October-November 2017

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Managing Changes in Senior Care

Understanding Mannose-Binding Lectin Deficiency
Medical Intervention for Depression

Transitioning IG Coverage to Medicare
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The Challenges of Aging with PI

SINCE THE FIRST discovery of a primary immunodeficiency (PI) disease (agammaglobulinemia) in 1952, the number of individuals diagnosed with PIs continues to expand rapidly. Today, there are more than 200 different forms of PI affecting approximately 500,000 people in the U.S. And, as the prevalence of PI grows, so does the number of discoveries about them, including the molecular causes. Currently, scientists have identified 320 molecularly defined PIs, with 22 genes known to be the cause. And, it is anticipated that as genetic research continues, some 500-plus genes will be identified.\(^1\)

What does all this mean? First and foremost, as the causes of PIs are discovered, there will be more targeted therapies developed to keep PI patients alive for far longer than previously possible. And, while this is extremely good news, it also brings with it some challenges. One that many immunologists are dealing with more frequently is how to transition senior PI patients into nursing homes equipped to deal with the special care they require. On the ground floor of this issue is immunologist Roger H. Kobayashi, who has co-authored with IG Living’s patient advocate, Abbie Cornett, the article “Moving Aging IG Patients into Nursing Homes.” As they explain, because nursing homes cater to seniors, they are equipped to deal with the common chronic conditions this population typically faces. But, PI differs by far from most of those illnesses. Not only do susceptible PI patients have to be gravely concerned about infectious diseases frequently circulating in nursing homes, but they have the added worry about how their immune globulin (IG) treatment may be affected — particularly if they are treated at home. Dr. Kobayashi’s and Abbie’s advice concerning some very specific items to consider when choosing a nursing home offers help to this growing PI demographic.

An equally challenging issue for aging patients treated with IG is insurance. At age 65, patients must enroll in Medicare, even if they have employer-sponsored insurance coverage. As our article “Transitioning IG Coverage to Medicare” explains, this transition “can be more complex than IG therapy!” As an update of an article topic that is covered every two years in IG Living, it reflects a substantial number of changes to Medicare reimbursement. Individuals must be very careful to choose the Medicare plan that will provide the best reimbursement, which will depend primarily on their diagnosis (PI or another), since coverage differs for each. In addition, we report on legislative issues that have complicated the reimbursement process for at-home treatment.

The challenges PI patients face day in and day out are hard enough, without the added burden of mental stress that can lead to depression. Indeed, as our article “Chronic Illness and Depression” highlights, approximately “one-third of individuals with a serious medical condition experience symptoms of depression.” Considering this, it is hoped that patients will benefit from this article that explains the warning signs of depression, as well as outlines various treatment options.

As always, I hope you gain insight from the information presented and enjoy this edition of IG Living.

Ronale Tucker Rhodes, MS

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ABBIE’S CORNER

IDF Says Goodbye to a Legendary Leader

By Abbie Cornett

I’VE ALWAYS ENJOYED attending the biannual Immune Deficiency Foundation (IDF) conference. It is always held at an exciting location, and it provides patients and their families amazing opportunities to learn about their diseases and to interact with others facing the same issues. This year’s conference was held in Anaheim, Calif., and included an evening at Disneyland. And, while it may have been the best conference I have had the privilege of attending, it was also bittersweet: Marcia Boyle, the founder and driving force behind IDF for more than 37 years, announced this would be her last conference because she is retiring. “Most of my professional life has been dedicated to IDF, thus my decision to retire was not an easy one, but I feel the time is right for me and my family,” she said.¹

Under Marcia’s leadership, IDF has helped thousands of patients. Marcia started IDF at her kitchen table after her son John was diagnosed with a primary immune deficiency disease (PI). She was concerned about the lack of information and support available to patients and their families. And, while she had few resources other than her dedication to the patient community, she grew IDF into an internationally recognized patient organization. Under Marcia’s leadership, IDF worked with immunologists and other specialists to create the first educational materials for patients and their families. Today, these materials are the leading sources of information for patients and families worldwide. But, Marcia didn’t limit her outreach to education; she has also led the way in patient advocacy. I first met Marcia at an immune deficiency advocacy day in Washington, D.C., where I was incredibly impressed with her and the IDF staff. Not only did Marcia seem to know everyone on Capitol Hill, but the day was perfectly organized, with meetings scheduled with everyone’s representative.

Through her efforts, IDF has achieved a number of legislative measures that have improved patients’ quality of life and access to care:

• The Medicare IVIG Access Act (HR 1845). Signed into law in January 2013, this act initiated a demonstration project to evaluate the benefits of providing payment and service items needed for in-home administration of intravenous immune globulin (IVIG) for the treatment of PI. Prior to its enactment, Medicare allowed patients with PI to receive home infusions under Medicare Part B, but did not cover the associated costs (nursing and supplies), thus rendering the benefit useless. Under the demonstration project, there is a per-visit payment for items and services needed for in-home administration of IVIG based on the national per-visit low-utilization payment amount under the prospective payment system for home health services.

• Newborn screening for severe combined immune deficiency (SCID). SCID is now part of the recommended federal uniform core-screening panel. Currently, 44 states are screening newborns for SCID, and five more will have pilot programs in place this year. Thus, as a result of advocating for adherence at the state level, 90 percent of all infants born in the United States will receive screening for this life-threatening condition.

• Champion of Change in Precision Medicine. In recognition of her leadership in the development of electronic personal health records and patient-powered, patient-focused research programs at IDF, Marcia, along with eight other healthcare leaders, was honored in July 2015 by the White House as a Champion of Change in Precision Medicine for initiating and supporting a program that was emblematic of then-President Obama’s Precision Medicine Initiative.

Under Marcia’s leadership, IDF has helped thousands of patients. Her years of service to the community will never be forgotten. Everyone at IG Living extends heartfelt thanks to Marcia. Enjoy your retirement. Your leadership and support will be missed! ²

Reference

ABBIE CORNETT is the patient advocate for IG Living magazine. She can be reached at patient advocate@igliving.com or (800) 843-7477 x1366.
Join the conversation! Connect with other immune globulin patients through IG Living’s Facebook page at www.facebook.com/IGLivingMagazine. See our daily posts of interesting articles and facts, as well as thought-provoking questions that you can chime in on. Following are some snapshots of what’s being discussed.

Was your diagnosis delayed by your doctor?

Yes, I was sick all my life. I went to doctor after doctor. Finally, when I was 25, my gastrointestinal doctor did lab work and found out I had CVID.

— Paula B

I was 36. I had several pneumonia infections as a toddler, ear infections all my life, congenital chronic obstructive pulmonary disease, chronic asthma and chronic bronchitis infections.

— Gina Marie G

It took a while with complaints from me asking why I was always sick to get a diagnosis at 43, but I’m so glad my allergist listened and took me seriously. I’ve been on intravenous immune globulin for seven-plus years and have never felt better!

— Ronda M

Is stress making you sicker?

Yes! Due to the medications I take for myasthenia gravis, my adrenal glands do not function (adrenal insufficiency). They are protectors of any kind of stress — good or bad. I have been extremely sick and have been in the intensive care unit due to this. Basically, it’s impossible to be completely stress-free. However, I am learning that I can stay away from situations that do not directly impact me and will make me ill.

— Judy S

My early years were stress-riddled. Sickness causes stress, and stress makes you sick; it can be a vicious cycle. Even with coping skills, stress is still affecting our bodies.

— Vicki DH

Do you worry about pandemics?

Early on, I panicked about everything. Then, I realized I wanted to live as normal a life as possible. I am still aware and cautious, but I do not let my disease define me or control me. On paper, I am sure my answer looks good, but there have been instances when I have really freaked out. Fortunately, my doctor has always encouraged me to talk to him about concerns and questions, so I have information and answers. Then, I am equipped to rationally think things through and come up with solutions for what might be ahead. I know I am very lucky to have all I need to access issues and then determine what my concerns should be and what actions I should be taking to minimize the concerns I have.

— Jenny G

No. If I get infected by something, I just hope it’s quick. I’m going to die sooner or later and heaven awaits, so I can’t waste my time worrying about something that might happen. Today’s troubles are sufficient for today. Dealing with a bad upper-respiratory infection that turned into bronchitis for the last two weeks while looking for a job has definitely had my attention enough to not worry about some pandemic that may or may not happen.

— Dana O
Leslie » SCIG is not yet FDA approved to treat CIDP, but some large-scale trials are underway. One trial of CSL Behring’s Hizentra has recently been completed, but study results aren’t posted yet. Based on those results, CSL Behring should soon be submitting a biologics license application to FDA for the indication; however, I don’t believe it will be granted until 2018.

