issues persist. I’m now on [subcutaneous immune globulin] and Cinryze and am doing better. Please advise doctors to test amino acid levels when on subcutaneous immune globulin products!

— Renee WL

Have you had trouble losing weight?

Yes. Trouble losing weight is an issue for me. I also have hypothyroidism and nasty high blood pressure. So, weight is always on my mind.

— Peggy Z

Yes, every winter, since I usually lose 20 pounds to 30 pounds when I go to my mom’s in Tucson, Ariz., and then regain some back home in Illinois. But, this year, I did not lose anything, and now I am up again due to weight gain from Gabapentin I am [taking] for shingles nerve pain. I weigh 60 pounds more than I should right now. I actually have gained weight in the last four years since starting Gammagard.

— Rachel D

Has your illness affected your mental health?

Yes. For years, I was in denial as to what I really was unable to do and had to come to terms with the “new me.” And, it's difficult when you look good on the outside [and your] autoimmune disease is destroying [you] on the inside. In general, people don’t see what is happening. The help I received was the best thing I did for myself. I had the best therapist who has now retired. Fortunately, my primary care physician gets me through tough times.

— Judy S

It does sometimes. Knowing that stress doesn’t help me with my disease, I try to always remember that and keep a positive attitude.

— Jenny G

It’s horrific. I still work and drag myself around. It’s getting harder and harder. My resentment is making me eat too much and drink too much. And, resenting people living a normal life is not my nature at all. I hate it and fight it every day. But some days, it wins.

— Larie D
Abbie » I spoke with one of our experts who believes, ultimately with education, there will be growth in utilization of this newer molecular technique. However, at this time, cultures are still the gold standard due to long-term usage with interpretable results and little controversy. While in many ways the genetic/molecular methods are potentially better, they still have issues. Contamination is much more likely, and therefore, specific controls and quality assurance methods must be in place. Further, the molecular techniques are much more sensitive. Sometimes, an apparent pathogen is found that may not be clinically relevant, so understanding how to interpret the results is very important.

Question » Why Aren’t More Doctors Using DNA Next-Generation Sequencing to Control Infections?

We’re told “culture, culture, culture” to control infections, but the traditional cultures are only 50 percent accurate, and they miss a lot of things. The new DNA Next-Generation Sequencing cultures are covered by Medicare and are readily available. Some doctors are using them successfully, and finding them helpful with recurrent and complicated infections. Why aren’t more doctors using them?

Abbie » Swelling and site irritation are the most common side effects of SCIG, and the swelling can last between 24 hours and 72 hours after the infusion. Hydration is an important factor to help with side effects. Our expert recommends a minimum of 64 ounces of clear fluids (water or Powerade) the day before, day of and day after SCIG infusions. He also recommends taking Tylenol and Benadryl pre-infusion to help with side effects.

In addition, he suggests if the site on your belly is a problem, your thighs might be an option depending on the amount of subcutaneous tissue you have. In fact, a lot of patients prefer the thighs over the belly. By infusing in the thighs, you have four sites (two in each leg). You just need to make sure to rotate the sites and move about one inch away from the previous stick each infusion. If you are still having problems at the site, you should consult your doctor because it’s possible other medications may be interacting with your SCIG.

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

ABBIE CORNETT is the patient advocate for IG Living magazine.
DiGeorge Syndrome: Series Summary

By Terry O. Harville, MD, PhD

Digeorge Syndrome (DGS) is a multisystem disorder resulting from the consequences of improper timing of the sequence of events during early phases of embryonic development, typically before pregnancy is recognized. Fetal alcohol syndrome produces many of the same features as DGS since it also disrupts early embryonic development, but DGS is typically a result of mutations affecting chromosome 22. DGS can be divided into complete DGS (CDGS) or partial DGS (PDGS) based on the immune system status. In CDGS, there is complete lack of the thymus and lack of T lymphocyte development. In PDGS, there is sufficient thymic tissue to allow for development of the T lymphocyte repertoire, even though it may take a few years to develop. In CDGS, donor thymic transplantation can allow for development of a T lymphocyte repertoire, and essentially normalization of immunity. Therefore, an immune disorder that was lethal in the past can now be managed with good immunologic outcomes.

Other structures of the body forming during early embryonic development can also be affected. Perhaps most notably is the heart. Indeed, when Angelo DiGeorge, MD, initially described this condition, the most important feature involved heart malformations. More severe cardiac malformations may arise when the disruption of the timing of development occurs earlier. This means these individuals may not have significant thymic development issues (since this completes late in the first trimester), resulting in PDGS or, in some cases, normal T lymphocyte repertoire development at birth. Due to improvement in pediatric cardiac surgical techniques, most heart abnormalities in DGS can be repaired or managed.

Because the parathyroid glands develop along with the thymus, abnormal parathyroid development occurs in parallel with abnormal thymus development, which can result in low serum calcium levels. Once recognized, this can be treated with parathyroid hormone replacement and calcium. Hypocalcemia was another of the original features of DGS described by Dr. DiGeorge.

Furthermore, other tissues and organs can be affected. Vertebral bodies and ribs may have some abnormal development. Also, kidney development may be affected. Due to disruption of the development of the esophagus, gastro-esophageal reflux is a major problem in most infants with DGS, requiring medical management and frequently requiring surgical intervention.

Development of the head region can also be affected, especially when the disruption in timing occurs earlier in the fetus. Many infants with DGS have a head and face described as “elfin-like,” with a somewhat “triangular” shape. The eyes may be slightly widely spaced. The lower jaw (mandible) is typically small, and the chin may be small and somewhat pointed, accentuating the triangular shape of the face. The ears may be somewhat low and rotated backward. The top of the ears may be slightly pointed and “notched” in appearance. These features do not create any particular medical issues. However, the Eustachian tubes can be small and tortuous, resulting in chronic ear infections even if the immune system is not adversely affected. Additionally, cleft lip and cleft palate can occur, but can usually be surgically repaired.

Central nervous system-related problems have been under-recognized in DGS. It is now clear that brain development is also disrupted. Unfortunately, there are no surgical or complete medical repairs for the brain. Thus, early intervention with neurocognitive evaluation and treatment is critical in infants and young children with DGS. Recognition that psychotic illnesses can develop as the patient grows older is also important so early psychiatric evaluation and treatment can be implemented.

DGS typically requires a team of physicians and services, initially including cardiologists, cardiothoracic surgeons, endocrinologists and immunologists to manage the condition early on. Occupational therapists, physical therapists, nutritionists and speech therapists are also very important in delivering optimal care for patients from the onset. In addition to medical and surgical driving early intervention, it must not be forgotten that neurologists and psychiatrists have a critical role in completing the care team for DGS patients, especially as they grow older.

In the next issue, we will begin a new topic of discussion.

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.
“I CAN’T DO anything.” “Nothing will ever change.” “No one understands what I’m going through.” We’ve all had thoughts like these at times, and they can leave us feeling depressed, anxious, angry or all of the above.

Some of us have thoughts about ourselves that really get us down, causing us to crawl back into bed and pull the blankets over our heads, or lash out at others. And, many of us try to control or change these thoughts by arguing with them or challenging them, replacing them with positive thoughts or even trying to get rid of them by distracting ourselves with TV, music, drinking or one of a million other ways.

Do these methods work? They may get these thoughts out of our heads for a while, but here’s the problem: They never go away forever. They always come back. And, many of us spend so much time and energy trying to control or get rid of these thoughts that we run out of time to focus on the things that bring us joy.

So, what is the alternative? Let those thoughts be there; they are going to show up anyway. Spend your precious time and energy on the things that really matter instead of fighting with your thoughts or running from them. This doesn’t mean you agree with the thoughts, you like them or you are happy they are there. It just means acknowledging they are there and letting them be. It means noticing the distressing thoughts when they pop into your head, and not giving them the power to control you.

Thoughts are just ideas our minds create. We can let them be there without allowing them to influence our behavior. For example, consider these thoughts: “I want to go meet my friend for coffee, but I’m too depressed.” “I want to go for a walk, but I’m too tired.” What do these two sentences have in common? The word “but,” which suggests only one part of each thought can be true at a time. In other words, I can’t meet my friend if I’m depressed, or I can’t go for a walk if I’m tired. However, in reality, both parts of each thought are true. We can feel depressed and want to meet a friend at the same time. We can be really tired and want to go for a walk at the same time. If you replace “but” with “and,” isn’t that more accurate? “I want to go meet my friend for coffee, and I’m too depressed.” “I want to go for a walk, and I’m too tired.”

Here’s the point: We can have the distressing thoughts and still choose our own behaviors, rather than letting the thoughts dictate what we do. You can have the thought that you’re depressed or tired and still meet a friend or go for a walk.

Some thoughts have such power over our lives that we find ourselves getting stuck to them and unable to move on. The trick is to learn to get unstuck. How? Changing “but” to “and” is just one of them. Another strategy is to visualize yourself relaxing by a stream with leaves floating down it. Put each unwanted thought on a leaf, and watch it float down the stream. The same thought may come up again and again. That’s common. Just keep putting each thought on a leaf, and let it float down the stream. The idea is you are noticing your thoughts and letting them float by, instead of trying to change or avoid them. (Googling “leaves on a stream” will turn up dozens of videos that take you through this exercise in detail.)

This strategy is called a defusion technique because it is meant to help you “de-fuse” or unstick yourself from distressing thoughts. There are many defusion techniques. Some involve using language, some employ activities like coloring and some concentrate on visualization. The trick is to find one that works well for you.

These strategies also take a lot of practice. They may not work the first time, and they aren’t the solution for everyone. However, if you’ve been struggling to get rid of or change distressing thoughts for a while, and they keep coming back, this might be worth trying.

The bottom line: The distressing, unwanted thoughts are already there. Unfortunately, they may always be there, and we can waste a lot of time and energy trying to control them. What we can manage is how we respond to the thoughts and how much we let them limit our lives. □

ERIKA LAWRENCE, PhD, LCP, is director of translational science at The Family Institute at Northwestern University, Evanston, Ill.
Making a Difference in Our Patients’ Lives...

so he can say “I do”  
so he can witness her first steps  
so he can cheer her on when she graduates  
so he can enjoy time with his grandchildren

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

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

©2019 Nufactor, Inc. is the specialty pharmacy subsidiary of FFF Enterprises, Inc., the nation’s most trusted distributor of plasma products, vaccines and other biopharmaceuticals.

Nufactor Specialty Pharmacy has earned the Joint Commission Gold Seal of Approval.

(800) 323-6832  
www.nufactor.com

nufactor.  
A SPECIALTY INFUSION COMPANY
Vamorolone: A Corticosteroid Replacement Under Study

By Michelle Greer, RN

CORTICOSTEROIDS, ALSO known as steroids, are the first line of treatment for a wide variety of diseases caused by inflammation, including autoimmune disorders such as neuropathies, multiple sclerosis, inflammatory bowel disease and arthritis. They were first discovered in September 1948 by Edward C. Kendall, MD, and Philip S. Hench, MD, after they injected a small amount of an experimental new drug, then named Compound E, in a 29-year-old woman hospitalized at the Mayo Clinic in Rochester, Minn., for severe rheumatoid arthritis that caused debilitating joint immobility. After two days and two additional injections, the woman’s symptoms disappeared, a result that stunned people throughout the world. In 1950, Drs. Hench and Kendall were awarded the Nobel Prize in Physiology and Medicine for their discovery.

Corticosteroids work by mimicking the effects of hormones in the body produced naturally in the adrenal glands, which are small glands that sit on top of the kidneys. When prescribed in doses that exceed the body’s usual levels, corticosteroids suppress inflammation. As such, corticosteroid use has increased substantially for arthritis and other inflammatory conditions. Yet, while they have clear benefits, the profound side effects associated with higher doses and prolonged use can cause more harm than good, and in patients with comorbidities, the side effects can be detrimental (see Table 1). Indeed, the higher the daily dose and the longer the duration of therapy, the greater the risk of side effects, which are caused by steroids’ subactivities that repress and/or activate other responses in the body.

Vamorolone: A New Steroid with Fewer Side Effects

Recently, a new medication was developed by ReversaGen BioPharma in an attempt to maintain the benefits of steroids while avoiding the negative side effects. Vamorolone is a dissociative steroid that chemically separates the aspects of efficacy (clinical benefit) from safety concerns (side effect profiles). To date, no other drug has this effect without the negative side effects, so vamorolone is frequently referred to as a “first-in-class” drug.

Originally called VBP15, vamorolone was studied for safety and tolerability of single and multiple doses of various increasing amounts in healthy adults. In the Phase I clinical trial, 86 healthy adult males were treated with single ascending doses (0.1-20.0 mg/kg) and multiple ascending doses (1.0-20.0 mg/kg/day for 14 days), with results showing vamorolone was well-tolerated at all dose levels. Pharmacokinetic and metabolism profiles were similar to prednisone, but biomarkers showed loss of side effects (bone fragility, metabolic disturbance, immune suppression) of traditional glucocorticoid drugs. Indeed, suppression of the adrenal axis was 10-fold less than prednisone.

Vamorolone and Duchenne Muscular Dystrophy (DMD)

Vamorolone is currently being studied as a treatment for DMD, a genetic neuromuscular disorder that is usually diagnosed around 3 years to 5 years of age and affects mainly boys. DMD is manifested by muscle weakness and wasting and results in a gradual decrease in the ability to walk by adolescence. Most patients die in the second or third decade of life due to cardiopulmonary complications. There is no cure for DMD, but corticosteroids are used in an attempt to improve muscle strength and function at the time of diagnosis and to prolong the ability to walk. Using vamorolone to treat DMD could...
be especially promising because bones are still developing in boys at diagnosis. And, where high doses and long-term use of current steroids can have a major negative impact on bone development, vamorolone could provide the benefits without this adverse effect.

