Get Connected

Your Complete Resource for Advocacy, Education and Support

On IGLiving.com

Features an easy-to-navigate design

Indepth content on IG-treated diseases and treatment

Connect with our Patient Advocate, Abbie Cornett

Read weekly blogs about issues related to living with chronic illness

Valuable Resources and more
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
   We Can Accomplish
   More Together
   By Ronale Tucker Rhodes, MS

6  Abbie’s Corner
   Living Well
   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:
   T Lymphocyte Development in the Thymus
   By Terry O. Harville, MD, PhD

10 Clinical Brief
    ALS: Where Are We Now?
    By Michelle Greer, RN

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

Columns

40 Let’s Talk!—
    Whitney Ward
    By Trudie Mitschang

42 Patient Perspective —
    The New Reality:
    This Is How It Really Is
    By Stacy Oliver

43 Life as a 20-Something —
    How to Manage Treatment Locally
    By Ilana Jacqueline

44 Parenting — Improving
    Communication with Your
    Chronically Ill Teen
    By Jessica Leigh Johnson

Features

20 Making Connections in the PI Community
   By Trudie Mitschang

24 Planning for Retirement with Chronic Illness
   By Abbie Cornett

28 Managing Complications of Chronic Systemic Corticosteroid Use
   By Bob Geng, MD

32 Good’s Syndrome: A Rare Disorder Associated with PI
   By Ronale Tucker Rhodes, MS

36 Expanding Uses of IVIG
   By Ilana Jacqueline

Sources

46 Product Guide
    Flu Season Safety Tips
    By Trudie Mitschang

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 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. ©2018 FFF Enterprises Inc.
MANY PEOPLE shy away from asking for help even when help is really needed. The question is: Why do we insist on toughing it out even when help is needed most? None of us likes to feel needy or weak. But, asking for help isn’t a sign of weakness; instead, it can be a sign of strength because we can all accomplish more together than we can alone. And, you’d be surprised at just how much help is out there for people with chronic illness, ranging from social support to planning and medical assistance.

Indeed, social support can help to combat feelings of isolation so often experienced by individuals with a rare and chronic illness. Social support can be found in many forms, from in-person to online options. In our article “Making Connections in the PI Community” (p.20), we share some examples of networking opportunities offered by national organizations such as the Immune Deficiency Foundation, Invisible Diseases Association and others. But, more importantly, we provide some real-world advice from people who have overcome feelings of isolation with the help of social networking groups. Of course, there are pros and cons of support groups depending on the type, but ways to overcome those as well. And, there are alternative ways to connect with others, including volunteering and fundraising.

Individuals with chronic illness also can feel isolated when it comes to planning for their future such as retirement. With the added costs associated with illness, saving for retirement may seem unattainable. Our helpful article titled “Planning for Retirement with Chronic Illness” (p.24) outlines attainable steps that can be taken to ensure monetary resources will be available when needed.

It goes without saying, doctors can and should be looked to for help when it comes to the effects prescribed medications have on patients. For primary immunodeficiency patients, the common treatment with steroids to control inflammation can result in complications, especially when taken long-term. In our article “Managing Complications of Chronic Systemic Corticosteroid Use” (p.28), immunologist Bob Geng, MD, discusses those complications and how they can be lessened. As he points out, doctors usually make every attempt to limit steroid use to the lowest possible dose for the shortest duration possible. When such adjustments cannot be made, he suggests other options available to manage and minimize complications.

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

By Abbie Cornett

USING THE TERMS “living well” and “chronic illness” in the same sentence might seem like an oxymoron. A reader might think you have either lost your mind or don’t understand what these terms mean. Yet, while at first glance they may seem counterintuitive, that doesn’t mean they are mutually exclusive.

No doubt, you already know what “chronic illness” means because you live with it every day or with a loved one who is ill. But, you might not be aware of how “living well” can be achieved or why it is so important. Statistics show chronic illness has become a major concern in the United States. Currently, at least 133 million Americans have a chronic illness, and that number is projected to grow to 164 million (nearly half the population) by 2025.1

In recognition of these rising numbers, the Centers for Disease Control and Prevention and the Arthritis Foundation teamed up to seek help from the Institute of Medicine to find ways to reduce disability and improve patients’ health and quality of life. As part of their mission, they explored what the idea of living well with a chronic illness means and proposed a definition: “The concept of living well reflects the best achievable state of health that encompasses all dimensions of physical, mental and social well-being.”2

What does this definition mean for patients who have a chronic disease that affects all aspects of their lives — from their physical well-being and independence to their personal relationships and employment? The first step toward living well starts with letting go of negative feeling such as guilt, anxiety and frustration. While all of these feelings are normal, they can negatively impact wellness. Instead, find ways to move toward acceptance of your new normal, and find ways to live your life in the healthiest most fulfilling way. This means something different for each person, but some good basic steps are suggested in an article published in Psychology Today:3

• Learn as much as you can about your illness.
• Be active in your own treatment.
• Maintain a healthy diet.
• Seek support when you need it.
• Stay in contact with your spiritual side.
• Find gratitude.

Add to these the following advice from people with debilitating conditions like fibromyalgia, lupus and chronic fatigue syndrome, as well as from scientists, meditation experts and great thinkers:4

• Let go of the blame; it’s not your fault you are ill.
• Distinguish your illness from yourself.
• Address envy; instead, be happy for your friends and family who don’t have a chronic illness.
• Honor your limitations; protect your health.
• Connect with universal suffering; all people suffer in some way.
• Let go of expectations.
• Find your tribe — people with whom you can connect.

There is no denying living with a chronic illness is tough, but it doesn’t mean you can’t still enjoy life. Take care of yourself in all ways. Let go of the past, and embrace the now. Live well!

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

References

I am always looking for answers or information regarding my disease. I was first diagnosed with an immune deficiency about 25 years ago, and I was totally overwhelmed. One thing I have learned since then is one thing can lead to another. The original diagnosis was just a peek into what my life would be like. I can’t worry about what might be, but I can choose to be proactive about anything that may occur.

— Jenny G

A lot more needs to be published about autoimmune diseases — not only for patients but for the people in their lives. Because, many times, the disease is inside and not as noticeable from the outside, it is overlooked by people. The physical and mental suffering is difficult. It is common when one has been diagnosed with one autoimmune disease that others will follow.

— Judy S

Stress affects me more than I want to admit. Unfortunately, I internalize a lot of things, which just makes things worse. A New Year’s resolution for me this year is to try to relieve some of the stress in my life.

— Jenny G

How do you plan for travel?

My husband and I sold our home, cars and most of our belongings, and we travel the world living out of our suitcases. I have one suitcase dedicated to my pump, HyQvia, supplies packaged in baggies for each infusion, all of my meds (pills), etc. I have daily/weekly pill cases for four weeks, and the rest of my pills in original bottles with the prescriptions, and I write how many pills are in the bottles on the outside so I have the correct count for the entire trip. We check the country websites for rules on bringing in blood products like immune globulin, so we receive permission ahead of time and paperwork explaining why I need all the products I have. We plan our stays in the U.S. for doctor visits, refills, dental and eye care, etc. and it is all coordinated with respect to tests/blood work and checkups every six months. My prescriptions are written so I can get a maximum of six months of medication for travel. It takes a lot of forethought and planning, but it’s all possible! Definitely get the Global Entry [pass]; it’s worth being TSA pre-checked every time you fly and [getting] quick entry through customs upon returning to the U.S.

— Florence CJ

Do you understand diseases of the immune system?

I am always looking for answers or information regarding my disease. I was first diagnosed with an immune deficiency about 25 years ago, and I was totally overwhelmed. One thing I have learned since then is one thing can lead to another. The original diagnosis was just a peek into what my life would be like. I can’t worry about what might be, but I can choose to be proactive about anything that may occur.

— Judy S

Stress has a very serious effect on me. I have adrenal insufficiency due to long-term steroid use for myasthenia gravis. My adrenal glands do not protect me. Therefore, when I’m under extreme stress, I have to inject Solu-Cortef. Many times, I have been admitted to the hospital under very ill conditions. Stress can cause me to be extremely ill.

— Judy S

Stress affects me more than I want to admit. Unfortunately, I internalize a lot of things, which just makes things worse. A New Year’s resolution for me this year is to try to relieve some of the stress in my life.

— Jenny G
**Abbie** I consulted one of our reimbursement experts who advises MG may be covered by Medicare Parts B or D, depending on the circumstances. If you receive infusions in a hospital, outpatient clinic or physician office, there is coverage under Part B, but it may be limited to exacerbations only, rather than maintenance therapy. You would need to check with the reimbursement/billing department in those facilities for their assessment. If you receive infusions at home, treatment may be covered under Part D. Typically, though, Part D only covers exacerbations as well. However, our experts have had some success in getting maintenance therapy approved on a patient-specific basis for those who have tried and failed on oral medication therapy or when oral medication therapy is contraindicated.

**Question**
Are IVIG Infusions Covered by Medicare to Treat Myasthenia Gravis?

I have myasthenia gravis (MG), and I have been receiving intravenous immune globulin (IVIG) infusions since 2006. My husband’s insurance has been covering treatment, but he wants to retire. I have Social Security Disability and will have to go on Medicare if he retires. According to an article in your October-November 2017 issue of IG Living, IVIG therapy is not covered by Medicare to treat MG. Is that correct?

**Abbie** Our reimbursement expert points out that SCIG therapy is only performed in the home setting. The reason the insurance company requires additional testing for SCIG therapy is probably due to the fact that specialty pharmacies supplying the medication are much more proactive about obtaining approval prior to treatment. In comparison, most doctor offices don’t require prior approval before treatment. But, that doesn’t mean the payer cannot deny or even recoup payments made for these medications. If you would prefer not to have to undergo new testing, you may want to consider continuing with intravenous IG (IVIG) therapy.

**Question**
Why Is Additional Testing Required When Changing from IVIG to SCIG?

I have been diagnosed with common variable immunodeficiency, and my insurance company has approved immune globulin (IG) therapy in my doctor’s office. I would like to be trained to perform subcutaneous IG (SCIG) infusions at home, but my insurance company is requiring me to have a pneumonia vaccine and to wait until testing is performed again before approving it. Why does it make a difference where I get treatment? How can they approve me for treatment in the doctor’s office and not at home?

**Abbie**

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.
THE THYMUS IS critically important for T lymphocyte development. When the thymus does not develop appropriately, immunodeficiency results. When it does develop appropriately, people are protected from infections and disease.

In the previous issue, we discussed the function of the T-cell receptor (TcR) that enables each unique T lymphocyte to recognize and bind to a specific antigen. Recombination of a specific set of genes results in the formation of millions of unique TcRs (each T lymphocyte has its own unique TcR). TcRs recognize and bind to specific antigens to activate T lymphocytes, which initiates an immune response. Among these TcRs will be some that are useless, some that will bind too tightly to self-antigens, and some that bind correctly to specific self-proteins known as human leucocyte antigens (HLAs). The role of the thymus is to select the correct repertoire of T lymphocytes based on the ability of each TcR to bind correctly with a person’s HLA proteins. I call this the “Goldilocks’ Rule.” When the TcRs bind correctly to the self-HLA proteins, this is known as positive selection. When they cannot bind to the self-HLA proteins, it is known as fail positive selection and they die from neglect (these are the useless ones).

To prevent autoimmunity, the thymus helps to eliminate developing T lymphocytes that bind too tightly to self-proteins. This is known as negative selection, and it occurs when a specific gene known as AIRE becomes activated in thymus cells. AIRE activation causes every person’s genes (some 21,000) to be expressed, which exposes the developing T lymphocytes to every protein an individual can produce. Thus, every self-antigen is then presented by the HLA proteins of the thymus to the developing T lymphocytes. Those that bind too tightly activate to self-proteins, which could result in autoimmune reactions and disease. But, if the appropriate response occurs, these T lymphocytes are induced to die by a process called apoptosis. In contrast, those that bind correctly survive and are selected as potentially useful to protect against infections and disease. By this process, T lymphocytes are educated to recognize self from nonself, and have adhered to the Goldilocks’ Rule.

More than 95 percent of all thymocytes (developing T lymphocytes) that produce a TcR will die since they are useless or bind too tightly to self. This efficient and effective quality control provides a useful repertoire of mature T lymphocytes to protect people, as well as prevents potentially harmful T lymphocytes from persisting.

Since most T lymphocyte development occurs in the developing fetus between about 12 weeks and 16 weeks of gestation, this illustrates why disruption of thymus formation during early fetal development results in the T lymphocyte immunodeficiency of DiGeorge syndrome. On the other hand, when this development functions correctly, people are born with a full repertoire of T lymphocytes that can provide immune protection for life. Consequently, some have thought the thymus has no further purpose after birth. However, we now know the thymus remains active, producing and educating new T lymphocytes for the entire life span, but at a much slower rate as people age. Fortunately, the presence of thymus activity throughout life allows for continuous development of new T lymphocytes to protect against the next new pathogen on the horizon.

In the next issue, we will discuss treatment of DiGeorge syndrome with thymus transplantation.
AMYOTROPHIC LATERAL SCLEROSIS (ALS) is a progressive neurological disorder affecting the motor neurons that control voluntary muscles responsible for walking, talking, swallowing and breathing. ALS is also called Lou Gehrig’s disease, named for a professional New York Yankees baseball player who was diagnosed with ALS in 1939 and died from it in 1942. There is no known cause for ALS, and there is no cure. Indeed, the disease is always fatal, usually in three to five years of onset. However, there have been some advances in treatments, and palliative care can extend life expectancy and improve quality of life.

Symptoms of ALS
Initial symptoms of ALS can mimic other neurological disorders. And, since there is no definitive test for ALS, it is critical to rule out other conditions to ensure an accurate diagnosis. It is not uncommon for ALS to be misdiagnosed, nor is it uncommon for someone to be diagnosed with ALS when suffering from another neurological condition. One of the more common conditions ALS mimics is multifocal motor neuropathy (MMN). Both begin with weakness in one limb and fasciculations (muscle twitching) that can occur throughout the body. However, MMN has some distinct differences upon neurological assessment, and it is treatable, typically with long-term immune globulin therapy.