It should be noted that some insurance companies are paying for SCIG to treat CIDP based on current peer-reviewed literature showing its efficacy. So, if the route of administration is initially denied, it can and has been successfully appealed.

Question » Is SCIG Approved by the FDA to Treat CIDP?

Leslie » Based on United States Pharmacopeia 797 guidelines, once IVIG is accessed, infusion should begin in one hour and should be administered in a continuous fashion until the infusion is complete. If the infusion is stopped for a short period due to side effects, and resumed shortly after (30 minutes to one hour), the infusion can continue. If the IVIG is completely stopped and doesn’t resume in a short period of time, it should be discarded — not placed in the refrigerator for administration at a later time.

Each product may have different language in its package insert. For example, Gamunex-C’s package insert states: “The GAMUNEX-C vial is for single use only. GAMUNEX-C contains no preservative. Use any vial that has been entered promptly. Discard partially used vials. Do not store after entry into bottle.”

Most manufacturers can provide extended stability (not sterility) data for product that has been transferred from the vial into a bag if that transfer has occurred using aseptic technique in a USP 797-compliant compounding environment; however, this is for storage prior to administration. So, once the bag has been accessed and hung, the same rules as above apply.

Dr. Riedl » Currently, several medications can be useful to relieve migraines post-infusion. The triptan drugs used for typical migraines are often helpful, but other antimigraine medications may also work well. Other treatment options include nonsteroidal anti-inflammatory drugs, acetaminophen, cyproheptadine and steroids such as prednisone. If headaches recur often, it’s worthwhile to try preventive measures such as predosing these medications prior to the infusion, slowing the infusion rate, ensuring aggressive hydration before and after the infusion and possibly switching IG products.

Question » How Can Post-Infusion Migraines Be Avoided?

Leslie » I frequently experience post-infusion migraines. How can I avoid this side effect?

Have a question? Email us at editor@IGLiving.com. Your information will remain confidential unless permission is given.
DiGeorge Syndrome: Further Discussion of Neurologic/Behavioral Issues

By Terry O. Harville, MD, PhD

IN PREVIOUS issues, we discussed features of DiGeorge syndrome (DGS) and partial DGS (PDGS) resulting from the consequences of improper timing of the sequence of events during early phases of embryonic development. We began with a discussion of the genes affected by chromosome 22q11.2 hemizygosity, and how some of these genes may be involved with the neurologic features of this complicated disease. In this column, we will continue discussion of the neurologic and behavioral issues.

In addition to the catechol-O-methyl transferase (COMT) gene previously discussed, the CRKL (CRK like proto-oncogene, adaptor protein) gene has been implicated in developmental problems occurring in DGS/PDGS. The COMT gene affects neurotransmitters in the nervous system, whereas the CRKL gene affects the migration of cells in the nervous system and the ways neurons grow and develop; it also affects the development of the genitourinary system (the organ system of the reproductive organs and the urinary system) and the heart. These alterations in the manner in which the “brain is wired” are thought to be largely responsible for the neurologic and behavioral features of DGS/PDGS.

As we also previously discussed, it is thought as many as a quarter of those with DGS/PDGS may develop more significant psychiatric disorders (primarily schizophrenia and bipolar disorder). And, unfortunately, 90 percent may exhibit some form of neurocognitive issues. The psychiatric disorders become more prevalent with aging, with some reporting an approximate 3 percent incidence in children, but as high as an approximate 25 percent to 30 percent incidence in adults. There also appears to be some correlation with poorer or lower IQ, resulting in an increased risk for schizophrenia illness (or similar) to occur. Since hallucinations are a major part of these psychiatric disorders, it is recommended that physicians managing patients with DGS/PDGS discuss these possibilities during routine evaluations. For example, patients should be asked: “Do you hear voices from someone who you cannot see or who does not seem to be there?” “Do the voices ever tell you to do things such as hurt someone or hurt yourself?” Additionally, it is always recommended that patients with DGS/PDGS have some form of a formal psychiatric evaluation to better judge the potential extent of neurocognitive disease. It is believed that earlier detection and beginning of treatment results in better outcomes.

Behavioral issues are also commonly present. Early on, behavioral issues were thought to be a manifestation of “acting-out” due to learning problems. It is now recognized that behavioral issues are manifestations of the alterations in the brain wiring and function, much like overactivity/hyperactivity, impulsiveness and emotional lability may be commonly found in isolation or in combination. Especially for children, behavioral issues, which can create problems in social situations and in school, are often mistakenly considered a consequence of lack of discipline when interpreted by those observing the behaviors. For example, an observer may make a comment such as: “A good spanking is what the misbehaving child needs.” But this is because the observer does not understand these actions are not learned behavior requiring disciplinary actions. Rather, they are the result of a physical and functional alteration of the brain likely requiring specific medications, as well as specific behavioral modification therapy. Therefore, once again, psychiatric intervention, counseling and therapy are likely needed to provide the most appropriate therapeutic intervention.

We will continue the discussion of the genetics of DGS/PDGS in the next issue.

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

By Elissa Ritt, DHSc

HUMAN PLASMA-DERIVED immune globulin (IG) has been used as antibody replacement therapy for patients with primary immunodeficiency diseases (PI) for decades. More recently, IG has become a first-line immunomodulatory treatment for patients with immune-mediated conditions such as chronic inflammatory demyelinating polyneuropathy. While generally considered a safe, yet invasive, treatment, IG is associated with adverse events of widely varying degrees.

While IG products are all composed of the same therapeutic plasma-derived immunoglobulin G (IgG), they are far from interchangeable. Product formulation, route of administration and, to a lesser degree, manufacturing processes can all affect tolerability and, ultimately, patient outcomes.

Current Brand Landscape

Healthcare providers have a number of brands from which to choose, each with its own product characteristics and adverse event profile. Currently, there are 13 intravenous IG (IVIG) products, five subcutaneous IG (SCIG) products and one facilitated SCIG (fSCIG) product.

Adverse Events

IVIG. Typical mild to moderate systemic adverse events often expected with IVIG treatment include headache, flu-like symptoms (fever, chills and nausea), back pain and skin rash. If these adverse events do appear, they often resolve with medication, a reduction in the rate of infusion or a change in brand. Medication for treatment and prevention can include diphenhydramine and/or acetaminophen.

Serious systemic adverse events, while rare, can also occur. These include aseptic meningitis, cardiovascular events, thromboembolic events, renal failure and anaphylaxis. If a patient experiences any of these, medical care must be immediately sought (Figure 1).

SCIG and fSCIG. The benefits of SCIG and fSCIG often include the avoidance of systemic adverse events. While the mild to moderate and severe adverse events experienced with intravenous administration can occur, they are rare with these products. Instead, side effects are usually infusion site reactions, including redness, swelling and discomfort at the site of the injection. While common, site reactions are usually transient and dissipate over time.

Manufacturing Processes

Because all IG preparations are sourced from human plasma, virus removal and inactivation processes must be employed. These procedures include treatment with solvent and detergent, enzymes, fatty acids or pasteurization, which can alter the fragile IgG molecule, resulting in biologically inactive fragments. While it is important to note the tolerability of existing IG products has not been compared in a head-to-head manner, experts suggest the resulting protein denaturation (loss of native structure) may affect tolerability.

Formulation

Lyophilized versus liquid. Though few lyophilized, or freeze-dried, IG products remain on the market, they can be prevalent in inpatient settings. These products must be reconstituted with a specified diluent prior to use, which can result in denaturation that may cause tolerability concerns for some patients. Fortunately, the vast majority of IG products on the market today are liquid preparations that do not require dilution.
Volume. The volume of IG products is determined by how much of the IgG protein is present in the solution. For example, a product containing only 5% IgG will require twice the fluid volume of a product that contains 10% IgG. Healthcare providers must exercise caution when prescribing higher-volume IVIG brands to fluid-sensitive patients. If a patient is a newborn, elderly, fluid restricted or has renal disease, a lower volume IVIG product should be chosen.

Sugar content. All IG products require a stabilizing agent. Historically, these agents were often a sugar (sucrose, glucose, maltose or sorbitol), but manufacturers shifted to amino acid stabilization after a disproportionate number of patients who received sucrose-stabilized IVIG experienced renal failure.7 Today, the majority of IG products are stabilized with amino acids, and those still stabilized with sugars are those that appear to be better tolerated by the kidneys.

Osmolarity/osmolality. These terms refer to the amount (volume or weight) of solute (all solutes; not just IgG) in a given amount of fluid. Osmolarity is the number of osmoles of solute in a kilogram amount of fluid, while osmolality is the number of osmoles of solute in a liter of solution. The human body has a physiologic osmolality of 258, and products that approach that number more closely mimic the osmolality of human blood.