To date, two Phase II studies have assessed the safety and tolerability of vamorolone in boys with DMD. The first, completed in November 2017, included 48 boys who were given a broad range of doses of vamorolone (0.25, 0.75, 2.0 and 6.0 mg/kg/day) for two weeks, followed by a two-week washout period. Findings showed vamorolone was safe and well-tolerated through the highest dose (6.0 mg/kg/day, which is about nine times the typical prednisone dose in DMD) and showed pharmacokinetics similar to prednisone. According to the lead author of the study, Laurie Conklin, MD, “Our use of blood biomarkers in this study was innovative. Based on blood testing, we were able to show that vamorolone has less potential for side effects than deflazacort and prednisone, but still shows strong anti-inflammatory activity.” The second, a 24-week extension study of the first that was completed in April 2018, examined using vamorolone in boys with DMD for more than two weeks to compare potential side effects compared with boys treated with other steroids. Two additional two-year extension studies are continuing to examine the safety and efficacy of vamorolone in boys with DMD and are comparing the side effect profile of vamorolone to that of currently available corticosteroid treatments.4

Vamorolone and Other Inflammatory Conditions

According to the researchers in the Phase I trial, “Vamorolone is being developed as a replacement for glucocorticoid treatment of Duchenne muscular dystrophy and other chronic inflammatory conditions. Vamorolone has shown efficacy similar or superior to prednisone in mouse models of muscular dystrophy, lung disease, inflammatory bowel disease and multiple sclerosis.” In addition, say the researchers, the potent immunosuppressive effects of glucocorticoids are considered an aspect of efficacy in certain clinical indications, including humoral autoimmunity disorders. But, since immunosuppression is important in treating various autoimmune conditions, vamorolone would need to be specifically studied in each of these conditions to determine its efficacy.3

Although vamorolone is currently being studied to treat DMD, it is not yet approved by the U.S. Food and Drug Administration for any indication in the U.S. However, there is a lot of optimism and excitement around this therapy for DMD. And, if approved, there will likely be additional research to assess vamorolone’s safety and efficacy for other conditions in which corticosteroids are the first line of treatment, especially when used in high doses and long-term.

MICHELLE GREER, RN, is senior vice president of sales for NuFACTOR Specialty Pharmacy.

References

<table>
<thead>
<tr>
<th>Side Effects of Corticosteroids</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Insomnia</td>
</tr>
<tr>
<td>• Infection</td>
</tr>
<tr>
<td>• Increased stomach acids leading to gastroesophageal reflux disease and gastrointestinal bleeding</td>
</tr>
<tr>
<td>• Increased hunger and/or thirst</td>
</tr>
<tr>
<td>• Nervousness, confusion and restlessness</td>
</tr>
<tr>
<td>• Mood swings and depression</td>
</tr>
<tr>
<td>• Headache</td>
</tr>
<tr>
<td>• Skin issues such as pimples, acne and thin/shiny skin</td>
</tr>
<tr>
<td>• Fluid retention, “moon face” and edematous feet/ankles</td>
</tr>
<tr>
<td>• Vision changes (glaucoma, cataracts)</td>
</tr>
<tr>
<td>• Changes in blood cell values and increased blood sugar</td>
</tr>
<tr>
<td>• Changes in heart rate and/or rhythm</td>
</tr>
<tr>
<td>• Confusion</td>
</tr>
<tr>
<td>• Difficulty breathing</td>
</tr>
<tr>
<td>• Facial hair growth in females (hirsutism)</td>
</tr>
<tr>
<td>• Changes in bone density leading to possible fractures</td>
</tr>
<tr>
<td>• Muscle pain, wasting and/or weakness</td>
</tr>
<tr>
<td>• Weight gain or loss</td>
</tr>
</tbody>
</table>

**Table 1. Side Effects of Corticosteroids**

**Table 2. Side Effects of Corticosteroids**

<table>
<thead>
<tr>
<th>Side Effects of Corticosteroids</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Insomnia</td>
</tr>
<tr>
<td>• Infection</td>
</tr>
<tr>
<td>• Increased stomach acids leading to gastroesophageal reflux disease and gastrointestinal bleeding</td>
</tr>
<tr>
<td>• Increased hunger and/or thirst</td>
</tr>
<tr>
<td>• Nervousness, confusion and restlessness</td>
</tr>
<tr>
<td>• Mood swings and depression</td>
</tr>
<tr>
<td>• Headache</td>
</tr>
<tr>
<td>• Skin issues such as pimples, acne and thin/shiny skin</td>
</tr>
<tr>
<td>• Fluid retention, “moon face” and edematous feet/ankles</td>
</tr>
<tr>
<td>• Vision changes (glaucoma, cataracts)</td>
</tr>
<tr>
<td>• Changes in blood cell values and increased blood sugar</td>
</tr>
<tr>
<td>• Changes in heart rate and/or rhythm</td>
</tr>
<tr>
<td>• Confusion</td>
</tr>
<tr>
<td>• Difficulty breathing</td>
</tr>
<tr>
<td>• Facial hair growth in females (hirsutism)</td>
</tr>
<tr>
<td>• Changes in bone density leading to possible fractures</td>
</tr>
<tr>
<td>• Muscle pain, wasting and/or weakness</td>
</tr>
<tr>
<td>• Weight gain or loss</td>
</tr>
</tbody>
</table>
Grants

**IDF Awarded $4 Million Grant for SCID Screening**

The U.S. Health Resources and Services Administration (HRSA) has awarded a $4 million grant to the Immune Deficiency Foundation (IDF) to design and implement an advanced screening and education program for people with severe combined immunodeficiency (SCID) in rural areas or underserved communities. The goal of the project, which will run from Aug. 1, 2018, through July 31, 2020, is to improve outcomes for infants with SCID detected through newborn screening by increasing awareness and knowledge about SCID, supporting state newborn screening programs, linking families (especially those living in medically underserved areas) to services and developing long-term follow-up strategies for infants identified through newborn screening. To develop and implement the program, IDF is partnering with the Association of Public Health Laboratories (APHL), the primary professional association responsible for supporting newborn screening, and RTI International, one of the world’s largest nonprofit research institutes.

Currently, 47 states have fully implemented newborn screening for SCID, which covers approximately 95 percent of births in the U.S. Yet, significant challenges remain for patients, families, clinicians and public health professionals, including communication between the screening community, healthcare providers and families; disparities in knowledge and care for patients with SCID in rural and underserved communities; and general awareness and knowledge about SCID and SCID newborn screening for all stakeholders. “While monumental advancements in testing and treatment of SCID have occurred over the past 40 years, critical gaps still do exist,” said John G. Boyle, president and CEO of IDF. “We are grateful to HRSA for its confidence in our ability to maximize and leverage the collective resources of IDF, APHL and RTI to have a transformational impact on improving the lives of people with SCID and helping to enhance the knowledge base of testing and treatment for these diseases.”


Research

**Experimental Tandem Lung-Bone Marrow Transplant Shows Promise as Treatment for PI**

A novel experimental tandem lung and bone marrow transplant has proved successful in seven patients with terminal lung disease who also had primary immunodeficiencies (PIs) or evidence of bone marrow failure. The seven patients were treated with the experimental protocol that involves bilateral lung transplant followed by bone marrow transplant from a cadaver lung donor.

The first procedure was performed on a teenage patient with a combined immune deficiency disorder in 2009 at Duke University Medical Center in Durham, N.C., who received bone marrow harvested from her lung donor four months after her bilateral lung transplant. Today, the bone marrow has fully engrafted and her immune system is normal. Four additional patients received the transplant protocol at the University of Pittsburgh Medical Center (UPMC), and a fifth has received the lung transplant and is scheduled to have her bone marrow transplant. Of these, three transplants were performed in patients with PIs and end-stage lung disease. These included a 14-year-old girl with IL-7R deficiency, a 39-year-old woman with common variable immunodeficiency and a 38-year-old male with severe combined immunodeficiency. Two of these patients successfully engrafted donor bone marrow. The teen has been off all immunosuppression therapy for more than a year. And, the male patient failed to engraft the donor bone marrow but was doing well 16 months post-lung transplant. A fourth 20-year-old male patient with interstitial lung disease and myelodysplastic syndrome with monosomy 7 and short telomeres had very advanced lung disease, bone marrow failure and myelodysplastic syndrome when referred to UPMC. After his lung transplant, he was discharged on room air and his lung function continues to improve. In July, he received a bone marrow transplant, and he is fully engrafted.

While, for now, the protocol will be used only in the sickest patients with end-stage lung disease and bone marrow irregularities who have no other treatment options, the researchers believe it could eventually be used in an even larger population of lung transplant patients to allow them to avoid lifelong immunosuppression.

Technology

IPOPI Launches App for Managing PI

The International Patient Organisation for Primary Immunodeficiencies (IPOPI) has launched PID Genius, the worldwide first mobile application for people with primary immunodeficiencies (PIs). PID Genius is a personal assistant for PI patients in search of an easy and dynamic way of keeping track of treatments, day-to-day symptoms, vaccinations, contacts and important documents. It also allows information to be stored in dashboards, facilitating discussion between patients and their medical providers. “We hope that with this mobile application, people living with [PIs] will see the management of their day-to-day life and treatment become simpler and easier to follow up, sparing them the trouble of managing with a big bulk of files,” said Jose Drabwell, a representative of IPOPI. The app is compatible with Android and Apple operating systems and can be downloaded at Google Play and the iOS App Store.

Medicines

FDA Approves Revcovi to Treat Pediatric and Adult ADA-SCID

The U.S. Food and Drug Administration (FDA) has approved Leadiant Biosciences’ Revcovi (elapegademase-lvlr) injection for the treatment of adenosine deaminase severe combined immune deficiency (ADA-SCID) in pediatric and adult patients. Revcovi is a PEGylated recombinant ADA (rADA) enzyme that eliminates the need to source ADA from animals and works by supplementing levels of ADA. Approval is based on results from two multicenter, open-label clinical trials that demonstrate Revcovi increases ADA activity, reduces concentrations of toxic metabolites that are the hallmark of ADA-SCID and improves total lymphocyte counts.

“For decades, physicians, patients and their families have relied upon enzyme replacement therapy as a lifesaving treatment for adenosine deaminase severe combined immunodeficiency, a disease in which the buildup of toxic metabolites can cripple children’s immune systems,” said Morna Dorsey, MD, MMSc, professor of pediatrics at the University of California, San Francisco. “Individuals with ADA-SCID are at an increased risk of severe and recurrent infections and often fail to thrive. By providing specific and direct replacement of the adenosine deaminase enzyme, Revcovi can reduce patients’ risk of potentially serious, life-threatening infections and their debilitating complications.”

Research

SCIG Is as Safe as IVIG for Patients with CVID and Concomitant AITP

A study that compared the efficacy and safety of intravenous immune globulin (IVIG) and subcutaneous IG (SCIG) treatment in patients with both common variable immunodeficiency (CVID) and autoimmune thrombocytopenia (AITP) found SCIG is at least as safe as IVIG; however, an IgG trough level under 7 g/l is a key factor for the development of AITP. The study, conducted at the Centre for Chronic Immunodeficiency in Freiburg, Germany, and the Royal Free Hospital in London, involved 61 patients between age 18 years and 71 years with CVID who had at least one bout of thrombocytopenia defined as a platelet count of less than 50,000/µL if bleeding episodes occurred or a platelet count of less than 20,000/µL without bleeding, one of whom was excluded because of a diffuse large B-cell lymphoma. Thirty patients were treated with IVIG and 31 patients were treated with SCIG. The researchers did not find a higher occurrence of thrombocytopenic events in CVID patients who received SCIG versus IVIG, but they did identify a low IgG trough level as a risk factor for AITP bouts.

Immune Globulin
Each Product Is Unique

By Ronale Tucker Rhodes, MS

While all IG products are comparably effective, they also have relevant differences that determine their tolerability by patients.

THE CURRENT AVAILABILITY of multiple immune globulin (IG) products gives providers many choices when prescribing this lifesaving therapy. The benefit of product choice, of course, is that it allows providers to match the best-suited product to the patient. And, this is extremely important because, while all products contain IgG (the most common protein in the body that helps ward off infections) and they all have comparable efficacy, they are not pharmaceutically equivalent. There are relevant differences between the current products on the market, considered third and fourth generation, that have evolved in terms of composition, resulting in decreased risk of infusion-related reactions and other adverse events. Product variations in sodium content, stabilizers, osmolality, IgA content, concentration and pH can affect the tolerability of a product for one patient versus another, based on both clinical conditions and comorbidities.1,2

When choosing an IG product based on the differences between each, the key factors a clinician considers are the patient’s body type, weight, conditions presenting in addition to the one being treated with IG (such as diabetes, high blood pressure or other heart disease), whether they are pregnant or postmenopausal, other medications taken, kidney function, and if there is patient history of blood clots or migraines. This information is particularly important for dosing recommendation and premedication selection, and it helps clinicians tailor patient-specific suggestions for tolerating therapy.3

Following is a review of the key differences among the products’ stabilizers, osmolality, IgA content and concentration.

Stabilizers

When intravenous IG (IVIG) was originally approved by the U.S. Food and Drug Administration in 1981, it contained no stabilizers, and patients often experienced undesirable side effects such as fever, chills, fatigue and chest, hip, joint and back pain, which were believed to be due to the formation of immunoglobulin aggregates. To resolve this issue, stabilizers were added, primarily sugars such as sucrose, maltose, glucose and sorbitol, and in some cases, glycine and albumin.4

The specific stabilizer used can play an important role in a product’s tolerability.5 Today, most IG products are no longer
stabilized with a sugar; however, a few still are, which can result in other adverse events. There is a strong association between renal failure and sucrose-containing products, rapid rates of infusion and diabetes. This is rare, and the cause of renal failure is unknown, but it is believed that it could be due to the fact that sucrose has the highest osmotic activity of the stabilizers in IG products. In addition, since sucrose is metabolized by an enzyme, called sucrase, that is found only in the intestine, when administered intravenously, sucrose is eliminated unchanged in the urine, possibly resulting in osmotic nephrosis. And, while cautious use of IVIG is recommended in patients at increased risk for adverse renal events, including those with renal impairment, diabetes mellitus, age greater than 65 years, dehydration or hypovolemia, sepsis, paraproteinemia or concomitant use of nephrotoxic drugs, they are not contraindicated in patients with renal insufficiency. In products stabilized with maltose, there is a possible interaction with strips that test for glucose in the blood. The maltose may cause an erroneous reading indicating glucose is high when it really isn’t. However, most test strips have been modified to prevent these erroneous readings when maltose is present.