ALS affects the muscles needed to move and, eventually, will affect those required to speak, eat and breathe. In addition to fasciculations, muscle cramping may be present. To diagnose ALS, various electrodiagnostic studies on nerves and muscles are conducted such as an electromyogram, a nerve conduction study and an MRI. Blood, urine and spinal fluid are also tested.

Treating ALS
Many physicians specialize in ALS, and most are located at ALS clinics and in academic settings, the majority of which are recognized by the ALS Association and designated as certified treatment centers of excellence, recognized treatment centers or other affiliated clinics. The differences between these sites are the service and clinical trial offerings. A list of these centers can be found at www.alsa.org/community/centers-clinics.

Various treatments are currently available during the different stages of disease progression. Early on, assistive devices such as a cane, walker or wheelchair may be needed. As the disease progresses, chewing and swallowing become more difficult, so dietary guidance is important. At some point, the patient and family may opt for a feeding tube. As mobility becomes more of an issue, attention needs to be paid to skin care, and physical and occupational therapy will need to be implemented. Speech therapy will be needed when speech becomes affected. There are also a range of assistive devices available to aid in communication.

Increasing difficulty with breathing is probably the most frightening symptom people with ALS experience. As the muscles in the chest weaken, they lose elasticity, and it becomes more difficult to pull air into the lungs. This can be uncomfortable, scary and interfere with sleep. Pulmonary function needs to be monitored consistently, and at some point, decisions should be made about various options that will provide assistance (see Options for Assistance with Breathing). It is up to the patient and family to determine how much assistance is desired.

Options for Assistance with Breathing
- Lung expansion exercises
- Noninvasive ventilation machines:
  - Bilevel positive airway pressure (BiPAP)
  - Average volume assured pressure (AVAP)
- Invasive ventilation via tracheostomy

More details on these options can be found at swallowingsystemscen.org/wp-content/uploads/2017/06/Living-with-ALS__Breathing.pdf.
Currently, many clinical trials are in progress with the aim of slowing disease progression or finding a cure. Clinical trials are imperative to determine viability of a treatment option. Unfortunately, many neurologists who specialize in treating patients with ALS see patients spend large sums of money on therapies that may not work, some of which even quicken disease progression. A list of trials can be located at www.alsa.org/research/clinical-trials and www.clinicaltrials.gov.

Facilities all around the world are conducting stem cell therapy trials for ALS. One theory of a possible cause of ALS is excess glutamate (an amino acid) around nerve cells, which creates an unhealthy environment for nerve cells, causing them to die. It is believed stem cells can be used to create healthier conditions that will eliminate excess glutamate.

One stem cell study currently recruiting is testing a therapy called NurOwn, an investigational therapy from BrainStorm Therapeutics. With NurOwn, autologous bone marrow-derived mesenchymal stromal cells, which are enriched from the patient’s own bone marrow, are transplanted, propagated ex vivo and induced to secrete neurotrophic factors. This means a specific type of stem cell from one’s own bone is harvested and encouraged via medical technology to develop into cells that promote the growth of nervous tissue.

In addition to treatments in clinical trials, two medications are currently approved by the U.S. Food and Drug Administration (FDA) to treat ALS. Riluzole (Rilutek) may slow disease progression in certain people by reducing glutamate levels. Riluzole comes in a pill and can cause dizziness and gastrointestinal upset, as well as affect liver function. Edaravone (Radicava) may reduce functional decline. FDA approved edaravone based on results of a clinical trial conducted in Japan that showed positive results. However, this therapy is a bit more complex. It is intravenously administered every day for 14 days each month. Side effects may include bruising at the IV site, and allergic-like reactions such as hives, swelling and shortness of breath. It also contains sodium bisulfite that can cause allergic reactions in people who are sensitive or allergic to sulfites.

Hope for the Future
ALS is a devastating disease for which there remains no cure. Fortunately, more treatment options are becoming available that can be individualized to the wishes of patients and their families. In addition, many patients agree to participate in clinical trials as a means of hope for their own condition and, selflessly, as a mission of hope for the future health of others.

For additional information on ALS and resources available, contact the ALS Association at www.alsa.org. 

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

Reference
IN THE NEWS

Initiative

PPTA Unveils ‘How Is Your Day?’ Global Initiative to Raise Awareness of Plasma Protein Therapies

In March, the Plasma Protein Therapeutics Association (PPTA) unveiled its new global initiative titled “How Is Your Day?” — Making the Difference with Plasma Proteins in conjunction with its annual International Plasma Protein Congress. The campaign is focusing on differentiating these unique therapies from traditional pharmaceuticals and building awareness on the value they provide for people living with rare, life-threatening, chronic and genetic diseases.

“How Is Your Day?” is a global initiative designed to unite people treated with plasma protein therapies worldwide. It focuses on individuals who live with these rare diseases and highlights the lifesaving difference that plasma protein therapies make in their lives by sharing their stories. Specific goals of the campaign include 1) a call for worldwide availability and access to safe and effective plasma protein therapies for all who need them, 2) supporting policies to improve patients’ access to therapies, 3) highlighting patient options and the unique role of plasma protein therapies and 4) highlighting the value, to patients and society, of plasma protein therapies.

“Access to plasma protein therapies is a critical issue for individuals facing these diseases because most of them can only be treated by medicines that are made from plasma donated by healthy, committed donors,” said Jan M. Bult, PPTA president and CEO. “While treatment options exist for individuals with bleeding disorders, many others use therapies derived from human plasma. ‘How Is Your Day?’ will discuss these differences and build greater awareness and understanding of the importance of plasma protein therapies for individuals and their families.”


Research

Study Shows SCIG Rapid Push Administration Less Preferred But More Cost-Effective

A study that compared the impact of pump and rapid push (syringe) subcutaneous immune globulin infusions on patients’ life quality index (LQI) showed the rapid push administration is preferred by some patients and is a cost-effective alternative to administration of larger volumes by pump once a week. Pump infusions are performed once a week and last approximately one hour, while rapid push decreases the duration of administration but requires more frequent infusions.

In the study, 28 primary immunodeficiency disease (PI) patients accustomed to weekly infusions at home by pump used either a pump or rapid push for three months each according to a randomized sequence for two periods. The main criterion was PI-LQI factor 1 (treatment interference), and the noninferiority ratio was set at 90 percent.

At the end of each period, the mean LQI factor 1 was 87.0 and 77.80 for pump and rapid push, respectively. There was a slightly larger effect of rapid push on treatment interference than with the pump so the primary endpoint could not be met. No difference was found on other LQI components, satisfaction or quality of life. Eight patients declared to prefer rapid push, while 19 others preferred the pump. Of rapid push infusions, 67.2 percent led to local reactions versus 71.8 percent of pump infusions. Both achieved similar trough IgG levels with similar incidence of infections. Rapid push, however, saved 70 percent of administration costs when compared to a pump.

IN THE NEWS

Research
Study Looks at Patient Satisfaction with Biweekly Hizentra Treatment

A study that investigated the efficacy of biweekly Hizentra compared with previous intravenous immune globulin (IG) or subcutaneous IG treatment regimens in patients with primary immunodeficiency (PI) disease found patients were either very (76.5 percent) or quite (23.5 percent) satisfied with Hizentra treatment. In the 12-month retrospective study followed by a 12-month prospective observation period, the main endpoints included pre-infusion IgG concentrations, proportion of serious bacterial infections, other infections, hospitalizations due to PI-related illnesses and days with antibiotics. Of the 35 patients in the study (mean age 26.1 plus or minus 14.4 years, 68.6 percent male), the mean pre-infusion IgG levels for prior IG regimens during the retrospective and prospective periods did not show any significant variations, the mean annual rate of SBIs per patient was 0.063 plus or minus 0.245, and there were no hospitalizations. According to the researchers, the “study provided real-world evidence on the efficacy of biweekly Hizentra in patients with PIs.”


Research
Study Pinpoints Rare Gene Mutation That Causes Primary Immunodeficiencies

Researchers have pinpointed rare mutations in the receptor interacting serine/threonine kinase 1 (RIPK1) gene that cause primary immunodeficiencies (PIs). The RIPK1 gene is essential in mediating cellular functions that stimulate immune cells responsible for fighting infections and mitigating the body’s reaction to inflammation. The scientists from the King Saud bin Abdulaziz University for Health Sciences and Alba University in Saudi Arabia made the discovery after sequencing the genomes of four patients suffering normal symptoms of PI such as viral and bacterial infections, as well as inflammation of the bowel and joints.

“Although RIPK1 has been extensively studied in animal models, we uncover, for the first time, its role in humans,” said the study’s lead investigator Sergey Nejentsev at the Department of Medicine of the University of Cambridge in the United Kingdom. “While RIPK1 deficiency in mice typically leads to multiple system defects, the effect of its deficiency in humans seems to be nearly exclusive to the immune system.” According to Nejentsev, their findings indicate transplantation of blood stem cells “can be an effective treatment for [PI] patients if performed early in life.”


Research
New 10% IVIG Safe and Effective in Treating ITP

A recent study found a new intravenous immune globulin (IVIG) 10% formulation was found to be safe and effective in adult primary immune thrombocytopenia (ITP) patients. The study included 81 patients aged 19 years or older (31 of whom were newly diagnosed, seven of whom had persistent ITP and 43 of whom had chronic ITP) with a platelet count of less than 20 x 10^9/L within two weeks of the start of study. Patients received the IVIG 10% at a dose of 1 g/kg per day for two consecutive days, and response was defined as achieving a platelet count of greater than or equal to 50 x 10^9/L at day eight. Results showed 61.3 patients (75.7 percent) achieved response and satisfied the predefined noninferiority condition. Median time to response was two days, and mean duration of maintaining response after completion of IVIG therapy was 9.13 plus or minus 8.40 days. Response rates were found to be dependent on the phase of ITP or previous treatment for ITP. The therapy was well-tolerated, and the frequency of mucocutaneous bleeding decreased during the study period.

Research

IVIG Safe and Effective for Treating SFPN

A study conducted to assess the safety and efficacy of apparently autoimmune small fiber polyneuropathy (aaSFPN) has found intravenous immune globulin (IVIG) reduces pain and improves organ function. The researchers hypothesized that small-fiber-targeting autoimmune diseases akin to Guillain-Barré and chronic inflammatory demyelinating polyneuropathy, for which IVIG is often prescribed off-label, could be a cause of aaSFPN. The study included 55 patients with aaSFPN in whom 27 patients had systemic autoimmune disorders, 20 percent had prior organ-specific autoimmune illness and 80 percent had abnormal blood-test markers of autoimmune immunity — but none had diabetes or other known cause of neuropathy. After being treated with IVIG for an average of 28 months, 77 percent of patients responded to treatment, with pain dropping on average from 6.3 to 5.2 on a 10-point scale, and internal organ function improving.

The researchers concluded the study provides proof-of-concept evidence that IVIG is safe and effective for rigorously selected SFPN patients with autoimmune causality, providing rationale for more trials.

Survey

AARDA Releases Findings from Autoimmune Disease Survey

A survey conducted by the American Autoimmune Related Diseases Association (AARDA) has found the vast majority of autoimmune disease (AD) patients do not believe U.S. federal and state elected officials understand that autoimmunity is a major U.S. health issue. The survey of 1,287 AD patients was conducted as part of AARDA’s annual “March is Autoimmune Disease Awareness Month” activities.

Major findings of the survey include:

• Ninety-four percent agree that “federal autoimmune disease research is significantly underfunded ($821 million) when compared to cancer ($5.4 billion) and heart disease ($1.7 billion). Increasing federal funding for autoimmune research should be a top national healthcare priority for the president and members of Congress.”

• A large majority are concerned that the following areas of healthcare policy will be an issue in the future, and nearly all believe these issues should be considered as the Affordable Care Act (ACA) is repealed and replaced, including: pre-existing conditions (75 percent and 91 percent, respectively); high cost/copays for medicine (78 percent and 93 percent, respectively); high insurance premiums/deductibles (76 percent and 92 percent, respectively); access to specialists (64 percent and 89 percent, respectively); and narrow provider networks (60 percent and 85 percent, respectively).

• Eighty-nine percent believe the president and Congress should work together to create legislation to prevent insurance companies from utilizing “nonmedical switching,” “step therapy” and other similar practices that take patients’ healthcare decisions out of the hands of their doctors.

• More than three-quarters do not believe the president, U.S. Senators and Congressmen, and local state elected officials are aware that there are more than 100 known autoimmune diseases (76 percent, 79 percent and 82 percent, respectively); autoimmunity is a category of disease like cancer and heart disease (75 percent, 78 percent and 80 percent, respectively); autoimmune disease represents a major U.S. health issue impacting 50 million Americans (77 percent, 79 percent, 81 percent, respectively); autoimmune disease is one of the top-10 causes of death for women under 65 (81 percent, 82 percent and 82 percent, respectively); and the economic impact of autoimmune disease represents more than $100 billion in annual direct healthcare costs (77 percent, 79 percent and 80 percent, respectively).

“Autoimmune disease patients suffer from a variety of serious, chronic illnesses that need careful and constant coordinated management, usually by specialists. Their concerns about changes to the ACA or any healthcare policy moving forward are real since these changes can have a devastating impact on their lives,” said Virginia T. Ladd, founder and executive director of AARDA. “As we begin to plan for our next 25 years of work, the survey also shows us that we have much work to do educating those who hold all the proverbial cards about autoimmune disease.”
Research

MG Impairment Index May Be Useful for Clinical Trials in Pure Ocular Disease

A new study has found the Myasthenia Gravis Impairment Index (MGII) is a sensitive tool for identifying treatment response and clinical meaningful change among patients with MG receiving prednisone, intravenous immune globulin (IVIG) and plasma exchange (PLEX). In the study, 95 patients with MG who were prescribed prednisone, IVIG or PLEX were compared with 54 control patients who received no treatment. Assessments using the MGII, Quantitative MG Score, MG Composite and MG Activities of Daily Living occurred at baseline and at three to four weeks after treatment. Those receiving prednisone, IVIG or PLEX demonstrated a significantly greater change in MGII scores. Specifically, patients receiving prednisone showed more change in the ocular domain when compared with IVIG/Plex, and in the generalized domain, those receiving IVIG/PLEX showed greater change in scores compared with those who received prednisone. According to the researchers, the results may be useful for clinical trials in pure ocular disease.