Products with high osmolarity/osmolality can cause the blood to be thicker or more viscous than normal. Hyperviscosity has been linked to thromboembolic events, myocardial infarction and kidney failure.6,8 Because of this risk, healthcare providers should seek out products with near physiologic osmolarity for elderly or newborn patients, patients with impaired renal function and patients with cardiovascular or thromboembolic risks.

IgA content. Immunoglobulin A (IgA) is another antibody present in most humans. An abnormal deficiency of IgA is relatively common among PI patients. While a very rare, an IgA-deficient patient’s body produces an antibody against the trace amount of IgA found in all IG products. This can be very serious, and can result in anaphylaxis. For these few patients, a low-IgA IVIG product is available.

Route of Administration

IVIG. IVIG is infused intravenously into a patient’s vein over the course of several hours by a medical professional. While this form of administration allows for a higher-volume infusion and requires less-frequent dosing than SCIG, many patients experience infusion-related adverse events described previously.

SCIG. With SCIG, the product is administered into and absorbed through the subcutaneous space. As previously noted, the systemic adverse events often associated with IVIG infusion are rare, though infusion-site reactions are common.

SCIG infusion may be preferable for patients who have intolerable adverse events with IVIG infusion, have poor venous access, prefer the convenience of shorter (but more frequent) and more flexible infusions, or prefer the independence of self-infusion in any location. Additionally, SCIG may be an option for patients who have previously had a reaction to IVIG due to anti-IgA antibodies.10

fSCIG. fSCIG has become a more recent option for patients. The IG preparation is administered in tandem with a recombinant human enzyme to facilitate the absorption of the IgG. Because of this enhanced absorption, fSCIG may be infused on a three-week to four-week schedule as opposed to the one-week to two-week schedule with traditional SCIG.

This boost in absorption may also make the infusion of larger doses more viable.

The safety of fSCIG is very similar to SCIG with a few exceptions: 1) Because of the larger volume infused, fluid may pool at the infusion site and cause discomfort; 2) Transient antibodies to the recombinant enzyme have been noted, though their clinical significance is unknown; and 3) There is some concern with male fertility, though the clinical significance is also unknown.

Clinical Outcomes and Product Selection

Currently, healthcare providers can choose from a vast array of IG products to offer their patients more options than ever. Formulation, route of administration and, to a lesser degree, the manufacturing processes of an IG product can all affect a patient’s clinical outcome. To optimize this outcome, healthcare providers should allow patient comorbidities, tolerability, lifestyle and preference to dictate product selection.

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Research

Studies Show Improved Treatment Satisfaction with SCIG vs. IVIG

Two Phase II/III prospective clinical studies, one in North America and one in Europe, report improved treatment satisfaction with a new human 20% subcutaneous immune globulin (SCIG) versus prior intravenous immune globulin (IVIG) treatment. The studies compared the treatments using the Life Quality Index instrument after patients completed IVIG and again after SCIG 20% treatment. In the North American study, patients received three months of IVIG prior to receiving 12 months of SCIG 20% treatment, and reported improvements in LQI of 33.5 versus 36.5. And, patients in the European study received 12 months of treatment with SCIG 20% following a three-month treatment period with either IVIG or SCIG 20%, and reported LQI improvement of 34.5 versus 39.0. In addition, patients “reported improvement relative to the IVIG therapy period after treatment with SCIG 20% in the therapy setting LQI domain, improving from 17.5 to 20.0 in the North American study and from 18.0 to 21.0 in the European study,” according to lead study author Lisa Meckley, PhD, of Shire Pharmaceuticals, and coauthors.


Resources

CGD Connections Launched to Support the CGD Community


Having CGD can be scary and feel isolating,” says Randall G., who lives with CGD and participated in one of Horizon’s workgroups. “CGD Connections provides a place to connect with those who know what it’s like to have or care for someone who has CGD, and includes information to help on the typical days, as well as how to find strength to tackle the hard ones. Collaborating with other CGD families in the development of this initiative raised shared challenges and led to the creation of resources that we hope will inspire and provide support for those at every stage of their CGD journey.”
Research

Chronic Illness Patients Have High Insurance Out-of-Pocket Costs

Researchers from VA Ann Arbor Health Care System, University of Michigan (U-M) Medical School and Penn State University have found that individuals with chronic health conditions who choose health insurance plans with a deductible spend hundreds or even thousands of dollars of their own money on their care, beyond what they spend to buy the insurance plan. The study analyzed data from 17,177 Americans under age 65 who were interviewed for the Medical Expenditure Panel Survey that showed just over 4,100 had a high-deductible health plan, and 44.5 percent had a chronic health condition, which made it more likely that health-related costs took up more than 10 percent of a chronically ill person’s total income. It also showed huge variations in the amount of out-of-pocket spending between patients who have the same condition, even for low-deductible plans. Yet, despite the out-of-pocket expenses, the study found few people with chronic illnesses said that costs or insurance coverage issues had gotten in the way of getting the care or prescriptions they needed.

Findings are based on data from 2011 through 2013, when many more employers started offering high-deductible plans and before individuals were able to buy their insurance on the HealthCare.gov marketplace. Since the launch of the marketplace, more than 90 percent of people shopping there have chosen high-deductible plans. “Increasingly, these plans have become woven into the fabric of health insurance in America, so it’s important to look at the impact of deductibles on people who need care on an ongoing basis,” said senior author Jeffrey Kullgren, MD, MS, MPH, a research scientist in the VA Ann Arbor Healthcare System and an assistant professor of general medicine at the U-M Medical School. “Not only on how they spend their money on care for their day in, day out health needs, but how that affects spending in the rest of their lives.”

Did You Know?

CSL Plasma has teamed with Wirecard North America to launch the first cash-back donor prepaid card, providing a one-time milestone reward and cash back for spending at local grocery stories and quick-service restaurants that is paid directly to the card. The cash-back option was introduced to support the platinum level loyalty tier of CSL Plasma’s new iGive Rewards Loyalty Program. “Our collaboration with Wirecard has allowed us to offer a new platinum level to iGive Rewards that offers more exclusivity and benefits to our valued donors,” said Robert Mitchell, CSL Plasma’s director of marketing and corporate communication. “With the personalized cash-back rewards prepaid card, platinum level donors receive additional points and other perks…. The evolution of the iGive program and platinum rewards is another way that CSL Plasma is delivering on its promise to give back to the donors who give their time to make sure plasma is available to make lifesaving therapy for patients in need.”
Shire has published three special editions of the quarterly magazine *Just Like Me* for 2017: Living with PI, Living with PI (for kids) and Understanding Insurance. Debuted in 2012, *Just Like Me* is a resource for children diagnosed with primary immunodeficiency disease (PI) and their families to inform them about serious topics related to their disease state in a fun and friendly way. The quarterly magazine is broken up into kids and teen sections, and also includes an insert for adults. Patients can begin receiving the quarterly and special editions by enrolling in Shire’s MyIGSource program at www.myigsource.com/enroll-primary-immunodeficiency-support-program.

**Resources**

**Three Just Like Me Special Editions Published for Children with PI**

John G. Boyle, son of the Immune Deficiency Foundation’s (IDF) president and founder, Marcia Boyle, who previously announced her plan to retire, will serve as the foundation’s new president and chief executive officer. John Boyle brings a wide base of nonprofit management experience to the position, having held a number of executive positions with leading national nonprofit organizations throughout his career and currently serving as the vice president of external relations at IDF. He was instrumental in developing a number of new initiatives for IDF, including the IDF Walk for Primary Immunodeficiency (PI) that launched in 2013 and has grown to 12 cities across the U.S., raising in excess of $1 million to support IDF programs and resources.

“It is an honor to build upon Marcia’s legacy of leadership as IDF continues to work to meet the current needs of the PI community, and to prepare for the community’s future needs,” said Boyle. “It is both an exciting and a challenging time for the people we serve: There are new advancements in treatment options on the horizon, but also growing uncertainty in terms of healthcare policy and insurance limitations. We have an amazing team at IDF, and I am thrilled to continue to work alongside them each day to reach more members of the PI community and to further advance our advocacy, education and research initiatives.”

**Industry**

**John G. Boyle Named President and Chief Executive Officer of IDF**

**Medicines**

**Shire Launches Pediatric Indication for HyQvia in Europe**

Following the recent marketing authorization by the European Commission, Shire (previously Baxter) has launched a pediatric indication for HyQvia (human normal immune globulin 10%) in Europe to treat primary and certain secondary immunodeficiencies. Shire commercially introduced the new indication across the member states of Europe beginning with Germany, Netherlands, Ireland, Greece, Slovakia, Denmark, Sweden and Norway. “We are pleased to bring pediatric patients a new therapeutic option as we build on our broad immunoglobulin portfolio for patients with immune deficiencies,” said Ueli Frankhauser, head, global product strategy. “We intend to expand the availability of HyQvia to more patients in additional geographies, with the goal of reducing the treatment burden for patients worldwide.”