**Osmolality**

Osmolality is the solute concentration contained in the IG solution; thus, the higher the osmolality, the higher the concentration of the IG solution. Higher osmolality solutions, also known as hyperosmolar, are typically seen with older lyophilized IVIG products. In contrast, today’s fourth-generation products have a more physiologic osmolality comparable to that of individuals’ blood because they have had amino acids glycine and L-proline added to them to help reduce the overall solute load, which can become elevated with sugars.

Hyperosmolar solutions tend to cause more local venous irritation at the infusion site. They also may be associated with an increased risk of thrombosis. Dehydration also can cause the blood to become hyperosmolar, which is one of the reasons people receiving IG therapy are encouraged to drink a lot of water before, during and after the infusion.

**IgA Content**

All IG products contain varying amounts of IgA (one of the five classes of antibodies found in the blood). IgA is not problematic for most people. However, in patients who are IgA deficient, IgA can cause the formation of anti-IgA antibodies that can cause anaphylactoid reactions upon infusion of IVIG, which would result from the IgE development against IgA. While the risk of anaphylactoid reaction in IgA-deficient patients is anticipated, the incidence is low given the total number of reactions reported compared with the overall number of patients. In fact, screening for IgA deficiency prior to IVIG infusion is not routinely recommended.

The amount of IgA in a given IG preparation may also influence the risk for common reactions that are milder such as fever, malaise, myalgia and headache.

**Concentration**

Today’s IG products come in 5%, 10% and 20% solutions. The solution percentage is the number of grams of IgG protein in an IG therapy solution. For instance, a 5% IG product contains 5 grams of IgG protein per 100 mL of solution, a 10% IG product contains 10 grams of IgG protein per 100 mL of solution, etc. The highest-concentration 20% solution can only be infused subcutaneously. Four of the higher-concentration 10% products can be infused subcutaneously, and all but one of them can be infused intravenously. The lowest-concentration 5% products are approved only for intravenous infusion.

Most of today’s products are available as a ready-to-use liquid formulation. However, there is one product that is lyophilized and requires reconstitution and pooling into an evacuated container for administration to the patient.

**Highly Improved and Tolerated Products**

With advances in manufacturing processes, today’s IG products are safer than ever before. However, every IG product has different pharmaceutical characteristics, and there is even variation from batch to batch of each product. It’s these differences that can influence patient tolerability. But with careful patient screening and understanding of the inherent differences in the products, clinicians can ensure that the most appropriate product is prescribed to the patient.

**Sources**


RONALE TUCKER RHODES, MS, is the editor of IG Living magazine.
As the number of approved IG products continues to evolve, so have delivery methods and infusion site options. Here are some tips for helping patients decide which are best for them.

By Abbie Cornett

**TODAY, PATIENTS TREATED** with immune globulin (IG) replacement therapy have options about how and where they receive treatment. Historically, this hasn’t always been the case. It’s been almost 70 years since a medical discovery changed the way many primary immunodeficiency diseases (PIs) and autoimmune diseases (ADs) are treated. This breakthrough occurred when Ogden Bruton, MD, chief of pediatrics at Walter Reed Army Hospital, successfully treated an 8-year-old boy with regular intramuscular (IM) injections of human plasma-derived IG. The boy had agammaglobulinemia characterized by low immunoglobulin (IgG) levels and repeated pneumonia. The new treatment resulted in increased serum IgG levels and an impressive reduction in the number of serious bacterial infections. While few appreciated its implications at the time, Bruton’s success was expanded on and IMIG dosages became the standard of care for PI.

Yet, despite the initial success of IMIG, these injections could only be given in limited amounts and were extremely painful. Thus, it was recognized patients needed a new method of receiving large doses of IG at one time. Fortunately, in the early 1980s, researchers developed the intravenous (IV) delivery method, which led to a rapid
increase in the amount of IG prescribed and the number of diseases it was approved to treat. Then, in the mid-2000s, the subcutaneous (SC) delivery method was developed, allowing for more frequent IG dosing and greater convenience in dosing schedules.

Today, IVIG can be administered at home and in a clinical setting, whereas SCIG is administered only in the home. Together, patients and their physicians should decide which product and delivery method are best for them and, if choosing the IV delivery method, in what setting it will be administered.

What IG Treats
IG was originally used as antibody replacement therapy in patients with PI to supplement the immune system. Since then, its use has expanded considerably to include treating and preventing various other conditions. Currently, IG is approved by the U.S. Food and Drug Administration (FDA) to treat:
- Chronic inflammatory demyelinating polyneuropathy
- Chronic lymphocytic leukemia
- Immune thrombocytopenic purpura
- Infections following bone marrow transplants
- Kawasaki disease
- Multifocal motor neuropathy
- PI
IG is also routinely used to treat a variety of ADs and neurological diseases that are not approved by FDA, including but not limited to:
- Guillain-Barré syndrome
- Lupus
- Polymyositis and dermatomyositis
- Multiple sclerosis
- Myasthenia gravis

A Historical Review
Because of the risk of adverse side effects, IVIG treatments were once administered only in hospitals as a safety precaution. However, expenses of these procedures soon became a major factor for insurance companies, and they began looking for less-costly alternatives to hospital infusions. In response, marketed IG products evolved rapidly to meet this need. These new IG products gave patients and clinicians additional safe IVIG and SCIG therapeutic options, which could be administered in a variety of settings, including the home, hospital, outpatient clinic and doctor office.

Site-of-Care Considerations
When making a decision about an infusion setting, it is important to remember the administration of IVIG is a complex undertaking. Providers and patients need to take multiple factors into account. First, providers need to decide which IG product is best suited to the patient to minimize adverse side effects from therapy. Therefore, each patient’s clinical characteristics must be considered when selecting an IG product. Second, when choosing an IVIG product, providers need to identify which patients need a higher level of care of a clinical setting, and which patients are more suited to receive treatment at home. Further, patients need to decide which option best fits their lifestyle, as well as any insurance coverage limitations.

Because of the risk of adverse side effects, IVIG treatments were once administered only in hospitals as a safety precaution.

Clinical Setting Considerations
For IVIG infusions, doctors may select a clinical setting if they feel additional medical supervision is warranted. In some instances, supervision may be needed because patients have experienced adverse side effects such as anaphylactic reactions, hypotension, seizures, pulmonary edema or aseptic meningitis. Other risk factors may include a health history of diabetes, age (65 years or older), coronary artery disease, hypertension, hyperviscosity disorder (including multiple myeloma, macroglobulinemia and polycythemia), thrombotic events and peripheral vascular disease.

Beyond safety reasons, infusing in a clinical setting allows patients to have more interaction with doctors and nurses who can monitor their health. This oversight is particularly important for patients with comorbidities and those who have been chronically ill for a long time and may fail to notice signs of infection or a declining disease state.
Important Safety Information

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

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

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

Hizentra is a prescription medicine used to treat:

- Primary immune deficiency (PI) in patients 2 years and older
- Chronic inflammatory demyelinating polyneuropathy (CIDP) in adults

Treatment with Hizentra might not be possible if your doctor determines you have hyperprolinemia (too much proline in the blood), or are IgA-deficient with antibodies to IgA and a history of hypersensitivity.

Tell your doctor if you have previously had a severe allergic reaction (including anaphylaxis) to the administration of human immune globulin. Tell your doctor right away or go to the emergency room if you have hives, trouble breathing, wheezing, dizziness, or fainting. These could be signs of a bad allergic reaction.

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

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

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

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

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

The most common side effects in the clinical trials for Hizentra include redness, swelling, itching, and/or bruising at the infusion site; headache; chest, joint or back pain; diarrhea; tiredness; cough; rash; itching; fever, nausea, and vomiting. These are not the only side effects possible.

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

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

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.
HIZENTRA®, Immune Globulin Subcutaneous (Human), 20% Liquid
Initial U.S. Approval: 2010

BRIEF SUMMARY OF PRESCRIBING INFORMATION

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

WARNING: THROMBOSIS

See full prescribing information for complete boxed warning.

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

INDICATIONS AND USAGE

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

CONTRAINDICATIONS

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

WARNINGS AND PRECAUTIONS

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

ADVERSE REACTIONS

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

DRUG INTERACTIONS

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

Based on March 2018 revision
Yet, there are drawbacks to clinical settings as well. One is increased exposure to infection. A study that compared rates of pneumonia and bronchitis in PI patients found those who received IVIG therapy in the home demonstrated significantly lower rates of infection than those treated in outpatient hospitals.\(^8\,9\) Infusing in a clinical setting also costs significantly more and can have quality-of-life implications. Because of scheduling requirements, patients frequently miss work or school, and their ability to travel is limited.

**Home-Based Infusions**

Home-based infusions may be beneficial to patients for many reasons. Unlike the clinical setting, home-based care offers many quality-of-life advantages such as convenience, privacy, and flexibility. When patients receive services in their homes, they have greater control over their living activities and enhanced options for the resumption of normal activities, including work, school and travel. And, because patients are able to schedule treatments around their lives, they have better adherence to medications and better overall outcomes. Another significant advantage of home-based therapy, as mentioned previously, is reduced exposure to pathogens, which can greatly lower the risk of infections for patients.

When infusing at home, patients can choose either IVIG or SCIG. In the past, IVIG outside the clinical setting was discouraged because of the risk of adverse reactions. Currently, those concerns have been alleviated with increased safety and training for home care nurses, so IVIG has seen a substantial shift to home-based therapy.

SCIG infusions, which have become increasingly popular in recent years, are given by slowly injecting purified Ig into fatty tissue just underneath the skin with the use of a mechanical infusion pump. SCIG is a good option for patients who have experienced serious side effects with IVIG. Common injection site reactions to SCIG include redness, swelling and itching, which are usually mild and go away after a day or two. In many cases, these reactions are generally worse with the first few infusions and diminish over time. Adverse reactions to SCIG are rare and usually mild, but can include headaches, feeling hot, nausea, diarrhea, sore throat, rash, increased cough and back pain. Extremely rare are serious side effects such as allergic reactions, kidney problems or blood clots.\(^8\)

When comparing a clinical site to home care, cost must also be considered. Infusing at home is much less expensive. One observational study conducted in 2014 noted a statistically significant difference when patients were switched from outpatient hospital care to home care, with the median price reduced from $6,916 to $4,188.\(^10\) These savings are considerable for insurance companies, healthcare providers and patients.

**Options Are Greatly Expanded**

IG therapy has come a long way since its first use in the 1950s. Today’s IG products are safer than ever, treating a host of diseases and offering patients new routes of administration and treatment sites. Patients and physicians should discuss which product is best for them and where to infuse by taking many factors into account, including the patients’ overall health, past adverse reactions and patient preference.

ABBIE CORNETT is the patient advocate for IG Living magazine.

**References**

AGING AS A health issue permeates the history of medicine. The Charaka Samhita Sanskrit text from India, dating from the time of the Roman Empire or earlier, prescribes light but nutritious meals for older patients. And, fifth century Greek physician Aëtius of Amida specialized in studying the health of the elderly. However, while doctors have always known aging affects all aspects of health, from general fitness to the body’s reaction to various illnesses, the formal scientific study of the specific changes in the body as it ages is a fairly recent development.

“Geriatrics” was first used as a term for the study of medicine among the elderly population in 1909 by Ignatz Leo Nascher, MD, of Mt. Sinai Hospital in New York City. This was after Ukrainian researcher Ilya Ilyich Mechnikov coined the term gerontology in 1903 to describe the larger, multidisciplinary study of aging across medicine, sociology, anthropology and other fields. However, gerontology was slow to gain academic acceptance, with the University of Southern California offering the first degrees only in 1975.

The link between aging and chronic illness is something that has long been recognized, or at least suspected. A fairly common example is prostate cancer: Doctors have known for decades that the older a man is when he develops prostate cancer, the more slowly it is likely to grow and the less likely it will spread. ‘Today, the National Institutes of Health’s (NIH) National Institute on Aging is funding research to look at

While research concerning the link between aging and chronic illness is still in the beginning stages, it is believed that discovering the underlying causes of aging will help to improve quality of life.

By Jim Trageser
how changes in the body as it ages affect the way a host of diseases progress, particularly chronic illnesses. And, NIH is funding the Geroscience Network to study not only aging, but the effects aging has on chronic illnesses and how those effects might be lessened. With the number of Americans over age 65 expected to double to almost 100 million by 2060, and with up to half of any person’s entire medical expenses coming in the last five years of life, this is an issue that will only grow in importance in coming years.

What Is the Geroscience Network?

The Geroscience Network is an affiliation of clinical institutions and researchers formed in 2013 by James L. Kirkland, MD, PhD, at the Mayo Clinic, professor Steve Austad, PhD, at the University of Alabama at Birmingham and Nir Barzilai, MD, at the Albert Einstein College of Medicine. Other member institutions include Harvard, Stanford, Johns Hopkins and Wake Forest universities. Funded by a grant from the NIH’s National Institute on Aging, the network ties together existing programs that conduct research about aging to help scientists map out promising new leads. It also shares research data across institutions, helps organize retreats for scientists to come together to plan future research strategies, and facilitates faculty exchanges to help nurture new research into aging. “Aging is the largest risk factor for most chronic diseases,” explained Dr. Kirkland in a 2016 interview provided by the Mayo Clinic. “The goal of our network’s collaborative efforts is to accelerate the pace of discovery in developing interventions to delay, prevent or treat these conditions as a group, instead of one at a time.”

In the summer of 2016, the Geroscience Network published six papers that laid out a road map for developing new drugs that target specific biological processes associated with aging, getting those drugs into clinical trials, developing standards for measuring “health span” and using mice as a vehicle for testing new theories on aging.

What Is Aging?

Asking “What is aging?” may seem a bit silly; it’s one of those common-sense concepts people grow up with. Aging is what happens to the body over time, and the external signs are embedded in our cultural view of growing older: Grandpa’s hair turns gray, Grandma’s face wrinkles and our favorite athletes see their physical abilities decline over the course of just a decade or so.