SUB-Q Needles and Skin Force Penetration

What Does it Really Mean to Patients?

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.

1264 Hawks Flight Court, Suite 200, El Dorado Hills, CA 95762 USA
TELEPHONE: 916.932.0071 | FAX: 916.932.0074
www.emedtc.com | sales@emedtc.com
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.
Immediately report to your physician any of the following symptoms, which could be signs of serious adverse reactions to Hizentra:

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

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

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

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

Please see 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.

Why Choose Hizentra?

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

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

Proven safety
Hizentra has an established safety profile and demonstrated tolerability. In clinical trials, the most common side effects were redness, swelling, itching, and/or bruising at the infusion site; headache, chest, joint or back pain; diarrhea; tiredness; cough; rash; itching; fever, nausea, and vomiting. These are not the only side effects possible.

Discover all the benefits Hizentra has to offer at Hizentra.com

Choose where you infuse

No IV infusions

Proven safety

Why Choose Hizentra?

Choose where you infuse

No IV infusions

Proven safety

Discover all the benefits Hizentra has to offer at Hizentra.com
HIZENTRA®, Immune Globulin Subcutaneous (Human), 20% Liquid
Initial U.S. Approval: 2010

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

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

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

INDICATIONS AND USAGE

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

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

For subcutaneous infusion only.

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
Research

Baking Soda May Help to Combat Autoimmune Disease

An animal study conducted by Medical College of Georgia (MCG) scientists has shown that drinking a daily dose of baking soda may help reduce the destructive inflammation of autoimmune diseases such as rheumatoid arthritis. The scientists found that by drinking a teaspoon of baking soda mixed with a half liter of tap water for two weeks, the population of immune cells called macrophages in the spleen, blood and kidneys shifted from primarily those that promote inflammation, called M1, to those that reduce it, called M2. Macrophages, best known for their ability to consume waste in the body like debris from injured or dead cells, call for an immune response early on. In the study, the lab animals’ problems were hypertension and chronic kidney disease. “Certainly, drinking bicarbonate affects the spleen, and we think it’s through the mesothelial cells,” said Paul O’Connor, MD, a renal physiologist in the MCG department of physiology and the study’s corresponding author.


Research

Study Finds Dermatomyositis Requires a Second-Line Treatment

Researchers at the University of Pennsylvania have found second-line agents need to be incorporated into treatment for moderate to severe dermatomyositis (DM). The study assessed the impact of care using a treatment algorithm to determine systemic treatment for 41 patients with skin-only DM (with clinically amyopathic DM and no lung involvement) who were treated between July 2009 and April 2013.

First-line treatments for skin-only DM are antimalarial medications hydroxychloroquine (HCQ), quinacrine and chloroquine. HCQ is administered to most patients for eight weeks, quinacrine is added if HCQ is ineffective, and chloroquine is used instead of HCQ in those in whom HCQ is ineffective or who have experienced a reaction to it previously. A second-line treatment such as methotrexate, mycophenolate mofetil or azathioprine are added if symptoms progress despite using antimalarial agents. And, if adequate control is not achieved within another eight weeks, patients are switched to another cytotoxic agent. Third-line agent intravenous immune globulin (IVIG) administered over two to five days each month is considered in patients who still do not respond, followed by oral calcineurin inhibitors. While systemic corticosteroid medications are used only to provide relief in cases with severe disease, topical agents such as corticosteroid or immunomodulator medications are commonly used.

In this study, 23 of the patients received antimalarial medications alone and 18 patients received second- or third-line agents. Ten (24.4 percent) patients were treated with HCQ alone, 22 percent were treated with HCQ and quinacrine, 4.9 percent were treated with chloroquine and 2.4 percent were treated with chloroquine and quinacrine. Initial disease severity and outcomes were assessed using the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI). After a median treatment duration of 24 months, just 14.6 percent of patients were managed with HCQ alone. The median final CDASI score for all patients was 13.5, with most patients failing to experience complete resolution of skin symptoms and showing mild disease activity at the final visit. Eleven of the patients received prednisone, which may have contributed to clinical improvements. Good results were seen in a small number of most refractory patients treated with IVIG. According to the researchers, “the results show that management of cutaneous DM often requires second-line agents because antimalarial medications alone are insufficient to treat most patients with skin-only disease.”

Making Connections in the PI Community

When living with chronic illness, you don’t have to go it alone; a support or networking group can help. From in-person to online options, choose the one that’s right for you.

By Trudie Mitschang
PRIMARY IMMUNODEFICIENCY diseases (PIs) affect people in different ways, but like anyone living with a chronic illness, individuals living with PI tend to battle feelings of isolation. In addition to seeking out the most effective treatment plan, it is vital to identify ways to reach out to others who are facing similar challenges. According to the Immune Deficiency Foundation (IDF), it is particularly important to build and maintain strong relationships, both inside and outside your immediate family, and to remain connected socially: “Individuals living with chronic illnesses, especially unusual or rare disorders such as primary immunodeficiency diseases, often feel isolated and that they are struggling alone. Contact with other individuals who live with these diseases is a way to both gather knowledge and acquire an important sense of connection with others who share your experience.”

Networking with National Organizations

An excellent place to begin looking for patient-to-patient connections is through recognized organizations created specifically for PI patients. IDF offers a number of programs and resources to get you started, including:

Peer Support Program. This program gives you the opportunity to interact with one of IDF’s trained volunteers who has personal experience living with PI. This free resource is for anyone personally affected by PI, including patients, parents or other family members, friends or caregivers.

IDF Friends. This private social network and discussion forum is exclusively for patients and family members who live with PI. The online community gives people affected the opportunity to offer and receive peer support, ask questions, make suggestions, share their stories and connect with others through words, pictures and video.

Get Connected Groups. The IDF Get Connected Groups are designed to connect individuals diagnosed with PI and family members in their local communities. The meetings can occur at a local community room, library, coffee shop or other venue. Through the groups, individuals and families can connect to share experiences, receive information and gain support. These groups do not include medical presentations or industry exhibits.

Common Grounds. This is a private social network and discussion forum exclusively for PI teens. This online community gives teens the opportunity to offer and receive peer support, ask questions, make suggestions, share their stories and connect with others.

Contact IDF at (800) 296-4433 for information on these and other opportunities to get involved at the local, regional and national levels.

Pros and Cons of Traditional Support Group

For many people, a health-related support group may fill a gap between medical treatment and the need for emotional support. While support groups may be offered by a nonprofit advocacy organization, clinic or hospital, they may also be run entirely by group members.

Formats of support groups vary, including face-to-face meetings, teleconferences or online communities. A lay person — someone who shares or has shared the group’s common experience — often leads a support group, but a group also may be led by a professional facilitator such as a nurse, social worker or psychologist. Some support groups may offer educational opportunities such as a guest doctor, psychologist, nurse or social worker to talk about a topic related to the group’s needs.

An excellent place to begin looking for patient-to-patient connections is through recognized organizations created specifically for PI patients.

One of the advantages of a traditional support group is the common experience among members often means they have similar feelings, worries, everyday problems, treatment decisions or treatment side effects. Participating in a group provides you with an opportunity to be with people who share common challenges and goals.

Benefits of participating in a support group include:
- Feeling less lonely, isolated or judged;
- Reducing distress, depression, anxiety or fatigue;
- Talking openly and honestly about your feelings;
- Improving skills to cope with challenges;
• Staying motivated to manage chronic conditions or stick to treatment plans;
• Gaining a sense of empowerment, control or hope;
• Improving understanding of a disease;
• Getting practical feedback about treatment options; and
• Learning about health, economic or social resources.

It’s important to note support groups, regardless of format, can have drawbacks as well. Common concerns arise from disruptive or domineering members, an excessive amount of complaining, lack of confidentiality, spreading of unsound medical advice and competitive comparisons of whose condition or experience is worse.

A strong and experienced facilitator can usually help the group steer clear of these types of pitfalls. Before joining a group, ask about the facilitator’s qualifications or training, clarify the confidentiality guidelines and then try it out for a few weeks. If it doesn’t feel like a good fit for you, consider a different support group or a different support group format such as online.

**Exploring Online Support**

Online support groups and social media pages offer a sense of community and connection that can alleviate feelings of isolation, especially for those who are housebound by illness and/or who do not live in close proximity to local networking groups. Many have discovered immediate connections and lifelong friendships thanks to the wonders of social networking. “Social media groups have been the best for me,” says Dona Darr, whose 16-year-old daughter lives with PI. “They are a wealth of information regarding the realities of dealing with the disease and navigating insurance, schools, etc. They can also give you perspective; through online networking, I learned there are many patients who deal with more severe challenges than the ones my daughter and I have faced.”

For Jessica Leigh Johnson, an IG Living columnist, blogger and mother of three boys with X-linked agammaglobulinemia (XLA) and one who passed away from XLA, social media has led to both online and real-world connections: “We’ve connected to families with our diagnosis through the XLA Facebook group. It’s an excellent way to connect with people who share your struggles. There are members from all over the world, so it’s not hard to find people who live in the same state or who even see doctors at the same hospital you do.”

Health writer Samantha Gemmell agrees: “When you are first diagnosed with a rare condition, Facebook groups can be a godsend. They are filled with people who understand, who can offer everything from a great specialist and day-to-day survival tips to someone to talk to when insomnia hits at 3 a.m. You are no longer lost in the storm. There are others to hold your hand and tell you ‘yes, that is a normal symptom,’ and ‘no, it’s not ridiculous to want to throttle your mother/sister/in-laws/friends when they say insensitive things or wonder about the validity of your condition.’”

Gemmell advises there are also downsides to relying on social media networking, noting members have a tendency to “overshare.” Differing opinions on treatment plans can also lead to unhealthy online bickering and even bullying that can leave you feeling angry, misunderstood and discouraged.

“We all have pride in what we can achieve and feel a greater connection to those who have faced similar circumstances,” says Gemmell. “But, when you post daily updates on symptoms, medications and bad days, you might be constantly focusing on your condition. Where you focus is where your energy goes, and chronic illness is a black hole for energy, constantly sucking up every little dreg.” In the end, Gemmell says she “unfollowed” a majority of the Facebook pages that initially offered a lifeline in the early stages of her illness, and today follows only those pages and resources that leave her feeling uplifted and inspired.

If you want to get started in the social network world, you can begin by using Google or another search engine to check for pages featuring the name of your diagnosis. To connect to larger chronic illness communities, check out one of the following support group pages, or follow them on Facebook, Twitter or Instagram:

**Invisible Disabilities Association (IDA).** IDA’s mission is “to encourage, educate and connect people and organizations touched by illness, pain and disability around the globe.” You can locate the association on Facebook and Instagram.

**I Told You I Was Sick Support Group.** This is a small closed support group, meaning you need permission from the site
administrator to join, and only approved members can view posts. If you are looking for an intimate and safe group to test the waters, this might be for you.

CrazyBoards.org. If you like your support groups more sarcastic and less politically correct, this site might fit the bill. It is primarily centered on mental health concerns, but it does have a space for chronic pain support. It also features an active chat room.

TreatmentDiaries.com. This is a good support group for people living with chronic pain. You connect with others by writing diary entries and waiting for those with similar experiences to comment on them. The site also allows you to list the specific conditions you’re living with, so you can get more personalized support.

But You Don’t Look Sick. Started in 2003, the mission of ButYouDon’tLookSick.com is “to help everyone with a chronic illness or invisible disability, in order for them to live their lives to the fullest and not feel isolated and alone.” The founder originated the “spoon theory” commonly referred to by chronic illness patients.

Volunteering, Fundraising and Other Ways to Connect

Volunteering for an organization that provides support for your particular illness can be a very positive way to meet others in the PI community, while also giving you a sense of purpose and accomplishment. Begin by doing some online research to identify opportunities that fit your particular needs, desires and physical limitations.

Many organizations offer various types of volunteer options, including working one-on-one with other patients, being part of a volunteer team, assisting a paid staff member or becoming a peer support mentor.

IDF offers a nationwide network of volunteer opportunities designed to increase awareness of PI. Depending on your time and interests, IDF can connect you with others who are passionate about everything from political advocacy and fundraising to educational awareness and even plasma center support.

If you are physically able, an awareness walk can also be a great way to connect with others in the PI community. IDF sponsors national Walk for PI events in major cities around the country. For those unable to participate in person, the organization also offers a “virtual walk” that gives you the opportunity to raise funds and partner with others who share your passion for raising awareness. “I have only been involved with IDF for two years, and the first thing I did was participate in one of their PI awareness walks,” says PI patient Whitney Ward. “That was the first time I saw there were people out there like me who completely understood what I was going through. Our diseases may have been a little different, but there were more similarities.”

Another national event that provides networking opportunities is the annual Invisible Disabilities Week (IDW) hosted by IDA each year in October. All events are free, and the IDA website offers numerous suggestions for how to get involved, including downloadable social media badges, IDA wristbands and accessories, photo and video contests and fundraising ideas. Each year, the organization also hosts a fundraising gala. Learn more at invisibledisabilities.org.

Reaping the Rewards of Networking

There are many national and online resources whose sole purpose is to help PI patients connect with one another. For those who have been recently diagnosed, reaching out can feel daunting, but the rewards can be significant. Sometimes it’s best to start small by asking your local healthcare provider, pharmacist or patient advocate to assist. “When our boys were first diagnosed, our specialty pharmacy actually set us up with three other patients/families with the same diagnosis,” says Johnson. “The pharmacy asked them if it would be OK if I called them, and they agreed. One mother of three XLA boys had also lost a child under the age of 1, so I felt an instant connection with her since we had so much in common. I emailed her whenever I had questions, and I was able to meet her in person when we were traveling on the East Coast where she lives.”