Research

Study Shows the Home-Based SCIG Therapy Setting Provides Higher Satisfaction Among Pediatric Patients

A study conducted in France that assessed quality of life and satisfaction regarding the route and place of administration of immune globulin (IG) replacement therapy treatment found that the home-based subcutaneous IG (SCIG) therapy setting provided higher satisfaction than the hospital-based intravenous IG (IVIG) setting among children. In the prospective, noninterventional cohort study, 44 children aged 5 years to 15 years who had been treated with IG for primary immunodeficiency disease (PI) for three or more months were followed over 12 months. Eighteen of the children were receiving hospital-based IVIG, two were receiving home-based IVIG and 24 were treated with home-based SCIG.

Quality of life was assessed with the child health questionnaire parent form (CHQ-PF), and satisfaction with treatment was measured with a life quality index (LQI) with three components: treatment interference, therapy-related problems and therapy settings. No difference was found on the CHQ-PF assessment. And, there was no difference between the LQI treatment interference and therapy-related problems components. However, the LQI therapy settings component was higher for home-based SCIG than for hospital-based IVIG. And, the LQI therapy settings component significantly improved in five patients who switched from IVIG to SCIG during follow-up when compared to patients who pursued the same regimen (either IVIG or SCIG).

Autoimmune Corner

Medicines

FDA Approves Renflexis, Biosimilar to Remicade

The U.S. Food and Drug Administration has approved Renflexis (infliximab-abda, Samsung Bioepis), the second biosimilar to Remicade (infliximab, Janssen Biotech). Inflectra (infliximab-dyyb, Celltrion) was the first approved biosimilar. The tumor necrosis factor blocker is an intravenous infusion (100 mg) indicated for the same indications as Remicade: Crohn’s disease, ulcerative colitis, rheumatoid arthritis (in combination with methotrexate), ankylosing spondylitis, psoriatic arthritis and plaque psoriasis. The most common adverse reactions that occurred in fewer than 10 percent of patients in clinical trials are infections (e.g., upper respiratory infection, sinusitis and pharyngitis), infusion-related reactions, headache and abdominal pain, which are similar to those seen with Remicade. In addition, Renflexis also comes with the same boxed warning as Remicade concerning the increased risk of serious infections.


Guidelines

AHA Updates Kawasaki Disease Diagnosis and Management Guidelines

The American Heart Association (AHA) has made its first update to the diagnosis and management guidelines of Kawasaki disease issued in 2004. The revised guidelines integrate new findings on the epidemiology, genetics, pathogenesis and long-term outcomes of the disease. Specifically, the revisions fall into four categories:

Pathogenesis. The new guidelines propose there are three linked pathological processes: 1) necrotizing arteritis, which “destroys the arterial wall into the adventitia, causing aneurysms,” 2) subacute/chronic vasculitis, “characterized by an asynchronous infiltration of lymphocytes, plasma cells and eosinophils with fewer macrophages that begins in the first two weeks after fever onset but can continue for months to years in a small subset of patients and is closely linked to the third process” and 3) myofibroblastic proliferation, which involves a unique medial smooth muscle cell-derived myofibroblastic process that can cause progressive arterial stenosis.

Diagnosis. Because a prompt diagnosis is critical, the revised guidelines include an updated algorithm outlining supplemental information that may facilitate the diagnostic process in cases lacking complete classic clinical criteria.

Treatment. In addition to administering intravenous immune globulin (IVIG) as soon as possible within 10 days of fever onset, the revised guidelines recommend IVIG for children who present after the 10th day with “ongoing systemic inflammation as manifested by elevation of ESR [erythrocyte sedimentation rate] or CRP [C-reactive protein] (CRP >3.0 mg/dL) together with either persistent fever without other explanation or coronary artery aneurysms (luminal dimension Z score >2.5).” The revision also includes recommendations for additional therapies, including corticosteroids, infliximab and cyclosporine in patients who are IVIG-resistant, as well as detailed recommendations for management based on coronary artery involvement.

Risk classification. A new classification of coronary artery abnormalities is based on Z scores as follows, with certain caveats:

- No involvement: always <2
- Dilation only: 2 to <2.5, a decrease in Z score during follow-up ≥1
- Small aneurysm: ≥2.5 to <5
- Medium aneurysm: ≥5 to <10, and absolute dimension <8 mm
- Large or giant aneurysm: ≥10, or absolute dimension ≥8 mm

The authors note that important gaps in evidence remain, and “until the cause and pathogenesis are defined, an exact diagnostic test remains elusive, and acute treatment remains somewhat empirical.”

The full guideline can be downloaded at circ.ahajournals.org/content/early/2017/03/29/CIR.0000000000000484.
The U.S. Food and Drug Administration has granted Octapharma USA orphan drug designation for Octagam (immune globulin intravenous [human] 10% liquid) for treatment of dermatomyositis. Orphan drug designation is given to drugs and biologics to treat rare diseases/disorders that affect fewer than 200,000 people in the U.S., or that affect 200,000 or more people, but whose sales are not expected to recover the costs of research and development of the product. Dermatomyositis is a rare acquired disorder characterized by chronic inflammatory and degenerative changes of the muscles and skin. Its main symptom is a skin rash accompanied or followed by progressive muscle weakness. The disease, which has an estimated incidence of 9.63 cases per million, is most common in children between ages 5 years and 15 years and adults in their late 40s and early 60s, but can occur at any age.  

Medicines  
FDA Grants Orphan Designation for Octagam 10% to Treat Dermatomyositis  

The U.S. Food and Drug Administration has granted Octapharma USA orphan drug designation for Octagam (immune globulin intravenous [human] 10% liquid) for treatment of dermatomyositis. Orphan drug designation is given to drugs and biologics to treat rare diseases/disorders that affect fewer than 200,000 people in the U.S., or that affect 200,000 or more people, but whose sales are not expected to recover the costs of research and development of the product. Dermatomyositis is a rare acquired disorder characterized by chronic inflammatory and degenerative changes of the muscles and skin. Its main symptom is a skin rash accompanied or followed by progressive muscle weakness. The disease, which has an estimated incidence of 9.63 cases per million, is most common in children between ages 5 years and 15 years and adults in their late 40s and early 60s, but can occur at any age. 

Many things need to be considered before choosing a nursing home, especially for PI patients who face considerably more challenges than the typical senior.

By Abbie Cornett and Roger H. Kobayashi, MD

UNTIL RECENT YEARS, individuals with a primary immune deficiency disease (PI) rarely survived to retirement age. Indeed, in the past, PI was thought to be a disease that occurred primarily in infants and young children, which led to many adults going undiagnosed and untreated. Today, however, because of advancements in early diagnosis, proper treatment and continued monitoring, PI patients are living much longer. In recognition of this growing PI patient population, the Consortium of Independent Immunology Clinics surveyed three of its member clinics, and found surprising results: The three clinics had almost 300 PI patients over age 65!
Because so many PI patients are living longer, they are reaching the stage of life when residential nursing care may be necessary. And, this is where things start to get complicated. Regardless of whether a person has a chronic condition, the decision to enter a nursing home can be challenging and stressful for patients and their families. It is a decision that needs careful consideration to ensure patients are well cared for and have a good quality of life. And for PI patients, these concerns are magnified.

While change is scary at any age, it can be particularly stressful for older PI patients. One of the biggest concerns expressed by seniors is a loss of control over their lives. For PI patients, the transition to a nursing home adds another element of concern: It can affect where and how they receive their lifesaving medication. For the transition to be successful, patients should include their loved ones in the decision process. A good way to ease the stress of the transition is to start a conversation with family members well before the move is necessary. That way, the family can be included in decisions regarding their care.

Any decision that affects the care of patients should also involve their physician, the best resource in helping assess patient needs and how those needs may evolve over time. Fortunately, most patients have established relationships with their doctors and can ask for their input.

Are Nursing Homes Qualified to Care for PI Patients?

Nursing homes are equipped to deal with the common chronic conditions related to aging. In fact, 80 percent of adults age 65 and older have at least one chronic condition, and many have more than one comorbidity. But, most older adults suffer from chronic conditions such as Alzheimer’s, arthritis, hypertension, etc. So, while nursing homes are used to dealing with these typical illnesses, they may not be as prepared for senior PI patients who have more complex medical needs.