At the clinical level, there is a solid road map of the changes that happen in a body. After hitting peak physical strength and condition at about age 30, the following happens:
• Metabolism slows (and, for many, this is accompanied by weight gain)
• Hearing clarity is lost
• The eyes become less able to refocus between different distances
• Muscle mass begins to decrease, and the heart becomes less efficient
• Blood vessels lose elasticity, and high blood pressure is more likely to develop
• Memory isn’t quite as sharp

All of these developments can influence the way a chronic disease affects a patient, whether it’s a malignancy, a neuropathy or an immunodeficiency disease.

**Which Chronic Diseases Are Most Affected by Aging?**

As pointed out by Dr. Kirkland, aging is the leading risk factor for most chronic diseases. Nearly all primary immunodeficiency diseases (PIs) are exacerbated by the aging process: As people grow older, their immune systems gradually lose efficiency, even in healthy people. This not only makes older patients with a PI even more susceptible to an infection, but it can also make it difficult for a physician to differentiate between a PI’s progression and the natural aging process — something doctors have recognized for several decades now.6

But, certain types of PIs are more predominant in older populations than others. One recent study found antibody (IgG) deficiencies comprise the overwhelming majority of PI cases among the elderly.7 These classes of PI occur when the body doesn’t make enough of a particular IgG protein. Therefore, after being exposed to a certain bacteria, they don’t respond by producing new IgG proteins to fight that specific infection. This leads to longer infections and a higher risk of a more serious condition developing, and will normally require the use of antibiotics or other treatment to assist the body in fighting the infection.

Peripheral neuropathy is another chronic condition that is statistically more likely to manifest as individuals grow older. Researchers are still sorting out causation from correlation, but it seems since most cases of neuropathy are caused by immune reactions, trauma, diabetes or alcohol abuse, the longer people live the more likely they are to encounter any of these possible triggers.8 When a peripheral neuropathy introduces numbness to the extremities, one result can be a higher risk of a fall. And, falls are the leading cause of death by injury among the elderly. Even without numbness as an associated symptom, the general loss of strength and impaired mobility caused by neuropathy can make falls more likely.9

Cancers may be the chronic disease most associated with longevity. And, like peripheral neuropathy, simply living a long time exposes a person to more instances of risk of developing the disease. With cancers, a long life also results in cumulative exposure to carcinogens.10 Not only do seniors have a greater chance of developing a cancer, but physicians also know that the older people are, the more challenging it can be to treat their cancers. Older patients are more likely to experience side effects to radiation or chemotherapy, the side effects are likely to be more severe and they take longer to recover from these treatments than do younger patients.

**The overarching goal of geroscience is to uncover new findings that will give physicians tools to address the underlying causes associated with aging, allowing them to more effectively treat any chronic diseases affecting patients.**

It is only in recent years, though, with the development of new technology that researchers have been able to dig deep into the body’s biology to find out what is occurring at the cellular, chemical and molecular levels in the body as it ages and what specific processes underlie these externally observable changes. Researchers believe it is not until we more fully understand the biological and chemical changes that drive the aging process that we’ll be able to more effectively treat age-related risk factors that can complicate chronic diseases in the elderly. And, right now, this process of discovering the underlying biological causes of the symptoms of aging is in a very early stage, with many answers yet to be discovered.
Surgery in older patients also carries more risks of complication, and the recovery times are significantly longer.

Treating any chronic illness is made more difficult when also managing high blood pressure, arthritis, diabetes or any of the other conditions that often accompany living into one’s 70s or beyond. And, there are drug interactions to consider since many seniors are on more than one maintenance drug.

What Advances Are Being Made?

The overarching goal of geroscience is to uncover new findings that will give physicians tools to address the underlying causes associated with aging, allowing them to more effectively treat any chronic diseases affecting patients. But, that will be a long process — one that is just beginning.

In one advance already being made, however, geroscience researchers are tackling a phenomenon known as senescence, which occurs when cells in the body stop dividing and reproducing, but also don’t die. These senescent cells, which were first discovered about 50 years ago, likely serve an as yet unknown biological purpose (perhaps slowing malignant cell growth) by churning out a variety of proteins (those not produced by normal cells) that affect neighboring cells. Senescence occurs throughout life, but in healthy young people, the immune system regularly kills off these cells so they can be replaced by normal cells of the same tissue type. New research suggests that as we grow older, senescent cells begin to build up, and the immune system can’t keep up with eliminating them. It is thought senescent cells are associated with conditions ranging from osteoarthritis to atherosclerosis, but a deeper understanding of their role is needed.11

On a more practical level, there are some promising developments for drugs that can target senescent cells while leaving healthy cells alone. A 2018 Mayo Clinic study on aging in mice found giving them drugs that kill senescent cells reversed many of the symptoms of aging.12 However, since senescent cells in different types of tissue (muscle vs. liver, for instance) produce far different proteins, it has been difficult to develop a test that can identify more than one type of senescent cell at a time, making measuring progress in killing off senescent cells throughout the body a challenge.

On another front, Grubeck-Loebenstein, MD, one of the researchers who first pointed out the challenge of differentiating a PI from normal aging, currently heads the Institute for Biomedical Aging Research at the University of Innsbruck in Austria. Its primary area of research now is to prevent T cell function from declining in the elderly.13

Another study about PI and the elderly examined whether subcutaneous immune globulin (SCIG) treatment could be as effective in the elderly compared to those in their age group using intravenous immune globulin (IVIG) treatment. With fewer side effects than IVIG, the SCIG option offers promise for elderly patients. The study found home-based SCIG is safe and effective in elderly patients with PI, most of whom can self-infuse. Infection rates were low, and no adverse events or difficulties in administering SCIG occurred that resulted in treatment discontinuation.14

The Future of Aging Research

Geroscience, geriatric and gerontology researchers all agree that prolonging “health span” is, if anything, more important to improved quality of life than simply prolonging “life span.” The goal is to lengthen middle age, not old age, so even if people don’t live longer, they will enjoy a better quality of life and encounter fewer medical expenses.15

An essential part of an increased health span is improved treatment options for chronic diseases. As researchers develop their initial explorations of the cellular and molecular causes of aging, future decades should see a rapid expansion of the effectiveness of care for older adults suffering from a variety of chronic conditions, ranging from cancer to PI. ■

JIM TRAGESER is a freelance journalist in the San Diego area.

References
Caring for the Caregiver

When a loved one becomes ill, it’s natural to want to step in and help. But, when the diagnosis is chronic and long-term, those thrust into the role of caregiver will need adequate resources and support to ensure they avoid becoming patients themselves.

By Trudie Mitschang

CAREGIVING RESPONSIBILITIES often sneak up on people. When a loved one is diagnosed with a chronic illness, seemingly overnight, caregivers’ time is no longer their own. It may begin with an extended hospital stay (and the hope that things will eventually “get back to normal.”) But, over time, there are follow-up doctor appointments, prescriptions to fill and errands to run, and gradually, caregivers realize they have inherited an ongoing commitment with no end in sight. The result? Life as they knew it becomes a thing of the past, and they enter a new normal that is anything but.

Defining the Role of Caregiver

Caregivers can be spouses, partners, adult children, parents, other relatives (siblings, aunts, nieces/nephews, in-laws, grandchildren), friends or even neighbors. Whatever the relationship with the person they are caring for, it’s important to add the title caregiver to the list of titles they embrace. While caregivers may be hesitant to formally label their new role, it is important to begin identifying themselves as caregivers so they can begin to search for resources to help navigate this new role.
Realistically, things can quickly get complicated as no one is solely a caregiver; individuals step into this role with other responsibilities already in place. They may be employed full- or part-time. When caring for a sick child, they may be raising other healthy children as well. If they are a caregiver for their spouse, they may also be fathers/mothers, friends, siblings, sons or daughters. Adding caregiving to that list can easily lead to frustration and exhaustion. They might need to navigate social service systems, call doctors while at work, advocate for the care receiver, and take care of their day-to-day needs, while trying to do all of those things for themselves and their families. Eventually, something has to give, and for many caregivers, it is often their own mental and physical health that begins to suffer.

"Most caregivers do not prioritize their basic needs for sleep, healthy diet, exercise and socializing," says Christina Mangurian, MD, MAS, professor of clinical psychiatry at the University of California, San Francisco School of Medicine, and mother/caregiver of a young child with Wiskott–Aldrich syndrome. "Doing these things helps them maintain their own well-being, which is directly related to how well the care recipient does. I encourage caregivers to think about ways in which they can incorporate more self-care into their daily regimen."

Few people step into the role of caregiver having been trained to do the broad range of tasks required. As a result, they may end up with back strain because they haven’t had the benefit of training from a physical therapist on how to correctly transfer someone from bed to chair or wheelchair to car. Or, perhaps they find themselves battling with a spouse who refuses to remain compliant with prescribed medications. These situations can make them feel isolated, but according to the Family Caregiver Alliance, they are extremely common, regardless of the care recipient’s diagnosis.¹

If caregivers are unsure the title fits them, here are some common tasks typically required of someone in a caregiver role:

- **Buy groceries, cook, clean house, do laundry and provide transportation.**
- **Help the care receiver get dressed, take a shower and take medicine.**
- **Transfer someone out of a bed/chair, help with physical therapy and perform medical interventions: injections, feeding tubes, wound treatment and breathing treatments.**
- **Arrange medical appointments, drive to the doctor, sit in during appointments and monitor medications.**
- **Talk with doctors, nurses, care managers and others to understand what needs to be done.**
- **Spend time handling crises and arranging for assistance — especially for someone who cannot be left alone.**
- **Handle finances and other legal matters.**
- **Be a companion.**
- **Serve as an unpaid aide on call 24/7.**

### Research Shows

**Family Caregivers of Any Age Are Less Likely Than Noncaregivers To Practice Preventive Healthcare and Self-Care Behavior.**

**Taking Care of Themselves**

We are all familiar with airplane safety instructions: Put on your own oxygen mask first before you assist anyone else. The same principle applies when caring for a loved one with a chronic illness. Caring for themselves is one of the most important — and one of the most often forgotten — things caregivers can do. When their needs are met, the person they care for will benefit, too.

Research shows family caregivers of any age are less likely than noncaregivers to practice preventive healthcare and self-care behavior. Regardless of age, sex, race and ethnicity, caregivers report problems attending to their own health and well-being while managing caregiving responsibilities.²

What’s more, caregiving can be an emotional roller coaster. On the one hand, caring for a family member demonstrates love and commitment and can be a very rewarding personal experience. On the other hand, exhaustion, worry, inadequate resources and continuous care demands are enormously stressful. Ironically, caregivers are more likely to develop a chronic illness than are noncaregivers, namely high cholesterol, high blood pressure and a tendency to be overweight. Studies show an estimated 46 percent to 59 percent of caregivers are clinically depressed.³ "The tragedy of a debilitating illness goes far beyond the patients themselves," explains Karl
Hawver, MD, a psychiatrist in Kensington, Md. “Caregivers suffer the loss of their dreams, just the way the patient does, and this can make them angry and resentful.”

These feelings of loss and disappointment can manifest in different ways. Some caregivers don’t acknowledge how upset they are, but instead develop physical symptoms such as headaches, back pain and chronic fatigue. “These are real physical ailments,” adds Dr. Hawver, “but they’re due to the underlying sadness. They won’t be cured until the true emotional problem is faced and dealt with.”

For other caregivers, there is an insidious, steady sapping of their energy and enjoyment of life. Slowly but surely, their lives become marked by unhappiness, and it all happens so gradually, they’re not even sure how or when they became so sad. “Caregivers can spend so much time giving care that they neglect tending to their own needs,” Dr. Hawver cautions. “The result is that their lives become devoid of gratification, and they sink into a profound despair.”

Of course, just because someone is a caregiver doesn’t mean he or she will automatically be afflicted with a mood disorder. Extreme stress is also common and can lead to many troubling symptoms, including:

- Feeling overwhelmed or constantly worried
- Exhaustion
- Getting too much or not enough sleep
- Gaining or losing weight
- Becoming easily irritated or angry
- Losing interest in activities previously enjoyed
- Feeling sad
- Having frequent headaches, bodily pain or other physical problems
- Abusing alcohol or drugs, including prescription medications

Too much stress, especially over an extended period of time, can harm health. It’s important for caregivers to find positive ways to alleviate stress. Reaching out for help and talking to friends and family can help. “I encourage caregivers to talk about the situation with friends, family, colleagues, spiritual leaders, counselors — whatever fits them,” says Dr. Mangurian. “Also, there is no shame in seeing a psychiatrist to get medication if they need it to get through a difficult time.”

Practicing Caregiving Stress Strategies

The emotional and physical demands involved with caregiving can strain even the most resilient person. That’s why it’s so important to take advantage of the many resources and tools available to help caregivers provide care for their loved one. It’s true, if caregivers don’t take care of themselves, they won’t be able to care for anyone else.

Here are some tips to help caregivers manage stress:

- Accept help. Prepare a list of ways others can help, and let the helper choose what he or she would like to do. For instance, a friend may offer to take the person they care for on a walk a couple of times a week. Or, a friend or family member may be able to run an errand, pick up groceries or cook.
- Focus on what you are able to provide. It’s normal to feel guilty sometimes, but no one is a perfect caregiver. Caregivers need to believe they are doing the best they can and making the best decisions they can at any given time.
- Set realistic goals. Break large tasks into smaller steps that can be done one at a time. Prioritize, make lists and establish a daily routine. Say no to requests that are draining such as hosting holiday meals.
- Get connected. Find out about caregiving resources in the community. Many communities have classes specifically about the disease a loved one is facing.
- Join a support group. A support group can provide validation and encouragement, as well as problem-solving strategies for difficult situations. People in support groups understand what caregivers may be going through. A support group can also be a good place to create meaningful friendships.
- Seek social support. Make an effort to stay well-connected with family and friends who can offer nonjudgmental emotional support.
- Set personal health goals. For example, set goals to establish a good sleep routine, find time to be physically active on most days of the week, eat a healthy diet and drink plenty of water.