For Ward, who has lived with a very rare type of PI since infancy, networking and becoming a part of a larger PI community brought a sense of emotional healing. “To know there are others who lived with the same medical issues I did made me feel less like an anomaly. I could just talk about what I was feeling and what I had gone through, I didn’t have to explain because they just knew. I gained a community of friends I never knew existed. To go from not knowing anyone similar to me to finding out there is a foundation that caters to your type of disease — it gave me joy, closure and an acceptance I had never experienced before.”

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.

References
Planning for Retirement with Chronic Illness

It can be difficult to plan for retirement with the added expenses of a chronic illness, but there are many steps you can take to ensure you are prepared.

By Abbie Cornett

IF YOU ARE like many Americans, you may not be preparing for retirement. A survey conducted by *Money* magazine in 2016 found one out of every three Americans has no savings, and 23 percent have less than $10,000 saved. This figure is particularly alarming since retirement is one of the biggest life expenses, even without a chronic illness.

Many expenses get in the way of saving money such as credit card debt, student loans, low wages and children. But, for those with a chronic illness, saving for retirement is even harder since a large part of your income goes to cover expensive insurance plans, doctor visits, medicines and medical supplies. Furthermore, if you are unable to work or can’t work full time due to your illness, there often isn’t enough money to save.

Actually, the population dealing with chronic illness and its expenses is larger than most people realize. According to statistics, 133 million Americans have at least one chronic illness. And, it is estimated by 2025, that number will have grown to 164 million people or nearly half the population. But, that doesn’t mean those with chronic illness can’t prepare for retirement. Following are some steps to getting started in the right direction.
Steps to Preparing for Retirement

1. The first step: Talk to your doctor. Your doctor can advise you on the likely progression of your disease and whether you will need special medical services, equipment or food. For example, if you have a neuromuscular illness, this may mean planning to make your home handicap-accessible in the future. Your doctor can also advise if you might have to reduce your work hours in the future or retire early due to your illness. This information is vital for helping you set a budget to plan for projected costs.3

2. Determine how much retirement money you will need. After you have learned as much as possible about the expected progression of your disease, it’s time to determine how much money you will need for retirement. While this may sound obvious, it isn’t. According to a study conducted by Age Wave and Merrill Lynch, 81 percent of Americans say they don’t know what they will need to fund their retirement.4

A common guideline for retirement planning is to aim for replacing 70 percent of your annual preretirement income. This amount can consist of savings, investments, Social Security wages and any other income sources such as a pension and part-time employment. A recent retirement survey estimates the average amount needed to retire is $738,400 with $260,000 of that amount allocated toward healthcare costs. Of course, this amount will differ from person to person based on individual needs and lifestyles.5

While close to $750,000 sounds like a lot of money to save, think about it in these terms: You don’t have to save that much by the time you retire, but you do need to save enough so that amount can grow to that total. For example, if you are 30 years old and want to retire at age 70, you have 40 years to save. Assuming an average rate of return of approximately 7 percent per year, you’ll need to save $5,463 per year or $738,400 by the time you are age 70. That’s just more than $455 per month.5

This nest egg will give you about $30,000 of income per year. To determine what your total income will be, add what you expect from Social Security wages each year, and adjust the amount. The Social Security Administration website has a number of calculators to help you estimate your benefits (see Retirement Planning Related Websites).

When deciding how much you need to save, remember that numbers are averages. You will need to estimate the age you plan to retire, as well as any additional amount you will need to take care of your illness. Similar to the Social Security calculators, there are a number of good retirement calculators you can utilize to help reach your goals.

3. Start saving. Next is to determine how are you going to reach your goal. According to Martin Shenkman, a CPA, attorney and author of Estate Planning for People with a Chronic Condition or Disability, patients should “simplify all of their finances by consolidating everything they can.” This means getting rid of unneeded bank accounts, setting bills to be automatically paid and uploading important documents. If all your information is consolidated in one place, it will be easier for you or your financial planner to manage. By making these simple changes, you can empower yourself to plan for the future and free up valuable time to concentrate on your health. Clearly, everyone’s circumstances are different, but that doesn’t mean the basic guidelines for planning for retirement are. The first step is to create a reasonable budget based on your individual needs and lifestyle. When creating the budget ask yourself these questions:6

- What is my illness going to demand (i.e., specialized medical equipment, home remodeling, home healthcare)?
- How much money do I need to be saving each month?
- How do I protect my future?
- How much do I need to include to enjoy life now?

Because of the unpredictability of chronic illness, there are going to be times when you have to revise your budget as needs change.

4. Stay the course. Consistently save money, even if it is a small amount. While it may not seem much at the time, the balance will build up. Trust me, your future self will thank you for every dollar you are able to save today. Two ways you can accomplish this are by prioritizing your spending and
paying down long-term debt. The less you owe, the less interest you will pay. Those interest payments can instead go toward savings!

5. Review your insurance. When diagnosed with a chronic illness, understanding insurance coverage becomes vital to your financial success. Not only do you need to be sure you are selecting the best healthcare plan to reduce out-of-pocket expenses and maximize treatments, you also need to understand disability, life and long-term care insurance.

To gain a better understanding of your needs, it’s a good idea to speak with an impaired-risk specialist who knows which policies would be best suited for you and to help find the right coverage.

“Never make a decision in a vacuum,” says Shenkman. Before purchasing new or additional coverage, thoroughly review your current policies to learn what they cover, how much they can give you and what your best options are. You might assume because you have been diagnosed with a chronic illness that life insurance is not an option or that you are stuck with your current coverage. This isn’t always the case!

To illustrate this point, Shenkman uses his wife’s life insurance. After she was diagnosed in 2006 with multiple sclerosis, he reviewed her policy and found out he would be able to convert it from a term life into a permanent life policy. If you can do this, you can ensure security for your loved ones if something happens to you. In addition, some whole life policies offer chronic care riders that allow the policyholder to receive a portion of the policy to manage expenses if he or she becomes chronically ill, while still having the security of a permanent life policy.

If you already have a whole life policy, make sure you understand it. Many policies have an accelerated death benefit that will allow you to unlock a percentage of the benefit while the policyholder is still living. Whole life policies have a cash value that you can borrow against. Either of these actions, though, will result in a reduced death benefit.

6. Decide whether you need a financial planner. Because of the complicated nature of retirement planning with chronic illness, you may feel your best option is to seek the help of a professional financial planner. If you choose to use a planner, finding the best one doesn’t necessarily mean finding one who specializes in clients with chronic illness. Shenkman advises finding a professional who has integrity and is willing to think outside the box. After you have chosen a planner, it’s up to you to provide details about your illness and what your future needs will be.

7. Plan your estate. Estate planning is one of the foundations of a good financial plan for a couple of reasons. First, it ensures your affairs are in order in case of your death. More importantly, an estate plan can help manage your finances and healthcare if you become incapacitated.

Two important parts of a good plan include a living will and a durable power of attorney. A living will (also known as an advanced directive) is a legal directive that states your wishes in writing about your medical and end-of-life care if you are unable to do so. With a durable power of attorney, you can authorize someone to handle your finances, pay bills and taxes if you become debilitated.

You Can Plan for Retirement Even with a Chronic Illness

While the above-mentioned obstacles may seem insurmountable when you first start planning for retirement, they aren’t! With a bit of financial education and careful preplanning, saving for retirement is possible even with a chronic illness.

ABBIE CORNETT is the patient advocate for IG Living magazine.

References

Retirement Planning Related Websites
- Social Security Benefits Planner: www.ssa.gov/planners/calculators
- Retirement Calculator: www.calculator.net/retirement-calculator.html?cagennov=51&retirementage=68&lifeexpectancy=85&ssn=1600&inflationrate=3&currentincome=80000&retiredincome=75&ctype=1&x=33&y=18
- Nerd Wallet: www.nerdwallet.com/investing/retirement-calculator
Help IG Living Magazine **Go Green**

Join our campaign to reduce unnecessary paper consumption!

**Here's how you can help:** If you can forgo receiving a hard copy of the magazine and utilize the digital version instead, go to [www.IGLiving.com](http://www.IGLiving.com) to select the Go Green tab to sign up for the electronic version and opt out of the print version.

---

**The Benefits of Going Digital for You!**

- Get notified and enjoy **earlier online access** to every new issue
- **Print individual articles** to keep or hand out to friends, family and care providers
- **Easily share articles** instantly on Social Media
- **Read the issues anywhere** at any time on all of your digital devices (smartphone, computer, iPad, tablet)
- Quickly **access all published articles**
While the prescription of long-term corticosteroids is often warranted, there are ways to mitigate and manage potential metabolic, immunologic and other complications.

By Bob Geng, MD

Corticosteroids are produced in the adrenal glands of the human body. They are important hormones that serve significant physiologic purposes in the regulation of the metabolic and immunologic systems. Their natural levels fluctuate throughout the day, and their production and release are highly regulated by the hypothalamus and pituitary glands.

Cortisol is the main corticosteroid naturally produced in humans. Due to important function of cortisol in regulating the metabolic and immunologic systems, many different synthetic corticosteroids have been created that mimic its function. Some of the most commonly used synthetic corticosteroids include prednisone, prednisolone, dexamethasone, betamethasone, hydrocortisone, methylprednisolone and triamcinolone. These can have very potent effects when used at high doses to treat a variety of immunologic and inflammatory disorders, and they have been widely used for a long period of time.

The efficacy of corticosteroids is often quite remarkable, but equally remarkable are the deleterious effects they may have on many organ systems — particularly associated with prolonged use. The purpose of this article is to highlight some of the commonly known and recognized complications of chronic systemic corticosteroid use and discuss ways to mitigate use and manage complications.

Adverse Complications of Chronic Corticosteroid Use

The complications of exogenous corticosteroid use can be grouped into several main categories, including metabolic...
changes, immunologic problems, ocular disease, skin health and behavioral changes.

From a metabolic standpoint, one of the biggest concerns is hyperglycemia secondary to impaired glucose tolerance and insulin resistance. This can lead to diabetes, or worsening of glycemic control in existing diabetic patients. Significant caution needs to be taken when prescribing high doses for diabetic patients. In addition to hyperglycemia, exogenous chronic corticosteroid use can lead to an increase in truncal obesity and body mass index. This is because corticosteroid use leads to appetite stimulation contributing to weight gain. Weight gain can have a further negative impact on glycemic control. Both hyperglycemia and obesity lead to the development of the metabolic syndrome, which will result in a negative impact on the cardiovascular system associated with hypertension, cardiovascular and cerebrovascular complications. In addition, corticosteroids can have a direct negative impact on the cardiovascular system by mimicking the effects of aldosterone to increase fluid retention.

Another aspect of metabolic disruption secondary to long-term corticosteroid use is growth and hormonal dysregulation. Chronic systemic corticosteroid use has been linked to linear growth retardation and menstrual cycle abnormalities. In addition, its use can have detrimental effects on bone health, leading to osteopenia and osteoporosis.

Beyond weight gain, chronic corticosteroid use is associated with an overall distortion of body appearance. A shift in distribution of body fat toward the face leads to the appearance of “moon face” (very rotund appearance of the face) and toward the posterior neck leads to the formation of a “buffalo hump.” Weight also increases in the abdominal region leading to the appearance of truncal obesity, and weight will often shift away from the extremities in both fat and muscle (atrophy of muscle). Overall, these body appearance changes can be termed as “Cushingoid features.” Fortunately, all of these tend to resolve upon discontinuation of corticosteroid use.

Chronic administration of exogenous corticosteroids will lead to adrenal suppression of endogenous production of cortisol. This means if exogenous corticosteroids are withdrawn abruptly or without tapering, patients may develop symptoms of adrenal insufficiency due to an insufficient level of endogenous cortisol.

The immunologic complication of exogenous corticosteroid use is mainly in immunosuppression. This is also the main purpose of therapeutic corticosteroid use since it is often administered to counteract an overactive inflammatory reaction. Indeed, corticosteroids can have a tremendous impact in suppression of both the innate and adaptive immune systems, potentially leading to a secondary immunodeficiency. Initial exogenous corticosteroid use can lead to a seemingly higher peripheral blood leukocyte count (white blood cell count) because it prevents the transit of these immune cells out of the bloodstream. However, long-term use of corticosteroids can lead to a decrease in T cell, B cell and neutrophil number and function due to the significant effects of suppressing the immune system. Chronic corticosteroid use has also been linked to low immunoglobulin counts and, in some instances, the need for immune globulin replacement therapy. The degree of immunosuppression may lead to a state of immunodeficiency that makes patients susceptible to a myriad of opportunistic infections (pathogens that would not normally be able to infect a healthy individual with normal immune function), as well as more severe manifestations of common infections.

Beyond metabolic and immunologic adverse complications, chronic systemic corticosteroid use has been associated with ocular disease such as cataracts and glaucoma. Special caution is needed for patients with those baseline conditions, and consultation with ophthalmology may be warranted. Skin health such as the development of significant thinning of the skin is also adversely affected by chronic corticosteroid use. Lastly, behavioral changes may be observed with corticosteroid use. Patients can demonstrate an increase in irritability and excitability. If corticosteroids are administered at night, they can interfere with sleep. In some instances, they have been associated with the development of “steroid psychosis” with frank hallucinations.

Managing Complications of Chronic Corticosteroid Use

Due to the myriad adverse complications associated with chronic corticosteroid use, all attempts are usually made by physicians to limit their use to the lowest possible dose for the shortest possible duration. During times of severe acute inflammation, it may not be possible to avoid the use of systemic corticosteroids, but the goal is short-term use. For patients who may need long-term immunosuppression due to persistent autoimmunity or inflammation, steroid-sparing agents are often used to mitigate the need for chronic systemic corticosteroid administration. Steroid-sparing agents are often other immunosuppressive medications that act through different pathways and mechanisms to achieve a similar targeted
clinical outcome as corticosteroids. Some steroid-sparing agents have the ability to decrease inflammation by blocking pathways that are not immunosuppressive.