Before selecting a facility, patients must make sure it is certified. For a nursing home to meet certification standards, it must comply with more than 180 regulatory standards, including proper management of medications, measures to protect patients from physical and mental abuse, and safe food handling. There are a couple of good options for checking facility ratings. The first is at Nursing Home Compare (www.medicare.gov/nursinghomecompare/search.html), which allows families to compare information on Medicaid- and Medicare-participating facilities. If a facility is not Medicare-certified, the second option is to go to the patient’s state website to access ratings (www.medicare.gov/NursingHomeCompare/Resources/State-Websites.html).

Things to Consider When Visiting a Nursing Home

Before deciding on a nursing home, patients and their families should visit a candidate facility at least twice: one planned visit and one drop-in visit. Dropping in at an unusual time or on a weekend is a good way to detect any hidden problems such as poor staffing or uncleanness.

Following are items PI patients and their families should specifically consider when choosing a facility:

- Staff: It’s important for patients and their family members to have confidence in the staff’s ability to meet the patient’s needs. When visiting a facility, it’s a good idea to talk to the staff and ask them how many hours per week they work on average. Many facilities require overtime and double shifts, and overworked staff can affect care.
- Cleanliness: As people age, their immune systems diminish. This decrease in natural immunity is compounded in senior PI patients who are very susceptible to infections regardless of age. Further, what might be an uncomplicated infection for others can have a severe and potentially life-threatening effect on geriatric PI patients, making cleanliness of the facility even more important. There should be a concerted effort on the part of nursing home staff to minimize the spread of infection through diligent hand-washing and avoidance of people with ongoing infections. Particular concern needs to be given to viral infections that can cause life-threatening illness in PI patients. Ask if there is a policy in place for the treating physician to be proactively contacted if patients are exposed to infection.

BEFORE SELECTING A FACILITY, PATIENTS MUST MAKE SURE IT IS CERTIFIED.

- Dining facility: When visiting the facility, plan on eating a meal there. Good food is an important quality-of-life issue, but it’s also vital for proper nutrition. While healthy dietary habits are important for everyone, they are especially key for PI patients. A lack of adequate nutrition can lead to many illnesses, including infections for which PI patients are already at risk.

Because all PI patients have been on antibiotics, which can affect their gastrointestinal tract, causing chronic diarrhea, bloating and malabsorption, they may have special nutritional needs. Therefore, patients should speak with the facility dietician to
make sure special dietary requirements can be accommodated. And, since each patient is different, coordination between the patient’s doctors and the facility is necessary.

- Transportation: All nursing home patients need transportation, especially for doctor appointments. This is particularly true for PI patients who, because of their complex medical needs, may require more frequent doctor visits. Fortunately, most facilities are generally equipped to deal with transporting patients, but it’s a good idea to verify that service is available.

- Training: Because PI patients can get very sick very quickly, staff at the facility must be trained to recognize when patients are ill. This is particularly important if patients have lost their ability to communicate.

**There should be a concerted effort on the part of nursing home staff to minimize the spread of infection through diligent hand-washing and avoidance of people with ongoing infections.**

**Treatment Location**

Virtually all older PI patients require immune globulin (IG) infusions. Typically, these infusion services are not currently available in nursing homes. Therefore, proactive measures need to be taken to plan for how and where patients will receive treatment. Here are some items to consider:

- If treatment is administered intravenously (IV) at a doctor’s office, infusion center or hospital, will the nursing home provide transportation to and from the site of care?

- If patients have been receiving IVIG at home, will the nursing home allow outside medical personnel into its facility to infuse patients? If it does not make these allowances, does it have staff properly trained to safely infuse patients?

- If patients have been receiving subcutaneous IG at home, will they be able to administer their own medication while at the facility? The Centers for Medicare and Medicaid Services guidelines require the facility to have procedures for the control and safe storage of medications for residents who self-administer them. Further, residents may self-administer drugs if an interdisciplinary team has determined this practice is safe.\(^1\)

**Treatment Costs**

Before transitioning into a nursing home, it is important to understand the costs. Nursing homes are expensive! The average daily charge for a shared room in 2016 was $225.\(^4\) These costs, combined with the expensive treatment for PI, can be devastating. IG treatment itself can vary between $40,000 and $80,000 a year for the medication and supplies. If patients are hospitalized or suffer complications, those costs multiple quickly. Compounding this problem is how complicated reimbursement issues have become. Before making any decision, it is imperative patients and families understand insurance coverage to avoid substantial unexpected expenses.

**An Ongoing Battle**

The decision to transition to a nursing home and the selection of a facility is only the beginning of the battle. Even after the transition is complete, family follow-up should be part of the plan. Family members must remain vigilant to ensure their loved ones continue to receive appropriate care. Visiting as often as possible ensures their well-being is monitored. Some red flags that signal things are not going well are sudden weight loss, bedsores, unexplained injuries, confusion and sudden changes in behavior.

Senior PI patients have spent a lifetime advocating for their care, fighting with insurance companies and caring for themselves. Their family members need to be prepared to be advocates for aging loved ones who can no longer advocate for themselves.

**Abbie Cornett** is the patient advocate for IG Living magazine, and **Roger H. Kobayashi**, is an allergist/immunologist in Omaha, Neb., a clinical professor at the University of California, Los Angeles, School of Medicine and a national consultant for the Immune Deficiency Foundation, and is on the executive committee for the Consortium of Independent Immunology Clinics.

**References**

NuFACTOR has the distinction of carrying all U.S.-approved immune globulin products. We are committed to exceptional customer service, product and patient safety, and secure product availability and affordability. Excellence is our standard, and we’ve earned the most respected name in homecare. Our customers know we care about them, and that makes all the difference.

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Options in Medicare coverage can be more complicated than IG therapy, but these guidelines can help to ensure a smooth transition.

By Michelle Greer, RN, and Leslie Vaughan, RPh

**IMMUNE GLOBULIN (IG)** is a complex therapy, both clinically and financially, that is used to treat rare and difficult-to-diagnose diseases. For some, IG is a lifetime therapy. And, while at one time this therapy was typically approved and reimbursed without question, today there are extensive medical policies in place that require a diagnosis to be proved and the medical need for IG justified.

Compared with all other insurance plans, Medicare probably varies most in its coverage policies for IG therapy. Therefore, patients who continue to receive IG therapy when they turn 65 or otherwise become eligible for Medicare need to know how to successfully transition to Medicare. In fact, changes in site of care and route of administration may be necessary to ensure therapy is continued without disruption and financial strain.

**Applying for Medicare**

To be eligible for Medicare coverage, patients must be age 65 or older and eligible for retirement benefits under Social Security, or a federal, state or local government employee. To be eligible for Social Security, individuals must have 40-plus quarters of Social Security-covered employment; receive benefits under a spouse’s work record and be currently married; or have received benefits under a former spouse to whom they were married for at least 10 years.

Individuals also may be eligible for Medicare if they are receiving disability benefits under Social Security Disability Insurance; have received railroad retirement benefits for 24 or more months; have end-stage renal disease; or have amyotrophic lateral sclerosis, also known as Lou Gehrig’s disease.
Some individuals will be automatically enrolled in Medicare when they turn 65, whereas others will need to apply. Those who are already receiving Social Security benefits and have enough work quarters will automatically be enrolled for Medicare Parts A and B when they turn 65 or on the 25th month of disability. All others will need to apply for Medicare. An individual who needs to apply for Medicare has a seven-month initial enrollment period to sign up for Part A and/or Part B. This initial enrollment period begins three months prior to the individual’s 65th birthday month, includes the birthday month and concludes three months after the birthday month. Starting the application process as early as possible can minimize any problems getting enrolled.

One of the most important things to consider when turning 65 is if insurance through an employer will continue. If patients or their spouses are still working and the employer has 20 or more employees, Medicare becomes the secondary insurance until they retire. If patients or their spouses plan to retire, and their employer’s insurance will continue, Medicare will become the primary insurance and will cover all approved charges at 80 percent, with the employer’s insurance generally covering the remaining 20 percent of approved charges. If the employer’s insurance will terminate, patients may consider obtaining a Medicare supplemental plan, since 20 percent of the cost of monthly IG therapy can be financially taxing.

For more detailed information, Medicare has a free booklet titled Medicare and Other Health Benefits: Your Guide to Who Pays First that explains all of the options. Another excellent free resource for learning about Medicare is a booklet titled Medicare and You. These booklets, as well as more comprehensive information on basic Medicare coverage, including eligibility, coverage criteria and plan options, can be found on the Medicare website at www.Medicare.gov.