A Helpful What Not to Do List

Toni Bernhard, JD, is the author of *How to Be Sick: A Buddhist-Inspired Guide for the Chronically Ill and Their Caregivers*. A frequent contributor to *Psychology Today* on topics related to living with chronic illness, Bernhard (whose husband is her caregiver) offers sage advice on what not to do when tasked with the role of caregiver. “Many people (including myself) have written about the need for the chronically ill to go through the same type of grieving process that’s triggered by other life-disrupting events, such as the break-up of a relationship or the death of a loved one,” she explains. “If you’re a caregiver, you need time to grieve, too. The drastic change in your life can be a shock. One day, you were free to go out whenever you wanted and hang out with whomever
you wanted. The next day, you were tied to the house and expected to understand how to take care of someone who may need help with the most intimate of life functions.”

Bernhard goes on to explain that in addition to grieving the loss of their own freedom, caregivers may be grieving the loss of the relationship they once had with the person they’re caring for. “In my life, except when we were at our respective jobs, my husband and I used to do almost everything together. Now, when he goes out, he almost always goes out alone.”

In addition to embracing the grieving process, Bernhard says it’s also vital to find moments of both frustration and joy, without guilt. “Let yourself have ‘bad’ days when, even though you’re doing what needs to be done for the person you care for, your heart isn’t in it and you wish you were free of the obligation and the burden. Don’t feel guilty if resentment arises now and then.”

Finding Relief Through Respite

No one can expend their energy, strength and time giving to someone else, especially in the demanding role of family caregiver, without replenishing their own reserves. Respite care brings temporary relief to primary caregivers from the continuing demands of someone with special needs. Respite care may be planned or emergency, in home or elsewhere, for a few hours or perhaps a couple of weeks. The purpose of respite is to allow caregivers to rest, recharge and remember that life exists beyond caregiving.

The question is, once they’ve decided they should take respite, how do caregivers begin to look for good respite care? A preferred respite choice for most people is having someone come into the home to allow the caregiver to leave for a few hours on a regular basis. In some cases, family members can meet this need. If this is not feasible, help from a community-based volunteer group or a home healthcare agency can be considered. The choice depends on the skill level that will be needed and on the ability to pay for respite services.

When a trained volunteer is not sufficient for respite (such as when medical skills are required or when volunteer groups are not a choice in the community), another option may be a home healthcare agency. Good places to start are local affiliates of the Visiting Nurse Associations of America. Visiting nurses essentially invented home healthcare more than a century ago, and since 1983, local groups have organized to offer help that is nonprofit, community-based and Medicare-certified. Working with a home care agency has a number of advantages. A primary one is while caregivers get some respite, a home care aide can actually provide more detailed bed and bath or other caregiving services, so when they return from respite, some of the heavy-duty work of caregiving has already been completed. Respite care is not covered by health insurance, whether Medicare or private, so caregivers should be prepared to pay out of pocket if they contact a home care agency.

Respite care brings temporary relief to primary caregivers from the continuing demands of someone with special needs.

Caregivers Are Not Alone

Caregivers new to their role can rest assured they are not alone. Estimates suggest two-thirds of the U.S. public expect to be caregivers in the future, and 43 percent report it is very likely they will become a family caregiver at a future time. According to the National Center on Caregiving, families typically provide 80 percent of the long-term care in this country, and the need for information, training and education for this growing population is significant. To meet the growing need, the organization has pioneered an online Learning Center (www.caregiver.org/caregiver-learning-center) that offers information, training, classes and more. According to the website, “By taking our most popular education programs and putting them online, we have brought the classroom to you. Our goal is to provide high-quality information and training that is accessible and convenient for family caregivers.”

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.

References
Purchasing Life Insurance with a Chronic Illness

Having a chronic illness is not necessarily a barrier to obtaining a life insurance policy. The key is understanding one’s options and what to expect.

By Ronale Tucker Rhodes, MS

SOME PATIENTS with a chronic illness are under the impression they cannot either obtain or afford a life insurance policy. In fact, a study conducted by Genworth Financial found between 39 percent and 54 percent of adults with pre-existing conditions have no life insurance. According to Ray Dinstel, senior vice president of underwriting at Genworth (an international financial services organization), the fear that the price of a policy will be too high because of their impairments prevents them from buying it. But, life insurance companies do not always preclude those with pre-existing conditions from obtaining life insurance.

Whether chronically ill patients are able to purchase a life insurance policy and how much it will cost depends on the type of policy and how the illness affects coverage. While companies typically group buyers into one of several rate classes based on their health and the risk they represent, many pre-existing conditions are not barriers to an affordable life insurance policy if they are controlled by medication or other treatment. And, many applicants with medically controlled chronic conditions can now get the preferred insurance rate.1

Life Insurance Types

There are different types of life insurance policies for which chronic illness patients can qualify, including term and whole (individual policies), group (through an employer), and simplified issue and guaranteed issue.

A term life policy is often called a “pure” life insurance policy because it’s designed only to protect dependents in case the policyholder dies prematurely. The policy lasts for a set number of years — usually 10, 20 or 30 — during which time the...
policyholder pays premiums to keep it active. If the policyholder dies while the policy is active, his or her beneficiaries receive the agreed-upon death benefit. Term life insurance is typically the lowest cost, and the premium remains the same throughout the life of the policy. When buying this type of policy, the number of years and the amount of the policy should coincide with the time frame the policyholder will be paying the bills and how much money the family would need if the policyholder were no longer there to provide for them.

Whole life insurance is permanent life insurance that combines life insurance and an investment component known as the policy’s cash value. Rather than providing coverage for a set period, it is in force until the policyholder dies, as long as premiums have been paid the entire time. The policyholder can borrow money against the account, but if the loan is not repaid with interest, the death benefit is reduced. In addition, the policy can be surrendered for cash, but then the policyholder will no longer have coverage. The upsides of whole life are the premium remains the same for as long as the policyholder lives, the death benefit is guaranteed and the cash value account grows at a guaranteed rate. And, some policies can earn annual dividends that can be taken in cash or left to earn interest, decrease the premium, repay policy loans or buy additional coverage. The downside of whole life is premiums can be six to 10 times as expensive as term life.\(^2\),\(^3\)

Both term and whole life insurance policies require a medical exam. Therefore, if a complicated health history due to chronic illness makes purchasing or affording an individual policy out of reach, the solution may be a group policy or a simplified issue or guaranteed issue policy. A simplified issue policy requires some medical questions but no medical exam, whereas group and guaranteed issue policies require neither. Individuals with pre-existing conditions can purchase group life insurance through their employers. While this policy is generally at least partially funded by the employer, employees usually have little or no choice of features, coverage is typically limited to one or two times the employee’s annual salary, and coverage is lost when the employee leaves the job. The upside, however, is it is available even to newly hired employees and, as mentioned, there is no required health exam.\(^4\),\(^5\)

Both simplified issue and guaranteed issue policies have higher premiums and lower benefit amounts than individual life insurance policies. Mostly, they are designed to pay for funeral expenses, which is why the policy amounts are small (typically ranging from $5,000 to $100,000), and they are often offered only to those between 45 years and 85 years old.

The benefits of a simplified issue policy include no medical exam, coverage is provided in days or weeks compared to months with other insurance policies, the death benefit is available immediately upon policy issuance, and it is less expensive than guaranteed issue life insurance. The downside is the policy has high premiums (sometimes two to four times higher than other life insurance policies).\(^6\)

A major difference with a guaranteed issue policy is, unlike simplified issue, there are also no medical questions to answer. If the premium is paid, the company is “guaranteed” to issue you a policy. The major drawback is the full death benefit is not available until after the policy has been in force for a specific period, usually one to two years. In some cases, if the policyholder dies within two to three years of purchasing the policy, the beneficiaries are refunded the premiums paid to that point. Some companies give a percentage of the death benefit, but the beneficiaries won’t get the full amount. In addition, it is probably the most expensive life insurance one can buy.\(^6\)

Whether chronically ill patients are able to purchase a life insurance policy and how much it will cost depends on the type of policy and how the illness affects coverage.

What to Expect from Underwriting
When it comes to chronic illness, life insurance companies take many details into consideration for individual policies, including the severity of the condition, treatment, age of onset and other variables. Carriers often rate a condition based on its category: mild, moderate or severe. Those with
mild illness will likely qualify for preferred (less expensive) rates, while those with moderate illness will likely qualify for standard (more expensive) rates. And, while severe illness is insurable, it usually comes at the highest cost.

When applying for an individual policy, preparing for the medical exam can be key.

Individuals who are able to control their condition with medication, diet and lifestyle, and those who were diagnosed at a younger age, are in a much better position for obtaining a policy. Other variables insurance carriers consider include the individual’s build, family history, background history and other medical conditions. For instance, if a person has asthma and smokes, he or she will be denied. Once all of the information is obtained by the insurance carrier, it will assign a rate class based on level of risk. This will determine the monthly premium rate (see below).\(^7\)

For a simplified issue policy, usually four or more questions will be asked to determine approval and cost (some companies will ask a few more questions, and with some, the questions will be slightly different): 1) Do you smoke? 2) Do you currently reside in a hospital or long-term care institution? 3) Have you been declared terminally ill? 4) Do you have AIDS or HIV? By answering “Yes” to smoking, individuals may still qualify for a simplified issue policy, but it will be more expensive. By answering “Yes” to any one of questions two through four, the policy will likely be denied. However, answering “No” to questions two through four will almost certainly gain policy approval.\(^6\)

There is no underwriting process for group or guaranteed issue policies.

How Are Rate Classes Determined?

As mentioned previously, life insurance companies use health ratings to determine what rate will be used to calculate an individual’s premium. The ratings generally fall into four classes: preferred plus, preferred, standard plus and standard. While these rating class names and the factors that determine one’s class can vary between insurers, many follow similar rules:\(^8\)

- Preferred plus: This is considered the best category. Most people in this category do not have a poor health history, are not taking any medications and are within the guidelines for height and weight. Only about 5 percent of people qualify for this class.
- Preferred: This is considered the second-best category. Individuals are deemed to be in excellent health with a few minor health issues that may be treated with medication such as controlled cholesterol, are within the guidelines for height and weight, may have a family history of cancer or other diseases, and participate in dangerous occupations such as risky sports or risky work occupations.
- Standard plus: Individuals are considered to be in better than average health, but may have minor health issues, may be slightly over the height and weight guide, but have no family history of diseases.
- Standard: Although this may be in the bottom of the four general life insurance health classes, it is considered to be the most common category. Individuals are of average health and take multiple medications. Their height and weight do not meet the guidelines, and they have a family history of cancer or other diseases.

In addition, individuals are advised to buy life insurance as early as possible since waiting to buy will always be more expensive. For example, consider a 30-year-old who waits 10 years to buy a policy at age 40:\(^9\)

- A 20-year, $500,000 term life policy will cost about $100 more per year.
- By waiting 20 years until age 50, rates for a 20-year, $500,000 term life policy will more than triple.

Improving the Chances of Approval

Individuals can take steps to improve chances of getting approved for a life insurance policy. According to Dinstel, people with chronic conditions must take medications, not have any significant symptoms or be otherwise healthy to receive an insurer’s preferred rate. And, they need to work with a broker who represents companies that ask whether a condition is controlled. In addition, people with impairments “shouldn’t stop at disclosing their condition,” Dinstel says. “They need to get on the phone with a company representative to provide their full information.” Otherwise, they could be quoted a rate that’s much higher than what they would qualify for if they had disclosed that their condition was under control.

When applying for an individual policy, preparing for the medical exam can be key. To improve the exam’s outcome, Dinstel recommends fasting 24 hours before the exam (to
help lower cholesterol slightly); avoiding alcohol and fatty and salty foods before the exam; refraining from caffeine the morning of the exam; taking medications to ensure cholesterol, blood pressure and other conditions are under control; refraining from a heavy workout the day before the exam (which can cause an elevated protein level); and getting a good night’s sleep the night before.

Furthermore, there are also some things that virtually ensure standard or substandard rates or denial. For instance, having diabetes or a history of cancer will qualify only for the standard rate, and for those who currently have cancer, they won’t even qualify for that rate. In addition, individuals who smoke will pay a lot more for life insurance even if they have a treatable medical condition that is under control. In general, Dinstel says smokers pay twice as much for life insurance as healthy nonsmokers do.

A Broker Can Improve Chances

Chronic illnesses should not be a deterrent to applying for a life insurance policy, and they are no longer a barrier to an affordable policy. What is crucial is to work with a broker who represents a broad range of insurance companies and who is specialized in working with impaired risk cases to find the right company. Some insurers have changed their underwriting guidelines so the majority of their applicants with medically controlled chronic conditions get the preferred rate.

RONALE TUCKER RHODES, MS, is the editor-in-chief for IG Living magazine.

References

Studies show EMED Soft-Glide® Needle Infusion Sets provide

- Easier needle insertion
- Facilitates 90 degree insertion
- Decreased insertion pain
- Decreased removal pain
- Minimization of tissue damage

*If you would like a copy of the needle comparison report please contact sales support.
Understanding Small Fiber

By Jim Trageser

SMALL FIBER NEUROPATHY (SFN) remains one of the least understood diseases. Doctors have a challenging time diagnosing it, and treatment options are limited. There is only a cure if a cause is known, and there is no vaccine to prevent it. In addition, researchers hold contradictory views on how SFN progresses, what the risks are, how to diagnose it and how best to treat it.1

As a June 2016 column in the journal *JAMA Neurology* states: “Despite three decades of intense study, SFN remains an enigmatic condition that is often difficult to diagnose and manage successfully. ... The precise diagnostic criteria for SFN are debated, and the relative role of specific symptoms, signs, specialized investigations ... and skin biopsy for measurement of intraepidermal nerve fiber density is uncertain and somewhat controversial.”

This lack of consensus about SFN in the medical community understandably leaves patients and their families confused and frustrated as they not only look for definitive answers, but an effective approach to ease what can be debilitating pain and other symptoms and restore their quality of life.

What Is SFN?