Another method of decreasing the use of chronic systemic corticosteroid use has been targeted localized corticosteroid application. These topical local administrations can take many different forms. For lower-respiratory diseases like asthma and chronic obstructive pulmonary disease, inhaled corticosteroids will deliver the medication directly into the airways, decreasing the amount of systemic absorption. For upper-airway diseases like chronic rhinosinusitis, intranasal steroids can deliver the medication directly into the nasal cavity in a concentrated fashion with minimal systemic absorption. For inflammatory ocular disorders, steroid eye drops can be given (although baseline intraocular pressures should still be checked prior to administration). For chronic inflammatory skin disorders such as atopic dermatitis and psoriasis, topical corticosteroid creams and ointments may be used to target the skin directly with less systemic absorption. For arthritis, corticosteroids can be directly injected into the joint space to concentrate the medication in the affected area while minimizing the systemic impact of adverse effects.

Unfortunately, localized administration of corticosteroids help to reduce long-term complications, but they do not eliminate them. Excessive use of high-potency topical steroids on the skin over a long period of time can still lead to skin thinning and atrophy. Long-term use of inhaled corticosteroids has been linked to linear growth suppression in children. Fortunately, additional steroid-sparing agents have been developed to also limit the use of localized corticosteroid administration.

Overall, close communication with patients’ healthcare providers is key when chronic systemic corticosteroid therapy is used. Potential metabolic complications can be monitored with routine lab work such as a comprehensive metabolic panel (which checks the blood glucose and liver enzymes that may be affected), the hemoglobin A1c (to screen for diabetes and prediabetes) and cholesterol testing. Routine physician follow-up is helpful to track blood pressure and weight since both are adversely affected by systemic corticosteroid therapy. For postmenopausal women, routine evaluation of bone density scans is helpful to screen for development of osteopenia and osteoporosis.

From an immunologic perspective, being vigilant about infections is crucial. If an opportunistic infection such as thrush or yeast infection arises, topical antifungal agents can be prescribed. There should be a low threshold to seek medical attention for any unusual, severe or prolonged infections since chronic corticosteroid use can lead to a degree of being immunocompromised. Routine complete blood counts and other immune surveillance labs such as quantitative immunoglobulin counts and lymphocyte subset flow cytometry may also be helpful to monitor and screen for any potential development of immunodeficiency secondary to chronic corticosteroid use.

Useful and Effective, But Caution Is Warranted

Corticosteroids are very potent and effective medications. However, due to their impact on so many organ systems, their long-term use can have significant detrimental consequences. Therefore, every effort should be made to limit their use to the lowest effective dose for the shortest duration possible. Sometimes, due to lack of alternative treatments and the ready availability of corticosteroids, high-dose administration for a short period of time is necessary to overcome overwhelming inflammation. However, every attempt should be made to decrease and gradually taper the use of systemic corticosteroids to minimize their negative impact. Localized administration, topical administration and use of safer alternative steroid-sparing agents should always be considered for all disease states. If there are no other good options besides the use of chronic systemic corticosteroids, then all attempts should be made to closely monitor patients metabolically and immunologically to ensure early detection of any potential adverse complications.

BOB GENG, MD, MA, studied medicine at Washington University School of Medicine in St. Louis, where he also completed his residency training in internal medicine. He is currently an assistant professor in allergy and immunology at the University of California, San Diego. Dr. Geng received his bachelor’s and Master of Arts degrees in Georgetown University’s School of Foreign Service.
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

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

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

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

(800) 323-6832
www.NuFACTOR.com

©2018 NuFactor, Inc. is the specialty pharmacy subsidiary of FFF Enterprises, Inc., the nation’s most trusted distributor of plasma products, vaccines and other biopharmaceuticals.
ALSO KNOWN AS immunodeficiency with thymoma, Good’s syndrome (GS) is a rare, adult-onset primary immunodeficiency (PI) characterized by low immunoglobulins (hypogammaglobulinemia) and a benign thymic tumor (thymoma). Eosinophils (white blood cells) may also be very low or undetectable in these patients.1

The association between immunodeficiency and thymoma was first recognized by Robert Good, MD, in 1954. It is classified as a distinct entity by the expert committee of the World Health Organization/International Union of Immunological Societies on PIs. Patients with GS may experience immunodeficiency prior to or after the diagnosis of a thymoma. And, while GS was noted in 7 percent of adults with PI attending a chest clinic, it is believed this figure is influenced by referral bias, with the actual incidence in PI patients more likely to be between 1 percent and 2 percent. Conversely, the incidence of hypogammaglobulinemia in patients with thymoma is between 6 percent and 11 percent. GS affects men and women equally, and while it can occur in children, it is extremely rare. Typically, GS is diagnosed in the fourth or fifth decade of life — much later than when most PIs are diagnosed.2

Symptoms of GS

While symptoms vary from person to person, the initial clinical features are varied (Table 1). The primary feature of GS is infection. Patients are susceptible to recurrent infections caused by bacterial, viral and fungal pathogens due to extensive hypogammaglobulinemia and lymphopenia (low levels of white blood cells). Most infections occur in the sinuses and respiratory tract, but infections can also occur on the skin and in the urinary tract. Diarrhea is also common. Sinopulmonary infections manifest with a cough, nasal discharge, fever and headaches, while diarrhea, abdominal pain, cramping and weight loss are typical signs of gastrointestinal infection. Other symptoms can include cytomegalovirus (CMV) retinitis (a sight-threatening disease), mucocutaneous candidiasis (yeast or candida infections), herpes simplex virus and human herpesvirus 8. In rare cases, central nervous system infection can occur.3

Patients with GS may experience immunodeficiency prior to or after the diagnosis of a thymoma. The second main feature of GS is tumor of the thymus gland (thymoma), with up to 42 percent of patients having a confirmed diagnosis prior to the onset of infections. In addition, thymomas are associated with a number of autoimmune disorders. Up to 50 percent of patients present with signs and symptoms of myasthenia gravis. But, other patients present with pemphigus, Sjögren’s syndrome, pure red cell aplasia and systemic lupus erythematosus. In up to 10 percent of patients, hypogammaglobulinemia may be severe even after successful removal of thymomas and is the main cause of mortality.3
Causes of GS
Since its discovery in 1954, little more has been found out about its pathogenesis. However, there are two possible pathogenic mechanisms for the association between antibody deficiency and thymoma. The first explanation is that “cytokines, possibly secreted by bone marrow stromal cells, may influence both thymic and B cell precursor growth and differentiation.” A second explanation “comes from studies of paraneoplastic phenomena in thymoma, such as pure red cell aplasia, which show that T cells or autoantibodies can directly or indirectly inhibit erythropoiesis [production of red blood cells]. T cells isolated from patients with thymoma can inhibit immunoglobulin production by B cells and pre-B cell growth in healthy controls.”

Diagnosing GS
Diagnosing GS can be very difficult due to its various symptoms that can be present at different periods.
GS is usually first suspected when a thymic tumor is detected. Unfortunately, there are cases when a thymoma is not detected, resulting in misdiagnosis. Indeed, GS is often misdiagnosed as common variable immunodeficiency due to lack of awareness and recognition. Because thymomas can be “a subtle feature on chest X-rays, and in one study, 25 percent of tumors were missed with a diagnostic delay of 41 months,” it is recommended to perform a computed tomography (CT) scan of the chest if a thymoma is suspected. A CT scan can define the extent and stage of the thymoma, as well as whether bronchiectasis is present.

In approximately half of the cases of patients with a thymoma, the history of recurrent infections precedes the detection of the thymoma. As such, GS should be suspected in PI patients aged 40 years and older. Patients who present with thymoma should have serum immunoglobulin values and B and T cell subsets measured. Even if these are normal, the measurements should be repeated every second year since cases of progressive immunodeficiency have been found. If immunoglobulin values are found to be low, response to the tetanus, diphtheria, pneumococcus and haemophilus vaccines should be measured. Failure to mount an adequate antibody response to these vaccines indicates an immunodeficiency is present.

Treating GS
Treatment for GS includes resection of the thymoma and immune globulin (IG) therapy to prevent infections. Yet, while removal will not cure the immunodeficiency, it may help other symptoms. If bronchiectasis is present, patients may need postural drainage, prophylactic antibiotics and, in some cases, more intensive IG treatment. Patients with stage 3 or 4 disease thymomas often require radiotherapy and combination chemotherapy.

IG therapy is the only way to prevent infections caused by PI. In one review of the efficacy of IG treatment for GS, 23 of 30 patients had a reduction in the numbers of bacterial sinopulmonary infections. Another study conducted just recently at a large tertiary referral hospital in Thailand investigated the clinical outcomes of GS patients after treatment with intravenous IG (IVIG) therapy from January 2005 through December 2015. Nine GS patients with a median age at diagnosis of 53 years presented with pneumonia and sepsis as the most common clinical manifestations. Six patients also presented with infectious organisms suggestive of cell-mediated immunity defects, including CMV, Mycobacterium tuberculosis, Mycobacterium...
abscessus, herpes simplex virus, pneumocystis jirovecii and Aspergillus. Mean serum IgG level was 317 mg/dL, eight patients had very low to undetectable B cells, and all patients had either low CD4 number or impaired T-cell function, and one had both. All patients received monthly IVIG replacement therapy at a dose of 0.4 g/kg. The mean trough IgG level was 881 mg/dL. After treatment, seven patients had favorable clinical outcomes, but two died due to septicemia.5

Late-onset diagnosis of GS can be problematic. In a case report published this year, a 57-year-old man was admitted to the hospital with a history of thymectomy due to thymoma six years previously. He developed weight loss and recurrent persistent diarrhea caused by isospora belli (an intestinal infection). His chest CT scan revealed bilateral bronchiectasis, and his labs showed hypogammaglobulinemia. After a diagnosis of GS, he was treated with monthly IVIG, but he lost his vision on the left side due to CMV retinitis; he also developed nail candidiasis.6

Treating GS patients who also present with autoimmune disorders can be even more complicated. In one case report in 2017, a 65-year-old woman was admitted to the hospital with ptosis (drooping upper eyelids) and abdominal muscle weakness. Based on the presence of anti-acetylcholine receptor antibodies, she was diagnosed with myasthenia gravis (MG). At the same time, invasive thymoma of Masaoka stage IVa was detected. After being treated with chemotherapy followed by high-dose corticosteroids, the thymoma regressed and the MG went into remission. However, several months later, the woman developed repeated bacterial respiratory tract infections, CMV infections and esophageal and systemic candidiasis. Lab tests revealed a marked decrease in IgG levels and severe reduction in B cells, as well as a decrease in the CD4+CD5+ T cell to CD4+CD5-T cell ratio indicative of deregulation of CD4-T cell activation, suggesting the patient had impaired humoral and cell-mediated immune responses. She was continued on antibiotics and given regular IVIG therapy.7

Clinical Outlook

GS is a rare association of thymoma and adult-onset immunodeficiency that is often difficult to diagnose and frequently presents with autoimmune disorders. Since it was first discovered, understanding of this syndrome has improved considerably. According to the most recent reports, the mean survival rate of patients with GS is 14 years, and overall mortality rates are between 45 percent and 57 percent. Ultimately, for better awareness of this disease and its early diagnosis and treatment, more research is needed to uncover its cause.

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

References


Organizations Supporting GS

Immune Deficiency Foundation
(800) 296-4433
idf@primaryimmune.org
www.primaryimmune.org

Jeffrey Modell Foundation (JMF)
www.info4pi.org

Canadian Immunodeficiencies Patient Organization (CIFO)
(877) 262-2476
info@cipo.ca
cipo.ca

Immune Deficiencies Foundation Australia
(800) 100-198
info@idfa.org.au
www.idfa.org.au

International Patient Organization for Primary Immunodeficiencies (IPOPI)
(44) 01503-250-668
Info@ipop.org
www.ipopi.org
Researchers are taking a closer look at intravenous immune globulin for its potential to stop the progression of multiple complex conditions from lupus to multiple sclerosis.

By Ilana Jacqueline

**THE LIST OF** conditions that intravenous immune globulin (IVIG) may potentially treat has grown exponentially since its first use to treat primary immunodeficiency disease (PI) in 1952. It is known IVIG protects against infections, modulates the immune system and reduces inflammation, but it’s not entirely understood why it works, not only for diseases it is approved to treat, but for many others that have failed to respond to conventional treatments.

To date, the U.S. Food and Drug Administration (FDA) has approved IVIG for only six indications: PI, idiopathic thrombocytopenic purpura, multifocal motor neuropathy, B-cell chronic lymphocytic leukemia, Kawasaki disease and chronic inflammatory demyelinating polyneuropathy. And, while the most frequent prescribers of IVIG therapy have been immunologists, today, specialists in neurology, nephrology, rheumatology, dermatology and hematology have all found clinical uses for the treatment. As a matter of fact, it is believed, and in some instances medical evidence has shown, that IVIG may be beneficial for treating many off-label indications, which, according to past estimates, represent 50 percent to 80 percent of total IVIG use. These indications include a host of complex medical conditions, including lupus, multiple sclerosis (MS), Alzheimer’s disease, dysautonomia, infertility and many others.

**The IVIG Process**

The manufacture of IVIG starts with plasma donations. In the United States alone, there were more than 38 million donations of plasma collected in 2016, according to the Plasma Protein Therapeutics Association (PPTA), more than double the 15 million donations collected just one decade ago. Worldwide, the total annual demand for plasma by pharmaceutical companies that manufacture plasma-based therapies is about 38 million liters. To meet this growing demand, most of the world’s plasma is collected in about 400 plasma donation centers scattered throughout the U.S., with some of it exported to other countries.

Once collected, plasma — 92 percent water and 8 percent proteins — must go through a fractionation process that separates and collects the individual proteins, of which 64 percent are albumin, 20 percent are immune globulin, 2.5 percent are alpha-1 antitrypsin, less than 1 percent are clotting factors, and 13.5 percent are others such as antithrombin, protein C, C1 esterase inhibitor, etc.

As part of the industry’s voluntary international standards program for manufacturers, known as the Quality Standards of Excellence, Assurance and Leadership (QSEAL), all plasma is held in inventory for 60 days before it can enter the manufacturing process. This allows for rigorous testing to identify, retrieve and destruct plasma donation from donors who are disqualified for various reasons such as having received a tattoo or piercing at the time of the original donation or failing to report foreign travel.