Choosing Medicare Benefits

The original Medicare plans include Medicare Parts A and B. There also is Medicare Part D (the Medicare prescription drug plan) for which patients can sign up. An alternative option to Parts A and B is Medicare Part C (the Medicare Advantage Plan), which is similar to an HMO and usually includes prescription drug coverage.

Coverage for IG varies based on patients’ diagnosis, where they currently receive therapy and whether they receive therapy via the intravenous (IVIG) or subcutaneous (SCIG) route.

Drug coverage for an immune deficiency diagnosis. IG therapy for an immune deficiency is 80 percent covered under Medicare Part B. This is the case whether patients receive IVIG or SCIG. However, any coverage changes should be confirmed for the site of therapy, including the hospital, physician office or home. There is broader coverage in the hospital and physician office than there is in the home. In the homecare setting, coverage is limited to 14 specific diagnosis codes:

- D80.0: Hereditary hypogammaglobulinemia
- D80.5: Immunodeficiency with increased immunoglobulin M [IgM]
- D83.0: Common variable immunodeficiency with predominant abnormalities of B cell numbers and function
- D83.2: Common variable immunodeficiency with autoantibodies to B or T cells
- D83.8: Other common variable immunodeficiencies
- D83.9: Common variable immunodeficiency, unspecified
- D83.0: Wiskott-Aldrich syndrome
- D81.0: Severe combined immunodeficiency with reticular dysgenesis
- D81.1: Severe combined immunodeficiency with low T and B cell numbers
- D81.2: Severe combined immunodeficiency with low or normal B cell numbers
- D81.6: Major histocompatibility complex I deficiency
- D81.7: Histocompatibility complex II deficiency
- D81.89: Other combined immunodeficiencies
- D81.9: Combined immunity deficiency, unspecified

Compared with all other insurance plans, Medicare probably varies most in its coverage policies for IG therapy.

Patients with an immune deficiency that is not identified by one of these 14 diagnosis codes may be covered under the Part D benefit, which is explained below.

Unfortunately, IG is not reimbursed very well under Medicare Part B. Prior to the passage of the 21st Century Cures Act in December 2016, reimbursement for SCIG received at home was adequate to cover the cost of immune globulin. Now, for most providers, Medicare reimbursement is below their cost to purchase IG.
Patients who receive IVIG in a physician office may be asked to change their site of care to a hospital outpatient setting if continuing to receive IVIG, or to change to a home setting to begin receiving SCIG or IVIG. Medicare publishes a new fee schedule every quarter that may impact the brand of IG patients receive. Based on these quarterly reimbursement rates for IG, providers may ask patients to change brands and/or routes of administration if reimbursement for their current product dips below the cost to acquire it.

There are six SCIG products: Gammagard Liquid (Shire), Gamunex-C (Grifols), Gammaked (Kedrion Biopharma), Hizentra (CSL Behring), HYQVIA (Shire) and Cuvitru (Shire). HYQVIA differs from the others because it is a combination product using IG and hyaluronidase. The hyaluronidase component makes it possible for patients to infuse monthly rather than the more frequent dosing that may be required when using traditional SCIG products. Medicare originally did not allow coverage for HYQVIA in the home setting under the Part B benefit; however, that decision has been partially reversed. The manufacturer of HYQVIA, Shire, recommends a dose ramp-up, which means patients start with a partial dose and increase the dose with each subsequent treatment until they reach a maintenance dose. Currently, coverage under Medicare Part B will not pay for the ramp-up phase in the home. Payment for the ramp-up phase is available only in the hospital outpatient and physician office settings. Once the patient is stabilized with the maintenance dose, Part B will cover ongoing doses in the home setting.

While Medicare beneficiaries in this position have options, they are more limited since the passage of the 21st Century Cures Act, and patients are advised to discuss their options with their physician and current provider of therapy well before transitioning to Medicare to develop a plan for continuation of care.

**Drug coverage for other diagnoses.** IG therapy for many other diagnoses is usually covered under Medicare Part B in the hospital outpatient setting or in a physician office. For those currently receiving IVIG in these sites of care, the same rules apply for transitioning to Medicare as they do for patients diagnosed with an immune deficiency.

For those receiving IVIG at home, the rules become more complicated. If patients keep their employer’s insurance, it’s possible no changes will be necessary. Medicare will be billed as the primary insurance; however, reimbursement will be denied as a noncovered benefit with a specific denial code, and then the secondary insurance will be billed. All deductibles and co-payments apply as they did when the employer’s insurance was in the primary payment position. This includes government insurance such as Tricare and Champus. However, one item that may not be covered is nursing services. To bill the secondary insurance, the provider must receive the correct denial code from Medicare. Since there is limited coverage for nursing services under Medicare Part A for homebound patients, home infusion providers are not able to bill nursing services and receive a denial to bill the secondary. Therefore, prior to making the change, patients should discuss how nursing services will continue to be covered with the current provider.

If patients who receive IVIG at home do not keep their employer’s insurance, one option that will allow them to continue IG therapy is to purchase Medicare Part D insurance, a government program for prescription drugs administered by commercial entities. Medicare Part D consists of many plans, so it can be complicated to choose one. All medications that are prescribed, including IG, should be considered when selecting a plan.

Patients can choose a standard benefit program that may have a lower premium but may not offer assistance through the different phases of coverage. Or, they can choose a plan that may have a slightly higher monthly premium but may have better assistance through coverage phases. The four coverage phases for a standard plan in 2018 are:

1) **Deductible:** This is paid 100 percent by patients up to a total of $405.

2) **Initial coverage limit:** For the standard benefit, patients pay 25 percent, and the plan pays 75 percent up to a total out-of-pocket cost of $3,750.

3) **Coverage gap:** In this phase, also known as the doughnut hole, patients are responsible for most of the charges; however, the drug manufacturer may provide payment assistance in the form of discounts. For brand-name drugs, the discount is 50 percent, and for generics, the discount is 56 percent. The out-of-pocket threshold (TrOOP) is $5,000 in 2018. TrOOP is defined by Medicare as “true out-of-pocket. TrOOP costs are the expenses that count toward a person’s Medicare drug plan out-of-pocket threshold. TrOOP costs determine when a person’s catastrophic coverage portion of their Medicare Part D prescription drug plan will begin.”

4) **Catastrophic phase:** Once patients (with the assistance of drug manufacturer discounts) have spent a total of $5,000, they become responsible for a smaller portion of the ongoing cost of the drugs, usually 5 percent of the total cost.

Again, there are options. Patients may qualify for Extra Help, a Medicare program to help people with limited income and resources to pay Medicare prescription drug plan costs. When applying for Medicare, it is important for patients to find out if they might qualify for this program. If they don’t qualify when first obtaining Medicare, patients should periodically recheck as their finances change to see if they qualify. In addition, some homecare...
providers may offer financial assistance programs. If patients are eligible, their financial responsibility can be reduced or waived. And, last, patient advocacy groups also may offer some assistance.

Guidance on selecting the right Medicare Part D coverage can be found at www.medicare.gov, or Medicare assistance can be obtained by calling (800) MEDICARE (633-2273).

The last option for patients who receive IVIG at home is to transition to a hospital outpatient setting where IVIG will be covered at 80 percent under Medicare Part B with the supplemental insurance plan covering the remaining 20 percent. Coverage criteria in the hospital outpatient and physician office setting are based on National or Local Coverage Determinations (NCD or LCD) published by Medicare. The NCD/LCD defines which diagnoses are approved for treatment with IG.

If patients choose to enroll in a Medicare HMO (Medicare Part C or Medicare Advantage Plan), they will automatically be enrolled in a Medicare Part D prescription plan in most cases, and the same rules apply as previously stated. It’s important for patients to understand this before choosing a Medicare HMO so they can make the best choice and have the least interruption in therapy.

If patients also have Medicaid, known as being dual eligible, they typically have the most options. Medicare is the primary insurance, Medicaid is the secondary insurance, and they will automatically be enrolled in Medicare Part D. Co-pays for dual-eligible patients are very low, usually in the $3 to $4 range. And, coverage may be 100 percent for infusions in the hospital or at home. However, if patients are infused in a physician office, they should check on their options.

Nursing and supply coverage for all diagnoses. In the physician office and hospital outpatient setting, nursing and supplies are covered under Medicare Part B. In the home, nursing for both IVIG and SCIG is covered under Medicare Part A if patients meet homebound criteria. If patients do not meet homebound criteria, nursing is not covered for the vast majority. Nursing may be covered at home under a Medicare Advantage Plan. Also in the home, supplies for IVIG are not covered, whereas they are covered for SCIG.