SFN is a type of peripheral neuropathy, which is a disease of the nervous system outside the brain and spinal column. It is a perplexing disease since it is defined by the damage it causes, rather than from what causes it. The damaged nerves that lead to symptoms of SFN are the small myelinated afferent fibers or the unmyelinated C fibers that extend into the limbs.2 (Myelin is a protective coating around larger nerves; SFN seems to involve nerve cells with minimal or no myelin.) These are the nerve cells that carry information on all sensation, temperature and pain to the brain.

If physicians and researchers struggle to craft a single, testable diagnosis for SFN (sometimes called small fiber sensory neuropathy), those suffering from it have no such difficulty describing it: terrible pain, often with no discernible cause.

Patients may experience hyperalgesia, which is a heightened sensitivity to pain in general. They may also develop hypoesthesia, which occurs when normal stimulation such as the feeling of a sheet over the legs suddenly provokes intense pain.3 In addition, they may experience allodynia, which...
Neuropathy

causes nonpainful sensations to become painful (i.e., lightly touching a limb producing severe pain).

Other symptoms may include sensations of burning heat or freezing cold, or the inability to tell the difference between heat and cold. Patients may experience tingling, numbness and/or feelings of electrical shock. And, oftentimes, these symptoms manifest or worsen at night or during rest.2

Patients with SFN can also have damage to their autonomic nervous system, which can produce the following symptoms:2,4

- Urinary problems
- Erectile dysfunction
- Nausea
- Lightheadedness
- Blurred vision
- Skin discoloration
- Changes in sweating
- Dry mouth or eyes
- Rapid heartbeat

Many patients with SFN also complain of irregular sleep habits, generally due to the onset of symptoms at night.

Patients can develop SFN at any age. And, for some, the symptoms will spread from their feet and/or lower legs into their hands and lower arms.

However, the progression of the condition is the subject of deep disagreement among researchers. A 2016 study conducted at Johns Hopkins University showed nerve damage spread from the lower legs at a fairly rapid rate.3 Yet, just a year later, a study at the University of Cologne in Germany determined the vast majority of cases of SFN will never worsen.5 Clearly, both of these studies cannot be accurate, and other researchers suggest much work needs to be done to expand our understanding of the causes, progression and treatment of SFN.

The 2016 Johns Hopkins study did seem to answer another question, though, about whether SFN is length-dependent. The results strongly suggest that loss of small fiber nerves is consistent across the legs, meaning it is length-dependent. However, it can also be non-length-dependent or even multifocal (present in more than one location in the leg). Other researchers commenting on the study theorize most cases of SFN first manifest in the feet and lower leg because there are fewer nerves in the feet than higher in the leg, so a consistent loss of nerve cells would be more noticeable there.6

What is known is, in rare cases, SFN can spread not only to the hands, but to the trunk and other areas of the body. In even rarer cases, it can be associated with a condition known as erythromelalgia, which increases blood flow to the affected areas (often the feet) without a corresponding increase in oxygenation. This causes the affected areas to feel hot or experience pain, and they may turn a deep red.8

Causes of SFN

As stated above, SFN is caused by damage to the small myelinated afferent fibers or the unmyelinated C fibers that extend into the limbs (the nerve cells that carry information on all sensations, temperature and pain to the brain). That damage, however, can be caused by a variety of conditions, including:

- Diabetes and prediabetes
- HIV infection
- Lupus
- Celiac disease
- Sarcoidosis
- Guillain-Barré syndrome
- Scleroderma
- Hepatitis C
- Lyme disease7
- Sjögren’s syndrome

In addition, certain drugs used in chemotherapy to treat cancer also may cause a form of SFN, a variant known as chemotherapy-induced peripheral neuropathy (CIPN).10

Roughly one-third of SFN cases are attributable to diabetes or prediabetes, making these the single largest cause. And, yet, even with all the above conditions known to lead to or contribute to SFN, there are many cases (between 20 percent and 50 percent) in which the underlying cause cannot be determined.11 These cases are referred to as idiopathic SFN.
Researchers also think there may be some hereditary conditions related to SFN. For instance, it is believed that, in some patients, there is an inherited gene malfunction behind SFN. Mutations in the SCN9A or SCN10A genes can affect the creation of sodium channels in nerve cells, impacting their ability to transmit electrical charges. Such mutations may cause up to one-third of all cases of SFN. Also, amyloidosis with mutations in the transthyretin gene can trigger SFN.

Diagnosing SFN

Because the initial symptoms of SFN are similar to other conditions, a firm diagnosis can take a bit of detective work. Indeed, it is often a process of elimination, removing other possible causes one by one.

In some cases, microscopic examination of the epidermal nerve fiber density in a skin biopsy from the affected area will show a lower-than-normal nerve cell count or the presence of weakened nerve cells. However, one recent study found nearly all patients with SFN had a loss of nerve density over time, meaning a definitive diagnosis may be possible over the course of several years. (When both small- and large-fiber nerves show damage, this is grouped into a different family of conditions known as polyneuropathy.)

In most instances, a physician will begin the diagnosis process by reviewing the patient’s overall health history. Any of the conditions that cause SFN, along with its symptoms, can give the doctor and his or her patient a good starting point from which to work. If the patient has not previously been tested for any of the conditions that can cause SFN, the doctor may order a blood test to check for diabetes, hepatitis C and/or HIV, and depending on any other symptoms or factors in the patient’s health history, the doctor may explore any of the other known causes of SFN.

In cases in which the patient is otherwise healthy with none of the possible associative conditions, the physician may order a skin biopsy. If that comes back negative, the doctor may order a quantitative sudomotor axon reflex test to measure the skin’s ability to sweat, a possible indicator of SFN. Other tests may measure heat or heat-pain thresholds.

What can be confusing to a patient and his or her family is the standard neurological examination given to flag most neuropathies will come back negative in an isolated SFN patient, meaning coordination, reflex and motor skills will appear normal. Light touch and vibration detection will also often be unaffected. And, standard nerve conduction studies will be normal. But, generally, if a patient exhibits all the symptoms of SFN and no other cause can be ascertained, a physician should proceed as though the patient has SFN.

What Is Not SFN

Burning feet syndrome remains a popular description of the symptoms of SFN in some regions of the United States. More formally known as Grierson-Gopalan syndrome, this condition was previously associated with a burning sensation in the feet. However, this is likely an outdated understanding of the symptoms, and researchers now believe this condition was in reality either SFN or, in specific instances, the result of extreme malnutrition (i.e., in prisoners of war or isolated small communities without access to adequate food sources).

Charcot-Marie-Tooth disease, a hereditary nerve disorder, can also cause pain in the extremities, although it will often affect the hands and feet. It generally leads to atrophied muscles in the affected areas, differentiating its symptoms from those of SFN.

Treating SFN

If the cause of SFN can be identified, neurologists can be very effective in treating the disease and improving the nerve density of the small fibers. But, in cases of idiopathic SFN in which there is no known cause, doctors cannot treat the disease. So, instead, treatment focuses on relieving symptoms as much as possible. For instance, controlling diabetes by managing one’s diet, losing weight and exercising regularly can help reduce pain over time in patients with diabetes or prediabetes.

For all cases, ceasing smoking and increasing exercise...
can improve blood flow throughout the body, thus increasing the amount of oxygen available to damaged nerve cells.\(^\text{13}\)

Medicines that have shown some efficacy in treating pain include anti-seizure medications and antidepressants.\(^\text{15}\) However, researchers caution that the available literature on the efficacy of pain treatments with SFN is limited and offers little guidance.\(^\text{2}\)

One recent study suggests one small subset of idiopathic patients — those with fibroblast growth factor receptor 3 antibodies — will respond well to intravenous immunoglobulin (IVIG), showing a marked decrease in both pain and numbness in the affected areas.\(^\text{17}\) Other studies indicate IVIG can help treat SFN linked to celiac disease and Sjögren’s syndrome.\(^\text{18}\)

Patients with CIPN have no specific treatments available, so pain management is the recommended course of action.\(^\text{19}\)

**Looking Ahead**

Research into the causes of SFN and more immediate, practical treatment options is ongoing. However, somewhat surprisingly given the number of people who suffer from SFN, the number of these studies is startlingly small: Fewer than three dozen are currently listed on ClinicalTrials.gov.

Some of the more noteworthy areas of research include exploring how chemotherapy drugs cause SFN. CIPN, which may affect one-third of all cancer patients, involves a class of drugs known as taxanes. A new study discovered how taxanes lead to nerve damage and death, and suggested a pretreatment that might allow cancer patients to avoid CIPN in the future.\(^\text{20}\)

Todd Levine, MD, director of Corinthian Reference Labs and a neuromuscular neurologist in Phoenix, Ariz., recently proposed dividing SFN patients into four subclassifications to allow physicians to better manage their condition:\(^\text{21}\)

- Those with sodium channel dysfunction
- Those with classic neuropathic symptoms
- Those with widespread pain
- Those with autonomic symptoms

While his ideas may not have gained widespread traction, they are indicative of the desire researchers and physicians have for more information and options at their disposal as they work with their patients to address this puzzling condition.

Other studies listed on ClinicalTrials.gov include one looking at whether the experimental sciatica pain reliever Vixotrigine might help in relieving symptoms of SFN, another examining whether IVIG might help even more SFN patients than the subgroup already identified with autoantibodies, and another examining the effectiveness of the experimental pain medicine VX-150, among others.

**More Knowledge Is Needed**

Without a fuller understanding of the causes of SFN, including deeper comprehension of the specific physiological changes behind this disease, researchers are unlikely to develop effective treatments, much less a cure, especially in light of the fundamental disagreements about the causes, methods of diagnosis and rates of progression existing among researchers and doctors.

For now, the process of diagnosing SFN from the exhibited symptoms and then working with a physician to craft an effective treatment regimen will remain a slow, frustrating process for patients and physicians alike.

**References**


JIM TRAGESER is a freelance journalist in the San Diego area.
Profile: Christina Mangurian, MD, MAS

By Trudie Mitschang

AS CAREGIVER for her son who was diagnosed with Wiskott–Aldrich syndrome as an infant, Dr. Christina Mangurian also wears several other hats. She’s a psychiatrist who delivers mental health services to patients and their families, a scientist who understands the healthcare system and a passionate advocate for improving care for all patients. Her experience with her son’s bone marrow transplant in particular taught her impactful lessons about the needs parents have for compassion and emotional support when navigating the care and treatment of a sick child.

Trudie: How old was your son, Anderson, when he was diagnosed with Wiskott–Aldrich syndrome?

Dr. Mangurian: Anderson was diagnosed with the most severe form of Wiskott-Aldrich syndrome, a genetically inherited immunodeficiency disease affecting one in 250,000 males, when he was just 4 months old. Children with severe Wiskott-Aldrich syndrome die early of infection or hemorrhage if not treated with a bone marrow transplant.

Trudie: Tell us about Anderson’s bone marrow transplant.

Dr. Mangurian: Anderson had a bone marrow transplant in August 2012 when he was 10 months old. Children receiving a transplant from an unrelated donor or via cord blood donation can be hospitalized for up to three months — all in an isolation room because of the risk of infection. My family had already endured a three-week hospitalization when my son was only 1 month old, so I knew it would be stressful on all of us. I had to develop a plan to take care of not only Anderson, but also my 4-year-old daughter, my husband and myself both physically and emotionally during this marathon. That’s why I’m passionate about helping other families do the same.

Trudie: What did this experience teach you about the healthcare system?

Dr. Mangurian: Despite all of our planning, the love showered on us by family and friends, and support from the transplant providers, nothing could prepare us for the reality of the hospitalization. Until I began “living” on the transplant unit, I hadn’t really understood what it was like to be on the other side of the patient-provider relationship. It opened my eyes to the universe of fear and vulnerability, where healthcare providers are in complete control and patients (even those who are doctors) are at their mercy across a wide logistical and emotional divide.

Trudie: What was it like being on the other side of the patient-doctor equation?

Dr. Mangurian: Feeling absolutely no control over this life-and-death situation, my husband and I struggled constantly with emotions such as vulnerability, fear and anger. As a psychiatrist, I knew these feelings were normal, but there was no mental health professional on the transplant team assigned to help us. Social workers were available, but they seemed to deal with the “problem cases,” not the easier ones such as ours. Child life specialists made my son’s daytime life as normal as possible, but they did not attend to my emotions. Thankfully, I had my own psychiatrist and my husband, parents and friends to turn to for the emotional support I needed. Many families are not so fortunate.

Trudie: Why do you think the treatment process for chronically ill children is so hard on families?

Dr. Mangurian: It struck me again
and again that, because of the increasing complexity of medical care, doctors do not seem to have the time to take the “emotional temperature” of patients or their caregivers during prolonged hospitalizations, nor are providers thinking critically about essential, patient-centered concerns to facilitate wellness. In my opinion, we were treated at the best pediatric bone marrow transplant unit in the country, but the empathic failures made me wonder about the emotional experience for families at other hospitals.

**Trudie:** What recommendations do you have for doctors in these situations?

**Dr. Mangurian:** I believe doctors who are treating children for prolonged periods should ask the parents how they are feeling about being in the hospital every day. If this is unreasonable given time constraints, then add a team member to fill this role. Again, there is plenty of evidence that children do better if their parents are emotionally grounded. By not being asked, parents may have the impression the doctors don’t care, and clinicians may miss important warning signs of parental depression or anxiety that could negatively affect outcomes for critically ill children.

**Trudie:** What was the turning point for your family?

**Dr. Mangurian:** About a week after the transplant, the chief of the pediatric bone marrow transplant unit visited us on the day he returned from vacation. Without ceremony, his first words to me were: “Hi, Mom, how are you doing? Do you need a hug?” As a psychiatrist, I was taught not to hug my patients, but at that moment, I did need a hug, and I needed it from him. I needed it from my son’s doctor, the person we were counting on to save his life, the person who was supposed to help us through this terrible ordeal. Although his bedside manner is not generally “touchy-feely,” our doctor knew I needed him to cross the doctor-patient-parent boundary. His small gesture took no extra resources and little extra time. Yet, it instantly made me feel better. And, that is what ultimately makes him not just a good but a great doctor.