Once the plasma is released from inventory, it is ready for fractionation. During the fractionation process, plasma is pooled from multiple
Donations, purified and processed in a specific order to extract specific plasma proteins that have a proven health benefit. The steps and regulations required to collect donated plasma and complete the manufacturing process that ultimately results in the final therapies takes between seven and nine months. Between weeks 0 and 4, the plasma is collected. Then, between weeks 4 and 12, it is batched and transported to the fractionation plant, where it is stored from weeks 12 through 16. During this period, “it is the combination of time, temperature, pH and alcohol concentration [that] allows the extraction of the specific therapeutic proteins.” At that point, the plasma is inspected and released for production. Production occurs between weeks 20 and 24. Then, between weeks 24 and 28, internal testing of the therapeutic proteins takes place, and the therapies are then released by FDA and shipped between weeks 28 and 32 to the wholesalers and end users.

**The IVIG Challenge**

In the U.S., there are currently six companies — BPL, CSL Behring, Grifols, Kedrion, Octapharma and Shire — that manufacture and market IVIG products. These include Carimune NF, Flebogamma 5% DIF, Flebogamma 10% DIF, Gammagard Liquid, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam 5%, Octagam 10% and Privigen.

With a half-life between three to four weeks and costs exceeding $100,000 annually, IVIG for off-label (unapproved by FDA) treatment is frequently a burden for providers to present enough scientific evidence to gain approval for the cost of treatment. For those fortunate enough to be treated with IVIG off-label, results are often not instantaneous, but often take several months and several infusions before any benefits are quantified, which can cause disruption in therapy. This was the case for Cece Collins, a patient with dysautonomia who did not respond to typical treatment options. “My doctor has championed the use of IVIG for me, but the insurance process has still been a nightmare,” says Collins. “We had to have mountains of documentation, including research articles. The medical director at the institution where I’m receiving treatment wants me to submit a review of my progress after every four infusions. But I won’t see results until I have at least six solid months of continuous treatment. Gaps in coverage mean that I’m only on my fourth dose, and I was just rejected from the next round of infusions.”

The process has been frustrating for both Collins and her physician who have had to manage her rapid decline. Unfortunately, the relevant studies have not been enough for insurance to provide continuity of care and to keep her stabilized.

**IVIG Possibilities**

Despite the challenges, many diseases are being studied for off-label IVIG treatment with varying results. Following is a brief review of a select few.

**Lupus.** An autoimmune disease that can cause damage to skin, joints and organs, lupus is a chronic condition thought to be caused by a combination of a person’s hormones, environment and genetics. The Lupus Foundation of America estimates 1.5 million Americans and at least five million people worldwide suffer from a form of lupus. The disease strikes mostly women of childbearing age, and can cause symptoms of hair loss, extreme fatigue, stroke, rashes and chronic pain.

FDA has approved the use of corticosteroids, antimalarial drugs, monoclonal antibody belimumab (Benlysta), Acthar injections and aspirin to treat lupus. However, because of these powerful immune-suppressing treatments, some patients experience lower antibody levels that can leave them vulnerable to infection. Treating these infections is a primary benefit of IVIG. However, IVIG can also help boost abnormally low platelet or low red blood cell counts. And, the use of IVIG can prevent a patient’s white blood cells from destroying platelets, which can cause autoimmune thrombocytopenia and autoimmune hemolytic anemia.

While IVIG is not a first-line treatment for lupus since it is time-consuming and expensive, for some patients, it may be their only hope at successful disease management. A 2015 study at the University of California, Irvine, tested the efficacy of IVIG in lupus erythematosus patients that yielded positive results. In the study, 15 patients were administered 500 mg/kg of IVIG per day on consecutive days up to a total of 2 g/kg per month for three months. IVIG was then discontinued, and the subjects were monitored for an additional six months for a possible relapse. Study results showed IVIG monotherapy achieved rapid and persistent decrease in disease activity, steady improvement of patients’ quality of life, low relapse rate and mild nature and short duration of relapses. In addition, since healing was maintained for months after treatment, the researchers concluded it is possible “IVIG triggered molecular events mediating the therapeutic action of IVIG that continued to unfold after the end of therapy.”

**MS.** An autoimmune disease that impacts the central nervous system, MS often impairs the spinal cord, optic nerves and brain.
This chronic, lifelong condition is progressive, meaning it can intensify over time and, in some patients, may become disabling. There is no cure for MS, but there are treatments available to help patients manage symptoms. These include corticosteroids to help reduce inflammation and disease-modifying drugs such as Betaseron, Axonex, Extavia and Plegidy.

While IVIG does not appear to slow the progression of MS, treatments may lengthen the time between relapses for those with relapsing-remitting MS. Doctors have also prescribed IVIG for patients with severe relapses that have not responded to corticosteroids.

In a 2008 meta-analysis of six clinical trials, researchers found results were consistent. While IVIG was well-tolerated, the studies could not substantiate a beneficial effect of IVIG in the studied doses, and the utility of IVIG for relapsing-remitting MS was still in question. However, the results did prove that IVIG can be considered as an alternative therapeutic option, second-line therapy or adjuvant therapy considering its positive beneficial effects.

Alzheimer’s disease. Though Alzheimer’s disease affects an estimated 5.5 million Americans and is the sixth-leading cause of death in the United States, its cause and cure have eluded scientists since its discovery by German physician Alois Alzheimer in 1906. This fatal progressive disease, which is the most common form of dementia, destroys brain cells and causes challenges with brain function and memory loss.

IVIG therapy for Alzheimer’s was thought to be very promising in its early stages. In 2009, the Alzheimer’s Association reported studies of medical records of 847 people who received IVIG treatments versus those of nearly 85,000 people who did not. The studies showed people who received IVIG had a 42 percent lower risk of developing Alzheimer’s disease over four years.

Discouragingly, in 2013, a more formalized study called GAP (Gammaglobuin Alzheimer’s Partnership) was conducted by the Alzheimer’s Disease Cooperative Study (ADCS), the National Institute of Aging and Baxter International. The Phase III trial measured the progress of 390 patients in 45 centers in the United States and Canada, and after 18 months of treatment, it failed to prove efficacy in reducing cognitive decline and stabilizing existing functional abilities in patients.

However, while most studies found on clinicaltrials.gov have either been completed or terminated, there are currently two active studies not currently recruiting, and another that began in December 2015 and is currently recruiting that is evaluating the effect of IVIG on brain scans for research purposes only (not for medical treatment). The study is broken down into three parts in which patients receive a single dose or placebo, multiple doses or placebo for up to 24 weeks and multiple doses or placebo for up to 72 weeks.

Interestingly, a new study posted on Oct. 24, 2017, purports to determine if changes in brain amyloid levels are evident three months after infusion of 0.4 g/kg of IVIG every 14 days times five infusions. The study is currently enrolling by invitation, and it is estimated to be completed by May 2019.

Dysautonomia. Presenting in several forms, including postural orthostatic tachycardia syndrome, neurocardiogenic syncope and multiple system atrophy, dysautonomia is an umbrella term used to describe the dysfunction of the autonomic nervous system. It is not a rare condition, as it is estimated to affect (in some form) more than 70 million people worldwide, according to Dysautonomia International. The condition can occur secondary to diseases like diabetes, rheumatoid arthritis, celiac disease, Parkinson’s and Sjögren’s syndrome.

Treatments for dysautonomia are varied. There is no cure, but patients may improve with treatment for an underlying cause of the disease. Treatment is often prescribed per symptom and can include responses to combat orthostatic hypotension. Patients are instructed to elevate the head of their bed, eat a high-sodium diet and can be prescribed drugs such as fludrocortisone and midodrine. Doctors are also exploring the use of intravenous saline therapy.

Jill Schofield, MD, a researcher on the topic of antiphospholipid syndrome and the use of IVIG in refractory autoimmune dysautonomia patients, describes these patients as having an underlying autoimmune disease, a family history of autoimmune disease and progressive worsening of dysautonomia symptoms over time, despite typical treatment. She has been prescribing a unique dose of IVIG to these patients: high-dose (1 to 2 gm/kg monthly) given slowly with aggressive hydration to reduce the risk of aseptic meningitis and thrombosis. On average, 88 percent of patients have responded to treatment within 5.7 weeks. “IVIG is highly efficacious in patients with refractory autoimmune dysautonomias,” says Dr. Schofield. “And, it is a fairly safe and well-tolerated treatment in these patients when given with pre- and sometimes post-hydration.” Dr. Schofield plans to release the results of her findings later this year.

Infertility. Pregnancy and autoimmune conditions can lead to a multitude of complications, including miscarriages. It is not understood why IVIG works for women with recurrent pregnancy loss, but it has been found to lower the incidence of
miscarriage. One theory is that IVIG combats natural killer cells (NK). Women who have never given birth and have high levels of NK cells in their peripheral blood have higher chances of pregnancy loss.14

With infertility affecting nearly 12 percent of women in the United States, it can be troubling to many who find themselves excluded from what some physicians feel is an ineffective treatment with a high price tag. The American Society for Reproductive Medicine and the American Congress of Obstetricians and Gynecologists claimed that after reviewing five studies in the 1990s, IVIG simply did not show enough evidence to suggest it could treat or prevent miscarriages. Still, some doctors vouch for its impact, and many fertility clinics still offer the treatment to couples in distress, as long as they’re willing to pay for it out of pocket.15

Alzahra Hospital Tabriz in Iran is currently recruiting for a study on immunomodulatory effects of IVIG on pregnancy rate or patients with recurrent pregnancy loss. A similar study on unexplained primary recurrent miscarriage and the use of IVIG is being conducted in Tokyo.16

Other disease states. There are currently more than 90 clinical trials recruiting patients to study the use of IVIG in different conditions, including:

• Small fiber neuropathy
• Kawasaki disease
• Chronic inflammatory demyelinating polyneuropathy
• Influenza
• Toxic shock syndrome
• Autoimmune epilepsy
• Spinocerebellar ataxia
• Demyelination in diabetes mellitus
• Sickle cell disease
• Post-polio syndrome
• Graft-versus-host disease
• Antibody positive psychosis
• Idiopathic inflammatory myopathy
• Sarcoidosis
• Dermatomyositis
• Myasthenia gravis
• Aloimmune thrombocytopenia

Looking Forward

Numerous researchers believe IVIG holds promise for treating many more diseases than those currently FDA-approved. And, clinical trials are key to uncovering its potential. According to Lilly Stairs, who serves on the board of the American Autoimmune Related Diseases Association and heads patient advocacy at Clara Health, patient participation in clinical trials is what truly makes or breaks awareness, availability and coverage of a new treatment. “Patients are key stakeholders in the clinical trials process and absolutely have the power to both improve and expedite them,” explains Stairs. “When sponsor companies include patients in the clinical trials design process, it exponentially improves enrollment, speed and outcomes because the trial is tailor-made to accommodate the needs of the patient.”

To help in expediting the process, it is critical that physicians make patients aware of clinical trials and the power of breakthrough research. “We need to work together to demystify clinical trials, which are often stigmatized and considered a ‘last resort,’” adds Stairs. “All patients deserve to know that clinical trials are a treatment option and can provide access to cutting-edge therapies. Greater awareness and participation will result in faster enrollment and ultimately a quicker pathway to approval.”

ILANA JACQUELINE is a dysautonomia and primary immune deficiency disease patient from South Florida who has been writing professionally since 2004.

References
Profile: Whitney Ward

By Trudie Mitschang

WEIGHING JUST 4 pounds 9 ounces at birth, Whitney Ward battled an endless array of health problems and endured multiple hospital visits as an infant and child, but doctors had no clue what was making her so sick. Then, following a series of “divine” appointments, Whitney was invited to be the subject of a dissertation at the National Institutes of Health (NIH). That invitation led her to become the first person diagnosed with what is now known as MAGIS syndrome — a name she came up with and submitted to NIH for consideration. Today, the busy college graduate is an aspiring writer who hopes her story will inspire others to live fully in spite of chronic illness.

Trudie: Your symptoms began at birth. Tell us about that.
Whitney: When I was born, I looked like a preemie who was 10 weeks early, even though I was full term. I had clubfeet and dislocated hips, and when I was just a few months old, I battled chronic ear infections and severe asthma. I was prescribed medication that helped, but not always. Sometimes, my asthma attacks got so bad, the only thing my parents knew to do was stick my head in the freezer. Or, if it was winter, they would take me outside to try to get my lungs to relax. As time passed, I also battled viral pneumonia and chronic ear and sinus infections. No one knew what was wrong, and doctors predicted I’d outgrow it.

Trudie: What led to your diagnosis of septic arthritis?
Whitney: I underwent a lot of testing and learned I had a form of autoimmune hemolytic anemia, which basically meant they had absolutely no clue what was wrong. My medical care was sporadic at best but eventually led me to Nationwide Children’s Hospital when I was 11 years old, where I have been a patient for 19 years. I gained a wonderful team of doctors that consists of my hematologist, immunologist and rheumatologist. Eventually, they were able to tell me I had combined immune deficiency complicated by autoimmune hemolytic anemia.

Trudie: What was life like prior to diagnosis?
Whitney: It’s funny, I was actually an athletic, rumble-tumble tomboy. I was quick, sturdy and strong. I have no doubt if I had been completely healthy, I would have become a college athlete. But because of my illness, I felt like I stood on the sidelines watching the lives of kids my age keep going while mine just stopped.

Trudie: Was it tough making friends?
Whitney: Growing up was a little lonely. When your illness is invisible, not everyone understands or believes what you’re going through. I felt all
people could see were my deformities. One thing my disease causes is excessive warts, and one time in junior high, a boy in my class told me I should be in the Guinness Book of World Records for having the most warts. The pain and humiliation I felt in that moment was almost more than I could bear.

Trudie: How did you stay strong?