For the last several years, HR 1845, the Medicare IVIG Access Act has been in place. The Act provides for a demonstration project, known as the Medicare IVIG Demonstration Project, to examine the benefits of providing coverage and payment for items and services necessary to administer IVIG in the home for patients with primary immunodeficiency disease (PI). The three-year project was scheduled to enroll up to 4,000 Medicare beneficiaries for whom it will allow some payment for nursing services and supplies. The project only applies to situations in which the beneficiary requires IVIG for the treatment of one of the 14 qualifying PI diagnosis codes. Patients receiving SCIG are not eligible for the project unless they wish to switch to IVIG. The demonstration project ended on September 30 of this year, and it is unknown at this time whether the project will affect coverage in the future.

One positive outcome of the 21st Century Cures Act for IG patients is the addition of payment for nursing and supplies effective in 2021. The amount of the payment has not yet been defined, but it can’t exceed the payment provided to physician office or hospital outpatient settings for similar services. Many patient organizations, providers and manufacturers are seeking a modification to the effective date to ensure access to care for patients receiving IG in the home setting.

Patients may qualify for Extra Help, a Medicare program to help people with limited income and resources pay Medicare prescription drug plan costs.

Know the Options!

Understanding coverage and the options involved with different sites of care and routes of administration is crucial as patients transition to Medicare. Especially when Medicare becomes their primary insurance, patients should be prepared to make changes in their care to optimize coverage. It truly seems that Medicare coverage can be more complex than IG therapy! But by discussing the coverage and options with someone knowledgeable about Medicare guidelines and IG therapy, patients can make the best choices for uninterrupted care when they consider these details well in advance of becoming eligible for Medicare.

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Editor’s note: This is an update of the article that appeared in the October-November 2015 issue of IG Living.
Temporary feelings of sadness and grief are a normal byproduct of chronic illness. But, when negative emotions linger and drift into hopelessness and despair, it may be time for medical intervention.

By Trudie Mitschang

**THE DIAGNOSIS OF** a chronic illness can alter a person’s life in more ways than one. The illness itself can lead to physical limitations, changes in activity levels and sleep habits, and even loss of employment and income. There are doctor appointments to schedule, medications to manage and, often, a barrage of emotions to navigate. With such an onslaught of physical and mental challenges, it’s common to experience feelings of sadness, grief or even anger. But, when feelings of intense sadness or hopelessness linger, it could be a sign of depression.

Statistics show that people living with chronic illness are much more likely to also experience symptoms of depression, and the risk is even higher for those who already have a history of depression. According to the American Psychological Association, depression is one of the most common complications...
of chronic illness. It is estimated up to one-third of individuals with a serious medical condition experience symptoms of depression. In fact, the rate of depression among individuals who have suffered from a heart attack is between 40 percent and 65 percent.\(^1\)

Unfortunately, because depressive symptoms can mimic the symptoms of many chronic illnesses, chronically ill patients dealing with depression are frequently misdiagnosed.\(^2\)

Here are some common warning signs of depression:
- Feelings of hopelessness and despair
- Changes in sleep patterns (insomnia or sleeping too much)
- Social isolation and withdrawal
- Weight loss or gain
- Loss of interest in hobbies and activities
- Frequent, unexplained crying
- Suicidal thoughts

While any chronic condition can trigger depression, the risk increases in proportion to the severity of the illness. And, when an illness leads to significant disruption in a patient’s life or daily routine, the odds increase even more. Treatment options for depression in chronically ill patients are the same as those for the general population and include psychotherapy, medication, lifestyle changes or any combination of these, depending on the patient and the severity of symptoms. For some, alternative therapies such as meditation, acupuncture or dietary supplements can effectively relieve milder symptoms. Seeing a physician for an accurate diagnosis of depression is always the first step to finding an effective treatment plan.

Talking It Out: How Behavioral Therapy Can Help

Because depression that is triggered by a life event like a chronic illness diagnosis is more likely to be linked to negative thought patterns and emotions (as opposed to a chemical imbalance), behavioral therapy is often an effective treatment choice.

Psychological therapy (also known as talking therapy) can help individuals battling depression by teaching them to change negative thought patterns and giving them specific coping skills. This type of intervention may also have long-term benefits that linger after therapy sessions have concluded, since a therapist can help patients identify thoughts, behaviors and triggers that could lead to a recurrence of depressive symptoms.

There are several types of effective psychological treatments for depression, as well as different delivery options. Some people prefer to meet one-on-one with a mental health professional, for example, while others find they get more out of a group therapy environment. “A behavioral medicine therapist will focus on how the chronic illness fits into the person’s life story, and where it may be causing problems. It all depends on what the person wants to work on in therapy,” says Tiffany Taft, PhD, co-founder of Oak Park Behavioral Medicine in Evanston, Ill. “Some people are newly diagnosed and are just trying to wrap their brains around the whole thing, so we can help with that through education and just giving the person a safe place to talk about their illness. Some people are anxious about their symptoms, so we help treat this anxiety by evaluating what’s triggering it and teaching relaxation techniques. Or, a person may be depressed and feeling isolated, so we help them cope and increase their social interactions.”\(^4\)

According to a report by the Institute of Medicine,\(^4\) mental health treatment goals for people with a chronic illness include:
- Learning to cope with the intense, sometimes debilitating, emotions related to their illness
- Changing behaviors to minimize the impact of their disease and maximize treatment protocol
- Managing disruptions their illness may cause to their work, school and family life

Types of Talk Therapy

There are several types of therapies that focus on changing negative thought patterns, and each uses specific intervention techniques. The most common therapies include cognitive behavior therapy (CBT), interpersonal therapy (IPT), mindfulness-based cognitive therapy (MBCT) and dialectic behavior therapy (DBT).

CBT is a structured psychological treatment that focuses on how the way people think (cognition) and act (behavior) affects how they feel. CBT is considered one of the most effective treatments for depression and has been found to be useful for a wide range of ages, including children, teens, adults and seniors.\(^1\) CBT
involves working with a licensed mental health professional to identify thought and behavior patterns that are either triggering depression symptoms or blocking the recovery process. “The basic idea of CBT is how we think affects how we feel emotionally and physically, and that affects how we behave,” says Dr. Taft. “I like CBT because it empowers people. We ultimately only have control over ourselves and how we think and react to what life brings us. So one of the first things we have clients do is start paying attention to their self-talk, especially as it relates to their illness.”

IPT is a structured psychological therapy that focuses on problems in personal relationships and the skills needed to deal with them. It is based on the idea that relationship problems can have a significant effect on someone experiencing depression, and can even contribute to the cause. In the case of a chronically ill patient, the illness can cause significant rifts in marriages, friendships and work relationships, leading to feelings of isolation and depression. IPT helps individuals recognize negative patterns in relationships and offers communication techniques and coping skills to improve relationship dynamics.

MBCT involves an intervention called mindfulness meditation. This technique teaches patients to focus their thinking on the present moment while paying attention to feelings, whether pleasant or unpleasant, without trying to resist or change anything. Initially, this approach focuses on physical sensations (like breathing in and out), but eventually it moves on to feelings and thought patterns. MBCT is thought to be effective in preventing depression from returning because it encourages patients to notice feelings of sadness and negative thinking patterns early on, before they become a habit.

One study noted that MBCT was especially effective when it came to treating individuals who had been unsuccessful with other interventions, including behavioral therapy and medication. According to the study authors, “The results of this preliminary audit suggest that MBCT is an acceptable treatment for patients who have only had a partial response to antidepressant medication and/or standard individual CBT. Further, for many patients, it appears to be effective in significantly reducing levels of depression, even in those who start with a more severe pattern, including suicidal depression.”

DBT is a psychological treatment that combines CBT with two additional techniques:

- Dialectics, which relies on discussion or dialog to explore and resolve issues
- Mindfulness, which encourages individuals to become more aware of and present in the moment, so that concerns about the future or rumination about the past do not interfere with their ability to enjoy life

DBT is particularly effective for patients who tend to view life and circumstances as polarized (right or wrong). The technique was originally developed as a treatment for patients suffering from borderline personality disorder, but in recent years, it has been proven effective for treating patients with a variety of symptoms and behaviors associated with mood disorders, including depression. Patients who decide to pursue DBT as a treatment option participate in both individual and group sessions during therapy.

The Medication Debate

When talk therapy is not enough, patients may be prescribed medication to treat depression. These medications, also known as psychotropic drugs, can be highly effective but are known to come with a variety of risks and side effects. For patients already dealing with a chronic illness and a potentially lengthy list of existing prescriptions and medications, adding an additional pharmacological intervention should only be considered under the close supervision of a medical professional.