**Trudie:** What advice do you have for parents/caregivers?

**Dr. Mangurian:** I think most caregivers are the experts when it comes to caring for their children. But, what most caregivers don’t prioritize is their need for sleep, a healthy diet, exercise and socializing. Doing these things helps them maintain their own well-being, which is directly related to how well the child does.

**Trudie:** How well did you follow your own advice?

**Dr. Mangurian:** Initially, during my son’s first hospitalization, I did not follow this advice at all. I got no sleep and no exercise for three weeks. This led to my first experience with postpartum depression. I recovered through the support of family, friends, psychotherapy and medication. I learned the hard way, and it forced me to develop a plan to manage my own well-being while my son was hospitalized for his bone marrow transplant.

**Trudie:** Put on your psychiatrist hat. How can parents/caregivers manage stress and anxiety when dealing with a family member’s illness?

**Dr. Mangurian:** This is very individual. I encourage parents to talk to friends, family, colleagues, spiritual leaders and counselors — whatever fits them. And, there is no shame is seeing a psychiatrist to get medications if they need them.

**Trudie:** What has this experience taught you about yourself?

**Dr. Mangurian:** I learned what really matters in life. I learned how fortunate I am to have so many people who love me, especially my husband, parents, extended family and friends. I have also learned I want to give back and help others navigate this experience in ways that are helpful to their families.

**Trudie:** Looking back, if you could give your younger self advice, what would it be?

**Dr. Mangurian:** I would tell myself that all of the planning we put in would pay off in the end. I would tell myself that things will be OK.

**TRUDIE MITSCHANG** is a contributing writer for IG Living magazine.
Well, I Wasn’t Expecting That to Happen

By Stacy Oliver

WE ALL HAVE ups and downs. Some days, we can take on the world. Other days, we dive under the covers in bed. This year has been full of change, and it’s not over. Sometimes there is no manual to help us navigate what’s coming; we just have to face it.

I might as well spit it out. I am going to die. Well, we all are; that’s life. But, some of us know a little more about our expiration date — like what’s on a carton of eggs. I might be good for seven days past my “best used by” date. I was diagnosed a week before my birthday with MSA-C (multiple systems atrophy). It’s a neurodegenerative autonomic disease that affects the cerebellum (the area of the hindbrain that controls movement, coordination, balance, equilibrium and muscle tone). The “C” stands for cerebellum. For years, I became used to my MMN (multifocal motor neuropathy) acronym, but now I have to learn new letters. My prognosis is I have five to seven years (that’s the norm) of decline. I use a walker, my speech sounds like I’m drunk or Chico Marx, and my gait and balance are way off.

I’m not working anymore; my blood pressure was so low I’m on pills to raise it. Who knew there was such a thing? I was thoroughly tested and, well, I wasn’t expecting that to happen. I actually felt bad for the doctor who told me the news. He was kind and compassionate. We had never heard of MSA-C, and he had to explain it to us. It was like those moments you see in the movies.

I started to feel like a unicorn. The doctor had never seen so many neurological diseases in one person. Of course, MSA-C isn’t as rare as MMN, which wins the prize for that. But, it is a rare disease, too! I had to be an overachiever? I couldn’t have something that is common and ordinary? At the moment, I’m still being treated with intravenous immune globulin (IVIG). It keeps me going, and I see my wonderful home nurse. I am very fortunate to have different specialists who like to work together. If I have a problem, they work the problem from their angle or specialty. If they don’t know what’s wrong, a colleague in a different area is brought in. It took a neurologist, ophthalmologist-neurologist, rheumatologist and autonomist to diagnose MSA-C.

I’ve only processed this for a few months, and I want to be transparent: The doctors don’t know where I am in the spectrum. A year in, probably. No one seems to really know what’s going to fail me first. I decided to take each day as it comes. I baked cookies for a bake sale and started thinking: “Maybe next autumn I won’t be able to do this.” Then, I stopped myself. “Be here now,” I said. I have today. This moment.

That’s what my husband and I did when we heard the news. We made immediate plans to go to New York City. I wanted to be somewhere different, feel the aliveness of all the people and see places I had only heard about in magazines and cultural references. We had a whirlwind three days and came home. (Our three doggies were very happy to see us.) Now what? Maybe some people drop everything and travel around the world. Maybe some folks climb a mountain. Everyone has their own path during this experience. For me, normalcy is important. I want to appreciate my day-to-day rhythm. I want to spend time with people in my life. I want to move and enjoy my body each day before I can’t move at all. I want to be with my husband as he pushes me in a wheelchair, even if it’s just sitting on a bench watching the beauty of the season.

This magazine is called IG Living. My IVIG still invigorates me, and I’m going to concentrate on the “living” part. I’ll keep writing my articles until I can’t or the powers that be decide they want a change. As I’ve seen my body change over the past 10 years, I’ll continue to see how this new disease develops. So, take it from me (please, my hands aren’t the most stable): Eat the cupcake, wear the shirt (don’t save it for an extraordinary occasion), put the lipstick on, go, do, be. I have to say I did! I don’t think I should have turned right when I should have gone left. As poet Adrienne Rich says in her “Moonlight Serenade” poem published in her book, Your Native Land, Your Life:

“We’ll dream of a longer summer
But this is the one we have:
I lay my sunburnt hand
On your table:
This is the time we have.”

STACY OLIVER was diagnosed in 2008 with multifocal motor neuropathy. When she isn’t writing her book, herding three pit bulls or trying to put eyeliner on straight, she is working on her super secret identity as Neuropathy Girl, who will one day save the world after an infusion and a nap.
Developing New or Unexpected Symptoms

By Ilana Jacqueline

THE FIRST TIME I experienced a seizure, I was at a party. I had a purse full of medications for all sorts of symptoms, but a seizure wasn’t one of them. Why would it be? I’d never had one. No doctor had ever indicated I would have one as part of my diagnosis, and I’d had no warning signs one was about to occur.

But, it happened anyway. It was humiliating and terrifying, and my brain felt like electrified Jell-O. I hoped it was a one-time episode, and after a checkup and hospitalization, I was told “Everybody gets a one-time free pass in life for seizures. It’s usually nothing and won’t happen again,” so I was discharged.

At my next-day follow-up appointment, I had my second seizure right in the office lobby. After my face-plant, I was a wreck of emotions and stress. I had so many questions and, just as with most of my diseases’ symptoms, I’m still not a perfect patient at coping with them. However, here’s what I have learned about what to do when the unexpected strikes.

Take a Minute
Call it your pity party with a curfew. After getting slammed with an unexpected new symptom, it’s OK to completely lose your cool for a minute, or even a day. But try to limit that moment of sitting on the couch in your pajamas crying into your sleeve to about a week and a half — max.

Spend that time researching (and then not panicking about what shows up in Google images) and reading other patient stories about how they got through it. And, research all the potential new specialists who are going to help guide you through this new phase.

Friends and family may be knocking down your door, but sometimes you just want to go through the shock alone. Other people might feel if they’re left alone for more than a minute, their brain is going to swallow them whole. Whatever your preference, your way is the right way for you.

Make a Plan
The shock has worn off, the pity party has all broken up, and now it’s time to start thinking about next steps. Are you going to start being social again? Get back to school or work? And, most importantly, how are you going to start making smart choices about how you handle this new symptom? Start by interviewing new specialists, reading up on helpful material and checking out free resources.

Maybe you can’t control this new symptom to the degree you’d like, but having a plan to help you deal with the physical and emotional ramifications of it can ease your stress.

Be Safe
If your new symptom presents as some kind of unexpected episode or attack, it’s a good time to think about safety systems.

If your new symptom presents as some kind of unexpected episode or attack, it’s a good time to think about safety systems.

If your new symptom presents as some kind of unexpected episode or attack, it’s a good time to think about safety systems.
ALL CHILDREN, as well as adults, get angry from time to time. It’s a normal, innate reaction hardwired into our brains as part of the fight-or-flight response, which kicks in when we feel threatened. For a child, sometimes a threat is physical such as an impending fistfight or an angry dog. But, there can also be emotional threats such as being made fun of or called a not-so-nice name. The resulting fear, hurt, disappointment, pain or grief can be quite upsetting, causing the child to lash out. Getting angry doesn’t remove the hurt, but it makes the child feel less powerless and temporarily eases the pain.1

Aggression Issues vs. Normal Childhood Behavior?

It’s normal for a child to have occasional tantrums or meltdowns. But when it becomes a pattern — when the child does these things repeatedly or can’t control his or her temper — it may be more than typical childish behavior.2

Here are some signs that a child’s emotional outbursts are a concern:2

- If the behavior is past the age at which tantrums are developmentally expected (up to about 7 or 8 years old)
- If the behavior is dangerous to the child or others
- If the child’s behavior is causing trouble at school, and teachers are reporting that he or she is out of control
- If the child has trouble getting along with other kids because of the behavior, and he or she is excluded from birthday parties or play dates
- If the tantrums and defiant behavior cause conflict at home and disrupt daily family life
- If the child is upset, feeling he or she can’t control the anger

There are many possible underlying causes of angry outbursts and aggressive behavior, including attention deficit hyperactivity disorder, especially if the child also experiences hyperactivity and impulsive behavior; anxiety or feelings of acute fear; trauma, neglect or chaos at home; learning problems (if the child acts out repeatedly in school or during homework time); sensory processing issues (for example, if the child is oversensitive or undersensitive to stimulation such as scratchy clothes or too much light or noise);2 and autism.

Calming an Angry Child

When parents are faced with the upsetting behavior of an angry child, it’s tempting to yell or lash out at him or her, but losing control will only make matters worse. The parents’ job is to try to restore calm. Only when the child is calm will he or she be able to learn and understand how to behave better. There are several ways to try to calm a child down:

- **Provide closeness and physical touch.** Even if it doesn’t seem like the child wants it, parents should be ready to show affection. Move physically closer to the child to curb his or her angry impulses. Sometimes, all that is needed for an angry child to regain control is a hug or other show of affection. However, if a child has severe emotional problems, he or she may have trouble accepting affection and will not appreciate it.3

- **Ease tension through humor.** Humor offers a distraction from whatever triggered the child’s anger. Getting the child to laugh offers him or her an opportunity to save face. However, it is never appropriate to use sarcasm, teasing or ridicule as a negative response to a tantrum.3

- **Provide physical outlets and other alternatives.** There is something to be said about “burning off steam” to stem feelings of anger. Give an older child half an hour to shoot hoops or jump on a trampoline to cool off. But, never send an angry child off alone to calm himself down. Parents should remain
in close proximity, without hovering. The parents’ goal is to restore a sense of safety, which requires a calm presence.1

**Use physical restraint.** Occasionally, a child may be so completely out of control that he must be physically restrained or removed from the scene to prevent injury to himself or others. This may also “save face” for the child (the less of the episode that is witnessed by others, the better). It’s important for the child to realize that physical restraint or removal from the scene isn’t punishment, but rather the parent’s way of saying, “You can’t do that.”

**Teaching a Child to Manage Anger**

Because of limited life experience, children lack the proper context for their upsets. What might be a small disappointment to adults can seem like the end of the world to children. And, since their frontal cortex, which helps them self-regulate, isn’t fully developed, children are even more prone to lashing out when angry.1 But, as children transition to adults, they’ll need to have the tools necessary to control their own emotions. Here are a few suggestions to help parents teach healthy anger management in everyday life:

**Start with yourself:** Parents should learn to control their own anger and stop modeling the behaviors they want to change in their child. It can be tempting to yell at a child when he or she is acting out, but if parents give in to that temptation, how can they expect their child to learn to control himself?1

**Help the child be aware of the “warning signs.”** Once a child is in a full-out tantrum or emotional outburst, managing the angry impulses that started it is almost impossible. But if parents can help their child notice when she’s getting annoyed and learn to calm herself, she’ll have fewer tantrums.1

**Find the triggers.** Parents may notice that certain situations, people or things consistently make their child upset. These are the child’s triggers, and parents should take note and make adjustments to the child’s routine to try to avoid as many of these triggers as possible. For example, if the morning routine is stressful, and getting out the door for school is a chronic cause of distress for a child, parents should help to ease this stress by giving time warnings, having the child lay out clothes and shower the night before or requiring them to wake up earlier.2

**Teach coping skills.** Parents can help their child by giving him or her tools to manage angry impulses. When the child is calm, parents should help him make a list of constructive ways to handle strong emotions. Together, they can practice these coping skills and then post the list somewhere visible or in multiple locations. The actions on the list should be age-appropriate, and may change as the child gets older. Some calming techniques are taking long, deep breaths; squeezing a squishy ball; putting on music and doing an “angry dance;” wrapping one’s arms around one’s own body instead of hitting someone else; and for an older child, drawing or writing on paper what he is angry about, and then ripping it into tiny pieces.1

**Reinforce good behavior.** Parents need to tell their child what behaviors please them and encourage the child’s positive efforts. During the day, try to make purposeful comments such as, “Thank you for brushing your teeth and getting ready for bed on your own,” or “I appreciate how you put away your backpack before you went outside to play.” The good behavior can then be rewarded with things like time on the iPad or going out for a special treat.

On the other hand, parents should give consistent consequences for behaviors they want to discourage.2 The child needs to know what the consequences are for negative behaviors such as time-outs, and parents need to show him they’ll follow through with these consequences every time.

**Teach the child to express herself verbally.** When a child is able to tell someone what makes her angry or upset, it helps her feel she has control and reduces acting-out behavior. Encourage the child to express these feelings using words rather than actions. For example, “I don’t like that you took my toy while I was playing with it. I don’t feel like sharing right now.”