Whitney: There were three things God gave me that helped me get through the heartache. The first was my family. My parents were always there to support and advocate for me, my sister was my best friend and cheerleader, and my grandparents were always there to lend a listening ear. The second was my church family. The third was my love for reading, which allowed me to escape and forget about all of the uncertainty.

Trudie: When did you become involved with NIH?

Whitney: I was a 22-year-old college sophomore, and my immunologist casually mentioned he sent my case to NIH. He said there was a new disease that had just been discovered called DOCK8, and I had similarities to it. He hoped I would fit the protocol. I was repeatedly cautioned it was unlikely the research would fit the protocol. I was repeatedly discouraged. I came over and presented me with a hug and bouquet of flowers and announced that instant, I received closure I never knew existed. They explained that when it came to naming a disease, the rules had changed. No longer did the disease consist of a person’s name, but the name needed to form an acronym that stood for the prominent symptoms of the disease. In the end, I came up with five names, and my favorite was MAGIS syndrome. It fit the scientific requirements; plus, MAGIS means more in Latin and is related to a Latin phrase: “To the greater glory of God.” To me, this was absolutely perfect because I wanted other patients to know they are more than their disease.

Trudie: When did you learn your name was selected?

Whitney: I attended Ian’s dissertation with my family, and at the end, he came over and presented me with a hug and bouquet of flowers and announced they had selected MAGIS syndrome! In that instant, I received closure I never had before. The years of pain, scary medical procedures, 26 surgeries, missed opportunities and a lonely childhood had not been in vain, but it had been for a greater purpose.

Trudie: What’s your treatment plan today?

Whitney: It’s been a long journey and lots of trial and error, but today, I take rituximab every six months and am treated with subcutaneous immune globulin.

Trudie: How has being involved with the Immune Deficiency Foundation (IDF) impacted your life?

Whitney: I have only been involved with IDF for two years, and the first thing I did was participate in one of their primary immunodeficiency awareness walks. I discovered there were people out there like me who completely understood what I was going through. I gained a community of friends I never knew existed.

Trudie: What is your day-to-day life like now?

Whitney: I am actually the healthiest I have ever been. Everyone who knows me realizes this is a complete and total miracle. In December 2011, I almost died; my hemoglobin levels dropped to 2.8, and from all appearances, the disease I had fought all my life was about to take my life. But miracle after miracle happened, and my health turned around so much quicker than my doctors ever imagined. I have never looked back. I have been on six mission trips, I’m a Sunday school teacher at my church, I’m active at my local gym, I help my grandpa harvest a garden, I volunteer for IDF, I am the founder and president of Peculiar Treasures (an online book club), and I now have many dear and special friends who are my prayer warriors. When I look back at where I was in 2011 to where I’m at now, I can see how God gave me beauty from ashes.

Trudie: What are your goals?

Whitney: I want to be married and have children. I have been asked what I would do if I passed on my gene mutation to my children, and I believe I would raise them the way I was raised and instill in them they are more than their disease. Careerwise, I am working hard to launch my writing career. I have a bachelor’s degree in creative writing with a minor in journalism. And, I write a weekly blog titled “More Than My Mountains,” which can be found at morethanmymountains.blogspot.com.

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.
PATIENT PERSPECTIVE

The New Reality: This Is How It Really Is

By Stacy Oliver

THERE ARE TIMES when changes happen in life subtly: the weather, time of day or growing older. We age from a baby to an adult and look back on pictures of ourselves and marvel. Somehow, the barren winter trees grow spring leaves, the bright sky turns dark, and you’re moving your way through the world first crawling, then walking and finally running.

It seems when I first had the symptoms of multifocal motor neuropathy (MMN), it was subtly abrupt. One day, my hands weren’t quite working the way they used to, but it was quickly evident to me. It was odd and I noticed it, but it was not dramatic enough to cause alarm. Same with the Sjogren’s syndrome symptoms. My dry eyes and mouth were apparent and annoying, but I couldn’t figure it out. Only over time did the symptoms of both diseases become worse enough for me to take action to find out what they meant and get diagnosed.

It’s been 11 years since I’ve been diagnosed with MMN. In the eight years since, I’ve added lupus and Sjogren’s to the list. I’ve had a chance to get “comfortable” and learn to live with my various conditions and their changing states. It’s as if I were a cake with multiple layers of filling, knowing exactly what I taste like, but now there’s a new mystery flavor. I seem to have a layer of filling nobody knows for sure what it is. With things staying the same for so long, coming to terms with these subtle changes has been a rude awakening. This is my new reality. A mirror was held up to my situation, and I had to see it for what it was.

Now that I look back, I really took it all in stride. I slowly had trouble with my balance and gait; I was falling more. For long walks, I’d use a cane. It was only after I literally fell on my face (breaking my sunglasses) that I realized this was serious. Then, I began having cognitive issues, slurring my words and, once, I forgot how to drive. I had to remind myself which was the gas pedal and which was the brake. There are other problems, too, but I don’t need to share my laundry list. Everyone reading this has their own. I think all of us have enough laundry to fill a laundromat.

What’s crucial is these were new symptoms. Having gone so long telling doctors the same story, I had to start paying attention again to my body. The story changed, and there were new questions doctors were asking me. There were new doctors to be seen. So far, there is no answer, and that’s OK. That is an answer. They are trying different approaches. Some days are better than others, but on the whole, 2018 has proved to be a new adventure with my body. There are now tasks that have abruptly changed: Driving is on hold (my sense of perception is wrong), speaking isn’t as fluid (foggy memory doesn’t help), and I am constantly trying to balance myself. Steady as she goes!

I walk full-time with a cane now. A year ago, heck even last October, I didn’t use one when I walked the dogs. I can’t walk my dogs now; they are too much of a fall risk. My head and one of my hands are trembling; I vibrate. One of my doctors put it well: He said I’m elusive. Sounds mysterious and out of a noir book. It’s like my brain yawned, acted weird and then closed up again. There is still so much to learn about the brain. The blood tests I was given didn’t even exist 15 years ago. What will researchers come up with tomorrow?

My advice: Be present and enjoy now, because it can change, and you can be faced with a new reality. A year ago, I thought my somewhat slow gait was an issue. Who knew a year later I’d need a cane for balance and I’d slow down even more. The Greek philosopher Heraclitus of Ephesus said: “Change is the only constant of life.” Bob Dylan said: “The Times They Are a-Changin’.” I am certain those phrases are true, so I’ll see what else the future brings.

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.

Be present and enjoy now, because it can change, and you can be faced with a new reality.
How to Manage Treatment Locally

By Ilana Jacqueline

IT’S BEEN MY observation that chronically ill patients are constantly breaking up with their local team of physicians. Then, finding a new one knowledgeable about their particular disease can be a challenge. Here is my advice for managing treatment with local physicians.

When looking for a doctor to help manage your condition locally, always try to go to someone who specializes in whatever field most closely relates to your disease (neurologists, hematologists, rheumatologists, etc.) and who works in a small practice with a small patient load.

While it’s great for the doctor to have some experience with your disease, if that’s not possible, then the most important qualities to look for is one who:

• Likes to read and stays updated on research in his or her field;
• Finds your case fascinating;
• Has the time to devote to you;
• Has an office staff who gets along with you and who will relay your messages quickly and accurately;
• Has privileges at your local hospital; and
• Is willing to spend between 45 minutes to an hour with you per appointment.

Wow. Let’s talk about that last one. That is a lot of time for a doctor to spend with a patient, right? Most doctors spend 15 quick minutes, and then they’re onto the next case. These are not the doctors for you. The kind you need do, however, exist. Time, follow-through and interest are the key factors to finding one.

Here are some problems you will likely encounter in your search and suggestions for how to solve them:

Problem: The doctor doesn’t think he’s got enough expertise to handle your crazy, complicated case.

Your approach: Talk him off the cliff, and let him know your expectation is not for him to have all the answers, but to have the initiative to explore, question and work with you as a consulting planner.

A generous flood of information usually sets the doctor straight.

Try saying: “I understand this is more complicated than your usual cases, and you feel I would be best served at a university, but I need to have someone local working on my case who can manage treatment. I can help you get a better understanding of my disease. I’m happy to put you in touch with my specialists or send you some articles or a summary of my medical history. What do you need on my end to help me get on track?”

Problem: The doctor doesn’t trust your opinion on treatments you think are worth trying and, instead, wants to stick with what little he knows about treating similar diseases.

Your approach: A generous flood of information usually sets the doctor straight. Print out articles and bring them in, give him brightly covered sticky notes with the names, emails and phone numbers of other doctors he can consult, bring up articles and organizations on your phone, show him a detailed account of past treatments you have tried and failed and, of course, keep a well-documented account of your case history.

Try saying: “I want to understand a little better why you feel this treatment won’t work for me? This study shows patients with my symptoms do well with this treatment. I think it’s worth exploring. I’m going to leave this research with you and this doctor’s phone number. Why don’t you reach out to him, and if you feel like it still isn’t a good move for me, we can look into something else.”

(Honestly, with this approach, the “light” either goes on in the doctor’s head and he wants to learn more, or the doctor loses his fight and is willing to take a second look at the information you’ve presented just to get you to stop bringing in more stuff for him to read! Uh, but you should keep doing that anyway.)

Problem: The doctor doesn’t want to consult with a specialist or your physician, make any calls, write emails or allow you to schedule a time to set up a phone call between the three of you during a scheduled appointment.

Your approach: If you’ve tried reasoning with the doctor about your expectations, and he still doesn’t want to make an effort, leave! Next!

A generous flood of information usually sets the doctor straight.

ILANA JACQUELINE is a 28-year-old dysautonomia and primary immune deficiency disease patient from South Florida. She’s been writing professionally since 2004 on everything from health and wellness to celebrities and beauty. Her blog www.letsfeelbetter.com is both a personal collection of anecdotes about life with chronic illness, as well as a resource for patients of all ages.
ADOLESCENCE PRESENTS many barriers between parents and teens, especially when it comes to communication. As teens strive for independence, they crave their privacy, and they may prefer to keep certain aspects of their lives to themselves. But when that teen has a chronic illness, his or her parents need to keep the communication lines open, particularly regarding matters of health.

As a parent of two teenagers, one of whom has a primary immunodeficiency disease, I know at least one way to cut off communication almost instantly. It’s a trick I’ve mastered over the years: nagging. Because I’m a worrier, I tend to bombard my 15-year-old son with questions about his health status: “How do you feel? Is your cough getting worse? Your voice sounds strained. Is that just puberty or do you have a sore throat?” More often than not, these questions are met with an eye roll. Usually he won’t even answer me. And, I can’t say I blame him. If the only thing I ever talk to my son about is his condition, I will annoy him to the point where he won’t come to me when he really has a problem. I want him to feel free to talk to me about his condition, or anything else he’s dealing with in his life.

So how do I make that happen?

During these difficult teenage years, how can parents keep up healthy levels of communication without coming across as intrusive or interfering?

Add a Dash of Praise

Parents often praise their children when they’re younger and constantly learning new things and mastering new skills. But, adolescents also need the self-esteem boost that praise brings. Even though teenagers might act indifferent when it comes to what their parents think, they actually still want and need their approval. Teens need to know their parents are proud of their accomplishments; they need to feel valued. Parents should actively seek opportunities to be positive and encouraging to their adolescent children. It’s good for the parent-teen relationship, especially if it has become strained.

Spend Time Together

Though verbal conversation might seem like the most efficient way to express feelings and get information, teens may not be too enthusiastic about having long talks with their parents. Thankfully, talking isn’t the only way to communicate. Spending time doing things with a teen might be a more effective way to build trust and open communication without talking about anything personal.

To open the gates of nonverbal communication,

Improving Communication with Your Chronically Ill Teen

By Jessica Leigh Johnson
parents and teens should spend time together doing things they both enjoy such as hiking, going to the movies or craft projects. Though they may prefer to keep their distance at times, teens need to know they can be around their parents and share positive experiences without having to worry they’ll sneak in intrusive questions or lecture them about something.¹

Keep Your Emotions in Check

Not all conversations with teens are calm heart-to-hearts. Sometimes, emotions run hot and voice levels escalate quickly, turning what started off as a conversation into an argument. At times, it may seem arguing is the only form of communication teens are capable of, other than texting. Parents can do their part to keep discussions from flaring up into arguments by controlling their own emotions. It might require some tongue-biting, especially if the teen is being rude, but nothing cuts off the stream of communication faster than a teen who feels attacked by his or her parents. Although parents may be justifiably angry or frustrated, yelling, screaming and verbal put-downs are not going to produce the desired results.² If the teen has lost his or her temper, parents must not respond in kind; they need to remember who the adult is. Adolescents are less able to control their emotions or think logically when they’re upset.³ One good strategy for keeping emotions under control is to count to 10 or take deep breaths before responding. If both the child and parents are too upset to talk, the conversation might have to be paused and revisited once everyone has had a chance to calm down.

Mix Conversation with Action

Gregory L. Jantz, PhD, founder of The Center for Counseling and Health Resources in Edmonds, Wash., suggests moving conversations out of the house and taking them outside — to the basketball court, baseball field, sidewalk around the neighborhood — anywhere the teen can be free to move around while talking, rather than feeling caged-in and cornered.

Forcing a teenager, especially a boy, to sit down and sit still while being berated with a long lecture will lead to nothing but distraction. According to Dr. Jantz, “Boys are generally spatial processors and, therefore, think best when they are active and moving.”² Today’s teens already struggle with a waning attention span. Talking in a more neutral, activity-filled environment will keep them more alert and engaged.

Take Time to Listen

Sometimes, asking direct questions isn’t the best way to get information about what’s going on in a teenager’s life. It may come across as prying. Teens are more likely to be open with their parents if they don’t feel pressured to share information. A casual comment about something that happened during the day might be the teen’s way of reaching out, and parents are likely to hear more if they simply sit back and listen.³

If parents are lucky enough to get their teen talking, they should try not to spout off an answer or give advice the second their child closes his or her mouth. This only tells the teen that rather than listening, the parents were too busy formulating a solution and didn’t really hear what they were saying. When it comes time to respond to what the teen has said, parents should start with empathy, not answers. They can even repeat back what their child has said: “You’re feeling stressed out about that test in Algebra tomorrow.” By repeating their words back to them, parents are showing their teens they hear them and are acknowledging how they feel.

Establish Healthy Communication Early

Teens with chronic illness don’t want to deal with their condition on their own. Even if they don’t say it often, they still need their parents. It’s comforting for them to know they have Mom or Dad around if they need help or advice, or if a medical problem arises. So, if parents take the time to establish healthy communication practices now, their teenager will be more likely to reach out to them when they need something, rather than keeping it to themselves. ²

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

References


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.
Flu Season Safety Tips

By Trudie Mitschang

INFLUENZA (FLU) is a contagious viral disease that typically occurs during the fall and winter months, a period commonly referred to as flu season that often begins in October and November. Most of the time, flu activity peaks between December and February and can last as late as May. Flu symptoms include cough, fever, sore throat, headache, chills, muscle aches and fatigue. While there are several different strains of the flu virus, all are transmitted from person to person via airborne droplets (typically when an infected person coughs or sneezes). For most people, contracting the flu is unpleasant and inconvenient, resulting in time off work and school and avoidance of regular activities. But, for those with a primary immune deficiency (PI), the flu can cause severe and even life-threatening complications, which is why taking precautionary measures to avoid contracting the flu is such a high priority for those in the PI community.

Be Safe, Not Sorry

Common-sense hygiene practices are critical to limiting the spread of the flu virus. Consider more frequent handwashing, the use of antibacterial hand sanitizers and even respiratory masks when visiting highly populated public areas such as shopping malls. Ask your primary care physician about additional ways to prevent exposure based on your specific illness and lifestyle.

Public health officials advise the most effective way to avoid an infection with influenza is to receive an annual flu vaccine. Influenza vaccines are safe, and contrary to a common misconception, they do not cause the flu. Because influenza viruses characteristically change or mutate from year to year, it is necessary to prepare a new vaccine formula for protection against the circulating flu strains of the current year. In a nutshell, that means getting a flu vaccine last year will not provide protection during this year’s flu season.

The Immune Deficiency Foundation advises all family members and other household members in contact with a patient with a PI receive the 2018/2019 influenza vaccine, which provides protection against four distinct influenza virus strains. Currently, there are two types of delivery methods for seasonal flu vaccine available in the U.S.: the inactivated or “killed” flu vaccine (the flu shot) and a live attenuated influenza vaccine (nasal spray). Both are highly effective in preventing the flu in healthy individuals, but the inactivated version is advised for PI patients. Ask your doctor about your specific situation to see if this vaccine is right for you.

You’re Sick, Now What?

If you are one of the unfortunate individuals who comes down with a case of the flu this year, you have several options. It’s important to note that antiviral medicines such as Tamiflu or Relenza are very effective if taken within the first 48 hours of symptom onset. Antiviral medicines have been found to shorten the duration of influenza and lessen the severity of symptoms.

“We want all our primary immunodeficiency patients to take influenza seriously,” said Dr. Ann Bullinger, PharmD, who leads the U.S. medical affairs team for CSL Behring’s immunoglobulin therapy area. “Check with your doctor about preventive measures, and be sure to get immediate attention if you show any signs of the flu.”

Once flu symptoms are in full swing, there is not much you can do but ride it out. In addition to getting lots of rest and drinking plenty of liquids, a number of products can help you feel a little less miserable. Look for over-the-counter treatments such as:

- Oscillococcinum, a homeopathic supplement in Europe that has gained popularity in the U.S. Studies suggest it may shorten the duration of the flu and ease symptoms, but there’s no proof that it prevents the flu.
- Elderberry extract has been shown to ease symptoms when taken within the first 24 to 48 hours after you start to feel ill. There aren’t any known side effects if you use it for five days or less; just don’t eat the plant itself as it can cause nausea.
- Over-the-counter cough and cold medicines like NyQuil and DayQuil are proven effective in relieving symptoms in adults. For high fever and body aches, both products come in an extra-strength “severe” formula. Side effects include drowsiness.

Flu season is without question everyone’s least favorite time of the year. Since the flu returns annually and is considered a common virus, it’s easy to not take it seriously. But, remember, when it comes to the PI patient population, the flu is nothing to sneeze at. Talk to your doctor about the best prevention options during the 2018/2019 flu season, and take necessary precautions now to keep you and your loved ones influenza-free.

Reference


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

Sambucol is a black elderberry extract that claims to provide natural immune system support. Developed by a world-renowned virologist, Sambucol is comprised of the same black elderberry extract that has been used in scientific studies. One bottle contains 30 chewable, berry-flavored tablets and is safe for daily use.  
$10.75; [iherb.com](https://www.iherb.com)

**Soothing Solution**

Vicks VapoRub can temporarily relieve bronchial irritation, as well as ease aches and pains in muscles and joints. The product contains active cough suppressants that are clinically proven to relieve coughs quickly. It is used by applying topically to the chest or neck. $4.89; [riteaid.com](https://www.riteaid.com)

**Daytime Relief**

DayQuil is an over-the-counter medication formulated to treat daytime symptoms of the cold and flu, including cough, headache, minor aches and pains, fever and sore throat, without causing drowsiness.  
$9.99; [vicks.com](https://www.vicks.com)

**Symptom Relief Minus Side Effects**

If taken at the first sign of flu-like symptoms, Oscillococcinum claims to reduce the duration and severity of body aches, headache, fever, chills and fatigue. The sweet-tasting pellets dissolve under the tongue and do not cause drowsiness. They are recommended for everyone ages 2 years and older.  
$24.99; [puritanspride.com](https://www.puritanspride.com)

**Concentrated Caplets**

Theraflu ExpressMax Severe Cold & Flu caplets combine four active ingredients to deliver “7-symptom relief.” In addition, a specially formulated coating provides a unique warming sensation. The product can help reduce aches, pains and congestion, while also reducing fever.  
$6.49; [target.com](https://www.target.com)
**BOOK CORNER**

**New and Useful Reading**

**Immune Deficiency Foundation Guide to Immunoglobulin Replacement Therapy for People Living with Primary Immunodeficiency Diseases**

Authors: Kristin Epland, FNP-C, and Elena E. Perez, MD, PhD
Publisher: Immune Deficiency Foundation

This guide was developed for patients and caregivers to help increase understanding of immune globulin replacement therapy. An increased understanding puts patients and caregivers in a better position to make informed decisions regarding care. Education about the therapy can also build confidence and create a level of understanding that can reduce a patient’s anxiety about a treatment. If patients and caregivers are well educated about the treatment, they will understand the goals they need to work toward to improve health and be more motivated to reach those goals, which will in turn improve patient overall health.

**I Am A Chronic Illness Crusader: An Adult Coloring Book for Encouragement, Strength and Positive Vibes: 20 Powerful Pages To Color (Courageous Coloring) (Volume 2)**

Author: Kathy Weller
Publisher: CreateSpace Independent Publishing Platform

This book offers support, empathy and creative therapy for those experiencing the pain and discomfort associated with chronic illnesses. It features single-sided coloring pages with motivational, encouraging and empowering sayings and affirmations specifically for those facing the daily challenges of chronic illness. Coloring is a highly creative and meditative activity that can have powerful therapeutic anti-stress and relaxation benefits. It activates the brain’s right hemisphere, reducing stress and promoting a relaxed, meditative state by focusing the brain on the “here and now.”

**Chronic Widespread Pain: Pill-Free Approaches to Move People From Hurt to Hope**

Authors: Martha and Donald Teater
Publisher: PESI Publishing & Media

This is a comprehensive and up-to-date book written about fibromyalgia (FM) and very common related but poorly understood illnesses. The author is an internationally recognized authority on FM who believes the condition is the key model to better understand and treat related chronic central pain conditions, including headaches, irritable bowel syndrome, chronic bladder and pelvic pain syndromes and oral-facial pain. Common sleep and mood disturbances shared by each of these pain conditions are reviewed. An in-depth discussion of the overlapping clinical and patho-physiologic features of these disorders provide a framework for treatment models relevant to each illness. Throughout the book, Dr. Goldenberg draws on his nearly four decades of professional experience, which includes extensive research in the field and evaluating 25,000 FM patient office visits over the past 30 years. In the last section of the book, he outlines a new paradigm for better understanding and managing these disorders.

**Treating Chronic Pain: Pill-Free Approaches to Move People From Hurt to Hope**

Authors: Martha and Donald Teater
Publisher: PESI Publishing & Media

Written by a mental health professional and a physician with more than 50 years combined experience, this skills manual will teach readers how to treat pain without pills and with confidence, using cutting-edge assessments, insights and interventions. Included are proven computer-based training applications to improve quality of life, mindfulness tools designed to reduce pain, nonopioid medical options and alternative therapies, a session-by-session treatment plan for individuals and groups, unique worksheets/guides/exercises, and case examples.
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 heartwarming 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
<table>
<thead>
<tr>
<th>Disease</th>
<th>WEBSITES</th>
<th>ONLINE PEER SUPPORT</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Ataxia Telangiectasia (A-T)</strong></td>
<td>• A-T Children’s Project: <a href="http://www.atcp.org">www.atcp.org</a></td>
<td></td>
</tr>
<tr>
<td><strong>Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)</strong></td>
<td>• GBS/CIDP Foundation International: <a href="http://www.gbs-cidp.org">www.gbs-cidp.org</a></td>
<td>• The Foundation for Peripheral Neuropathy: <a href="http://www.foundationforpn.com">www.foundationforpn.com</a></td>
</tr>
<tr>
<td><strong>Evans Syndrome</strong></td>
<td>• Evans Syndrome Research and Support Group: <a href="http://www.evanssyndrome.org">www.evanssyndrome.org</a></td>
<td></td>
</tr>
<tr>
<td><strong>Guillain-Barré Syndrome (GBS)</strong></td>
<td>• GBS/CIDP Foundation International: <a href="http://www.gbs-cidp.org">www.gbs-cidp.org</a></td>
<td></td>
</tr>
<tr>
<td><strong>Idiopathic Thrombocytopenic Purpura (ITP)</strong></td>
<td>• ITP Support Association – UK: <a href="http://www.itsupport.org.uk">www.itsupport.org.uk</a></td>
<td></td>
</tr>
<tr>
<td><strong>Kawasaki Disease</strong></td>
<td>• American Heart Association: <a href="http://www.heart.org/HEARTORG/Conditions/More/">www.heart.org/HEARTORG/Conditions/More/</a> CardiovascularConditionsofChildhood/Kawasaki-Disease_UCM_308777_Article.jsp#.T1T2boEPWE0</td>
<td></td>
</tr>
<tr>
<td><strong>Mitochondrial Disease</strong></td>
<td>• United Mitochondrial Disease Foundation: <a href="http://www.umdf.org">www.umdf.org</a></td>
<td>• Mitocure (personal account): <a href="http://www.livingwithspss.com">www.livingwithspss.com</a></td>
</tr>
<tr>
<td><strong>Multifocal Motor Neuropathy (MMN)</strong></td>
<td>• The Foundation for Peripheral Neuropathy: <a href="http://www.foundationforpn.com">www.foundationforpn.com</a></td>
<td></td>
</tr>
<tr>
<td><strong>Multiple Sclerosis (MS)</strong></td>
<td>• All About Multiple Sclerosis: <a href="http://www.mult-sclerosis.org/index.html">www.mult-sclerosis.org/index.html</a></td>
<td>• Genetic Alliance: <a href="http://www.geneticalliance.org">www.geneticalliance.org</a></td>
</tr>
<tr>
<td><strong>Peripheral Neuropathy (PN)</strong></td>
<td>• Neuropathy Action Foundation: <a href="http://www.neuropathytaction.org">www.neuropathytaction.org</a></td>
<td>• CCX (personal account): <a href="http://www.livingwithms.net">www.livingwithms.net</a></td>
</tr>
<tr>
<td><strong>Primary Immune Deficiency Disease (PI)</strong></td>
<td>• Immune Deficiency Foundation: <a href="http://www.primaryimmune.org">www.primaryimmune.org</a></td>
<td></td>
</tr>
<tr>
<td><strong>Stiff Person Syndrome (SPS)</strong></td>
<td>• Scleroderma Research Foundation: <a href="http://www.srfcure.org">www.srfcure.org</a></td>
<td></td>
</tr>
<tr>
<td><strong>Scleroderma</strong></td>
<td>• IDP Discussion Forum: idfopatient.org/forum</td>
<td></td>
</tr>
<tr>
<td><strong>Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)</strong></td>
<td>• PANDAS/PANS Network: <a href="http://www.pans.net">www.pans.net</a></td>
<td>• International Scleroderma Network: <a href="http://www.sclero.org/support/forums/a-to-z.html">www.sclero.org/support/forums/a-to-z.html</a></td>
</tr>
<tr>
<td><strong>Myasthenia Gravis (MG)</strong></td>
<td>• Myasthenia Gravis Foundation of America (MGA): <a href="http://www.myasthenia.org">www.myasthenia.org</a></td>
<td></td>
</tr>
<tr>
<td><strong>Myositis</strong></td>
<td>• The Myositis Association: <a href="http://www.myositis.org">www.myositis.org</a></td>
<td>• Scleroderma Support Forum: curezone.com/forums/s.php?f=404</td>
</tr>
<tr>
<td><strong>Scleroderma</strong></td>
<td>• Scleroderma Research Foundation: <a href="http://www.srfcure.org">www.srfcure.org</a></td>
<td></td>
</tr>
<tr>
<td><strong>Stiff Person Syndrome (SPS)</strong></td>
<td>• Living with Stiff Person Syndrome: <a href="http://www.livingwithspss.com">www.livingwithspss.com</a></td>
<td></td>
</tr>
</tbody>
</table>
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.ff ENTERPRISES.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@ff ENTERPRISES.COM
Want priority access on FLU VACCINES for the 2019-2020 season?


- 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

MyFluVaccine.com
(800) 843-7477 | MyFluVaccine.com

Brought to you by FFF Enterprises, Inc., the nation's largest and most trusted distributor of flu vaccines and critical-care biopharmaceuticals.