**Seek Help When Needed**

It takes a lot of effort on the part of parents to teach their child how to properly respond to feelings of anger and negative emotions. If after a few months of teaching coping skills and offering suggestions for alternative behaviors, parents don’t feel like things are changing, it would be wise to seek professional help. An experienced therapist can help the child work through deeper feelings, and develop his or her ability to manage emotions.1

**References**


**JESSICA LEIGH JOHNSON**

is a stay-at-home mom and mother of four kids, three of whom have X-linked agammaglobulinemia. She is a member of American Christian Fiction Writers and has written one book about the loss of her son to a primary immunodeficiency.
SPRING IS coming, but for now, cold weather and its associated illnesses still linger, causing the more than 26 million Americans who suffer from asthma an influx of incidents. Seasonal cold, dry air, in addition to the abundance of circulating sinus and upper respiratory infections, can be a troublesome mix for asthmatic lungs and increase asthma symptoms. In the U.S., the number of individuals diagnosed with this medical condition is growing, making it one of the nation’s most common and costliest diseases, according to the Centers for Disease Control and Prevention.

Identifying Different Types of Asthma

Asthma occurs when the airways become inflamed, restricting the ability to breathe. The three most common types of asthma, all of which cause shortness of breath, include exercise-induced, nocturnal and allergic.

Originally labeled exercise-induced asthma, this type is now referred to as exercise-induced bronchoconstriction, since exercise itself does not directly cause asthma. Coughing, wheezing, chest tightness and shortness of breath are the most common symptoms.

All asthma sufferers also experience nocturnal asthma. As the name implies, symptoms include sleepless nights filled with coughing, wheezing or chest pains. Nocturnal asthma requires a more effective long-term treatment.

Airborne allergens — from dust mites and pet dander to pollen, mold or pollution — are the most common triggers of allergic asthma. When these elements are inhaled by someone with an allergy, the body’s shields are alerted leading to wheezing, coughing and other asthma-related symptoms. Individuals who suffer from allergic asthma should see an allergist or immunologist to decipher what is triggering the attacks.

Managing Asthma

Although there is no cure for asthma, there are ways to manage it. The best route is to identify and avoid triggers, take medications to prevent further symptoms and have a plan in place for future asthma attacks. In addition, identifying the type of asthma a patient suffers from will help physicians determine the best treatment.

For those who suffer worse symptoms depending on the season, it’s possible a more natural approach could be effective. This could include eye drops and nasal decongestants such as a saline nasal rinse to flush out molds and allergens.

Now or Later: How to Treat Asthma

There are two types of asthma medications: long-term and quick relievers.

Long-term medicines, which come in pill and inhaler forms, are needed daily. The most common forms are inhalers and nebulizers because both send medications directly into the lungs. Nebulizers change the medication from a liquid to a mist and are often used in infants and small children who have a difficult time using inhalers.

Since asthma can strike at any time, quick-relief medications that help relax muscles and open the airways when a sudden attack occurs may also be needed.

Breathe Easier

Breathing is a bodily function few think about daily; it just comes naturally. But for the population that suffers from asthma, the ability to inhale and exhale can be stifled at any moment. Thankfully, a variety of treatment options are available to help asthma sufferers breathe and rest a little easier.

HEATHER BREMNER CLAVERIE is a contributing writer for IG Living magazine.
Filters can prevent some pollutants from entering living spaces, but pet dander and dust mites can be produced inside the home. An effective air cleaning device such as the Rabbit Air Minus A2 Air Purifier can help trap and remove these. In addition, the air purifier monitors ozone levels and has wireless capabilities. It can be hung on the wall and outfitted with a customized panel with images ranging from Vincent Van Gough and Claude Monet or Peanuts’ Snoopy and Hello Kitty for the younger crowd. Starting at $549.95; Rabbitair.com

Asthma is the leading chronic illness in children. Since young children have a difficult time using inhalers, nebulizers can help open their airways. The Mabis Margo Moo Compressor Nebulizer helps calm and entertain children while also treating their asthma. It comes with a cute barn-shaped docking station and a handle for easy transport. $64.99; natallergy.com

Some natural or homeopathic medications can help with respiratory issues or asthma. One product that works well to clear sinuses is Simply Saline Nasal Relief Spray by Arm & Hammer. This product naturally softens mucus so it drains easier and helps breathing easier. This spray is drug-free and is safe to use with prescription medications. $7.26; Target.com

Scents, perfumes and dyes in many commercial household cleaners can flare up respiratory issues. And when asthma is an issue, it’s necessary to clean often. Bona Free and Simple Hardwood Floor Cleaner is free of dyes and will effectively help keep dust at bay. Starting at $9.99; Amazon.com

Anti-inflammatory drugs such as inhaled steroids, which are by prescription only, can help provide long- and short-term relief from asthma by helping to reduce swelling and mucus. Flovent HFA contains a medicine called fluticasone propionate, a synthetic corticosteroid (corticosteroids are natural substances found in the body that help fight inflammation). When inhaled regularly, it can help prevent asthma symptoms. It can be used by patients 4 years and older. www.flovent.com/about-flovent-hfa/index.html

Mold spores, dust, pollen and viruses can trigger attacks in asthmatic individuals. Ensuring air filters are certified allergy and asthma friendly, according to the Asthma and Allergy Foundation of America, is a good way to make sure they will adequately remove pollutants from the air. Filters should be changed every 30 to 60 days. The Filtrete Elite Allergen Reduction Healthy Living 2200 filter will attract and capture large particulates, making the air safer. $44.73/two pack; Amazon.com
**Book Corner**

**Crohn’s and Colitis: Understanding and Managing IBD (Third Edition)**

*Author: Hillary Steinhart, MD, MSc, FRCP(C)*
*Publisher: Robert Rose*

This book, which aims to help patients deepen their knowledge about their disease, has been recognized by practitioners and patients for its information and its supportive and easy-to-follow approach. This new edition features updated content based on the most current research and standards for diagnosis and treatment, along with information on genetics-based drugs and naturopathic treatments. It is written as a tool for improving quality of life for anyone diagnosed with inflammatory bowel disease. The book also includes information on underlying causes, clinical features and effective treatments, as well as informative charts, case studies, “Did You Know?” boxes and answers to frequently asked questions.

**Death and Chronic Illness in the Family: Bowen Family Systems Theory Perspectives**

*Authors: Peter Titelman and Sydney K. Reed*
*Publisher: Routledge*

Rooted in Murray Bowen’s family systems theory (a theory of human behavior that views the family as an emotional unit), this edited volume provides conceptual ideas and applications useful to clinicians who work with families facing chronic illness or the death of a family member. The text is divided into four parts: Part I provides a detailed overview of Bowen’s theory perspectives on chronic illness and death and includes Murray Bowen’s seminal essay “Family Reaction to Death.” In Parts II and III, chapter authors draw upon Bowen theory to intimately explore their families’ reactions to and experiences with death and chronic illness. The final part uses case studies from contributors’ clinical practices to aid therapists in using Bowen systems perspectives in their work with clients.

**Chronic Illness and Long-Term Care: Breakthroughs in Research and Practice**

*Author: Information Resources Management Association*
*Publisher: IGI Global*

This book features current research on the diagnosis, monitoring, management and treatment of chronic diseases such as diabetes, Parkinson’s disease, autoimmune disorders and many more. Highlighting a range of topics such as medication management, quality-of-life issues and sustainable health, it is intended as a reference source for hospital administrators, healthcare professionals, academicians, researchers and graduate-level students interested in the latest research on chronic diseases and long-term care.

**The Port**

*Author: E.H. Sonny*
*Publisher: Amazon Digital Services*

When a boy becomes sick, the Masked Ones steal him from his happy graveyard home and place a port in his chest so they can summon him at will. Raw and poignant illustrations reveal the boy’s conflicted relationship with the port and a medical system that heals his body but damages his spirit. In a story both cathartic and hopeful, The Port speaks to the importance of patient empowerment at all ages, especially when managing a chronic illness and/or enduring repeated hospitalizations.
Download the IG Living eBook today—now available for iPad, Nook and Kindle!

“You can lament what is lost to you, whether it’s opportunity, a person or your health, but clinging to anger is no way to experience life.” — Rebecca Zook in “Life Lessons,” excerpted from Chronic Inspiration.

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

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

— Jenny Gardner

Chronic Inspiration can be purchased on iTunes, Amazon and Barnes and Noble.com
**RESOURCE CENTER**

**Ataxia Telangiectasia (A-T)**
- Website: [www.atcp.org](http://www.atcp.org)

**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)**
- Website: [www.gbs-cidp.org](http://www.gbs-cidp.org)
- Website: [www.niehs.nih.gov/research/resources/imacs](http://www.niehs.nih.gov/research/resources/imacs)

**Evans Syndrome**
- Online Peer Support: [www.evanssyndrome.org](http://www.evanssyndrome.org)

**Guillain-Barré Syndrome (GBS)**
- Website: [www.gbs-cidp.org](http://www.gbs-cidp.org)
- Website: [www.muscleatatlas.org](http://www.muscleatatlas.org)

**Idiopathic Thrombocytopenic Purpura (ITP)**
- Website: [www.itpsupport.org.uk](http://www.itpsupport.org.uk)
- Website: [www.fda.gov](http://www.fda.gov)

**Kawasaki Disease**
- Website: [www.kdfoundation.org](http://www.kdfoundation.org)
- Website: [www.emedicinehealth.com/kawasaki-disease.htm](http://www.emedicinehealth.com/kawasaki-disease.htm)

**Mitochondrial Disease**
- Website: [www.umdf.org](http://www.umdf.org)
- Website: [www.mitoaction.org](http://www.mitoaction.org)

**Multifocal Motor Neuropathy (MMN)**
- Website: [www.foundationforpn.com](http://www.foundationforpn.com)

**Multiple Sclerosis (MS)**
- Website: [www.nationalmsociety.org](http://www.nationalmsociety.org)
- Website: [www.msfocus.org](http://www.msfocus.org)
- Website: [www.nationalmsociety.org](http://www.nationalmsociety.org)

**Peripheral Neuropathy (PN)**
- Website: [www.neuropathyaction.org](http://www.neuropathyaction.org)
- Website: [www.nmfs.org](http://www.nmfs.org)
- Website: [www.nimh.nih.gov](http://www.nimh.nih.gov)

**Primary Immune Deficiency Disease (PI)**
- Website: [www.PrimaryImmune.org](http://www.PrimaryImmune.org)
- Website: [www.idffriends.com](http://www.idffriends.com)
- Website: [www primaryimmunedeficiencynetwork.org](http://www.primaryimmunodeficiencynetwork.org)

**Scleroderma**
- Website: [www.scleroderma.org](http://www.scleroderma.org)
- Website: [www.nhgtt.org](http://www.nhgtt.org)

**Stiff Person Syndrome (SPS)**
- Website: [www.aarda.org](http://www.aarda.org)
- Website: [www.livingwithspss.com](http://www.livingwithspss.com)

**Pemphigus and Pemphigoid**
- Website: [www.pemphigus.org](http://www.pemphigus.org)
- Website: [www.primaryimmune.org](http://www.primaryimmune.org)

**Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)**
- Website: [www.spas.org](http://www.spas.org)
- Website: [www.pandasnetwork.org](http://www.pandasnetwork.org)
- Website: [www.muscleatatlas.org](http://www.muscleatatlas.org)

**Myasthenia Gravis (MG)**
- Website: [www.myasthenia.org](http://www.myasthenia.org)
- Website: [www.sclero.org](http://www.sclero.org)

**Online Peer Support**
- Forum: [www.gbs-cidp.org/forum/main-forum](http://www.gbs-cidp.org/forum/main-forum)
- Forum: [www.idffriends.com](http://www.idffriends.com)
- Forum: [www.sclero.org/support/forums/a-to-z.html](http://www.sclero.org/support/forums/a-to-z.html)

**Websites**
- [www.foundationforpn.com](http://www.foundationforpn.com)
- [www.gbs-cidp.org](http://www.gbs-cidp.org)
- [kidshealth.org/parent/medical/heart/kawasaki.html](http://kidshealth.org/parent/medical/heart/kawasaki.html)
- [www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsofChildhood/Kawasaki-Disease_UCM_308777_Article.jsp#.T1T2boePWE0](http://www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsofChildhood/Kawasaki-Disease_UCM_308777_Article.jsp#.T1T2boePWE0)}
BioSupply® is the online product ordering platform by FFF Enterprises, Inc., the largest and most trusted distributor of plasma products, vaccines, biosimilars and other specialty pharmaceuticals and biopharmaceuticals. Visit www.ffenterprises.com to learn more about us.

BioSupplyOnline.com makes ordering your products easy, fast and convenient!

Available Products
- Albumin/Plasma Protein Fraction
- Coagulation Products
- Hyperimmune Globulins
- Immune Globulins
- Influenza Vaccines & Treatment
- Specialty Biopharmaceuticals & Pharmaceuticals
- Other Vaccines
- Surgical Sealants
- Ancillary Products
- Oncology
- Biosimilars
- Generic Injectables

We are proud to be an accredited NABP® Verified-Accredited Wholesale Distributor for all authorized U.S. plasma products manufacturers.

BioSupply is a quick and easy-to-use platform offering instant access to the critical-care products you need when you need them. Our customer-driven online portal empowers you to order what you want, when you want it, with just one click so you can better manage your inventory. With over 28 counterfeit-free years, you know you are buying from a trusted leader in the industry. BioSupply offers:

- At-a-glance access to your account information
- Links to view open orders and ordering history
- Shortcuts to frequently purchased products
- FFF Sales Team contact information
- Detailed product pages
- Product alternatives if products are back-ordered or unavailable
- Convenience and accessibility to drop-ship products
- Shopping Cart feature displays account number and shipping address to minimize purchasing errors
- My Favorites feature for frequently ordered products
- BioVision reporting tool provides analysis of purchasing patterns

For ordering support, contact our Wow! Customer Care team:
P: (800) 843-7477 | Emergency Ordering available 24/7/365
F: (800) 418-4333
E: customerservice@ffenterprises.com
Want **priority access** on **FLU VACCINES** for the **2019-2020 season**?

Then take advantage of our NEW inclusive **MFV Loyalty Program** and enroll for the 2019-2020 season when you go to book your flu vaccine orders for the 2018-2019 season through MyFluVaccine.com.

- Priority access on advance released flu vaccines
- No minimum purchase
- Hassle-free
- Guaranteed booking for the products you need

**YOU PICK THE PREFERRED DATE • YOU PICK THE QUANTITY • WE DELIVER**

(800) 843-7477 | MyFluVaccine.com