Psychotropic drugs, also known as antidepressants, typically work by changing or balancing the amount of important chemicals in the brain called neurotransmitters. Some mental health issues show improvement when neurotransmitters such as dopamine, serotonin and norepinephrine are increased or decreased. Psychotropic drugs are usually prescribed by a psychiatrist, a psychiatric nurse practitioner or a primary care physician (as opposed to a behavioral therapist). In some states, clinical psychologists may be able to prescribe antidepressants.6

For situational depression, such as the type triggered by a diagnosis of chronic illness, psychotropic drugs in combination with psychotherapy may be recommended, with the medication serving as a supplement to rather than a replacement for therapy.
The most commonly prescribed psychotropic drugs include:

- Duloxetine (Cymbalta)
- Trazodone (Desyrel)
- Venlafaxine (Effexor)
- Fluoxetine (Prozac)
- Sertraline (Zoloft)
- Escitalopram (Lexapro)

As with all medicines, the use of antidepressants can lead to side effects. Some, like jitteriness, strange dreams, dry mouth or constipation, may go away on their own. Others, like sexual dysfunction, may linger. It’s important to note that side effects from antidepressants vary from patient to patient and can sometimes be alleviated by simply switching medications. Anyone embarking on a treatment plan using prescription antidepressants should be closely monitored by a physician who can keep tabs on unwanted side effects and make dosage adjustments as needed.

How Lifestyle Changes Can Help

Patients diagnosed with chronic illness have typically experienced unwanted lifestyle changes such as loss of mobility, digestive problems that lead to dietary restrictions, and poor sleep quality. These changes alone can trigger circumstantial depression. While advice to “get out more” or “eat healthier” can sound trite in the face of serious depression, the fact remains that in milder cases, lifestyle adjustments have proven extremely beneficial.

Eating to beat the blues. Individuals suffering from mild depression can benefit from avoiding foods high in refined sugar and saturated fats. Instead, they should incorporate healthy foods containing omega-3 and omega-6 fatty acids, including those found in fish, nuts, fresh fruits and vegetables and olive oil.

Amino acids are also a healthy dietary aid when it comes to banishing depression. Neurotransmitters, the messengers in the brain, are made of amino acids and play an important role in mental health. Foods rich in amino acids include animal proteins; legumes, nuts and seeds; dairy products; and whole grains.

The role of exercise in mental health. Physical activity increases your body’s production of natural antidepressants. According to the Mayo Clinic, exercising for just 30 minutes a day three to five days a week can help alleviate depression without the need for medication. Regular exercise eases depression in a number of ways, including:

- Releasing feel-good brain chemicals that may ease depression (neurotransmitters, endorphins and endocannabinoids)
- Reducing immune system chemicals that can worsen depression
- Increasing body temperature, which may have calming effects

Addressing Depression as a Part of the Healthcare Plan

For the chronically ill, successful treatment of depression is an important component of the overall healthcare plan. Addressing mental health issues leads to improved symptom management and quality of life, and in many cases, higher degrees of compliance when it comes to other methods of care. According to the National Institutes of Health, more than 80 percent of depressed people can successfully be treated using talk therapy, antidepressant medication or both.

FOR THE CHRONICALLY ILL, SUCCESSFUL TREATMENT OF DEPRESSION IS AN IMPORTANT COMPONENT OF THE OVERALL HEALTHCARE PLAN.

When considering treatment options for depression, especially if it was triggered or exacerbated by a chronic illness diagnosis, doctors are advised to first do a thorough review of the treatment plan for the chronic disease itself. By taking an inventory of current medications, addressing patient compliance issues and even meeting with caregivers and family members to address emotional support concerns, mental health improvements may be attained without the need for specific depression-related intervention.

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.

References

Important Safety Information

Hizentra treats various forms of primary immunodeficiency (PI) in patients age 2 and over.

WARNING: Thrombosis (blood clotting) 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 thrombosis, your doctor will prescribe Hizentra at the minimum dose and infusion rate practicable and will monitor you for signs of thrombosis and hyperviscosity. Always drink sufficient fluids before administration.

Tell your doctor if you have had a serious reaction to other immune globulin medicines or have been told you also have a deficiency of the immunoglobulin called IgA, as you might not be able to take Hizentra.

You should not take Hizentra if you know you have hyperprolinemia (too much proline in your blood).

Infuse Hizentra under your skin only; do not inject into a blood vessel.

Allergic reactions can occur with Hizentra. If your doctor suspects you are having a bad allergic reaction or are going into shock, treatment will be discontinued. Immediately tell your doctor or go to the emergency room if you have signs of such a reaction, including hives, trouble breathing, wheezing, dizziness, or fainting.

Tell your doctor about any side effects that concern you. Immediately report symptoms that could indicate a blood clot, including pain and/or swelling of an arm or leg, with warmth over affected area; discoloration in arm or leg; unexplained shortness of breath; chest pain or discomfort that worsens with deep breathing; unexplained rapid pulse; and numbness or weakness on one side of the body. Your doctor will also monitor...
Before being treated with Hizentra, inform your doctor if you are pregnant, nursing or plan to become pregnant. Vaccines (such as measles, mumps and rubella) might not work well if you are using Hizentra. Before receiving any vaccine, tell the healthcare professional you are being treated with Hizentra.

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.

Voice2Voice gives you an opportunity to connect with others who have been in your shoes. Dealing with PI can be a challenge, and knowing someone who truly understands what you’re going through can mean a lot.*

It’s good to know you’re not alone.

Sign up at voice2voice4pi.com

You may also call 1-877-355-IGIQ (4447)
Monday–Friday, 8 AM to 8 PM ET
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.

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

- Thrombosis may occur with immune globulin products, including Hizentra. Risk factors 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.

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**INDICATIONS AND USAGE**
Hizentra is an Immune Globulin Subcutaneous (Human) (IGSC), 20% Liquid indicated for the treatment of primary immunodeficiency (PI) in adults and pediatric patients 2 years of age and older.

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**DOSE AND ADMINISTRATION**
For subcutaneous infusion only.
Administer at regular intervals from daily up to every two weeks (biweekly).

**Dosage (2.2)**
Before switching to Hizentra, obtain the patient’s serum IgG trough level to guide subsequent dose adjustments.

- **Weekly:** Start Hizentra 1 week after last IGIV infusion
  
  Initial weekly dose = Previous IGIV dose (in grams) × 1.37
  
  No. of weeks between IGIV doses

- **Biweekly:** Start Hizentra 1 or 2 weeks after the last IGIV infusion or 1 week after the last weekly Hizentra/IGIV infusion. Administer twice the calculated weekly dose.

- **Frequent dosing (2 to 7 times per week):** Start Hizentra 1 week after the last IGIV or Hizentra/IGSC infusion. Divide the calculated weekly dose by the desired number of times per week.

- Adjust the dose based on clinical response and serum IgG trough levels.

**Administration**
- Infusion sites – 1 to 4 injection sites simultaneously, with at least 2 inches between sites.

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**ADVERSE REACTIONS**
The most common adverse reactions observed in ≥5% of study subjects were local reactions (i.e., swelling, redness, heat, pain, and itching at the injection site), headache, diarrhea, fatigue, back pain, nausea, pain in extremity, cough, rash, pruritus, vomiting, abdominal pain (upper), migraine, and pain.

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

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

**USE IN SPECIFIC POPULATIONS**
- Pediatric: No specific dose requirements are necessary to achieve the desired serum IgG levels.

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**DOSAGE FORMS AND STRENGTHS**

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Based on October 2016 revision
Between five and 30 out of every 100 people have a deficiency that many have never heard of: mannose-binding lectin deficiency, also known as MBL deficiency. People with this condition have low levels of MBL in their blood. Part of the complement system that helps protect the body from infection, MBL plays an important role in defending the body against invading microorganisms, including yeasts, parasites, viruses and some types of bacteria. MBL is produced by the liver and released into the blood, where it recognizes mannose residues present on a wide range of common pathogens and binds to them. This binding initiates complement activation of the lectin pathway, which is the first part of the complement system to react to invading microbes.

“In many individuals, MBL deficiency is asymptomatic (since the other pathways of complement activation are still intact), but in some people, it can lead to recurrent bacterial infections,” says Bob Geng, MD, MA, assistant professor in the division of allergy and immunology at the University of California, San Diego. “However, deficiency in MBL can predispose individuals to develop more severe symptoms if they have chronic inflammatory conditions. In addition, MBL deficiency in addition to another known defect of the immune system can lead to more severe conditions.”
severe and frequent bacterial infections” such as pneumonia and meningitis.

MBL deficiency is diagnosed with a blood test, and is then classified as mild, moderate or severe based on how low the MBL levels are. Depending on the type of infection, symptoms vary in frequency and severity; and treatment is based on the severity of symptoms.

**MBL Deficiency in Children**

Since the 1970s, researchers have recognized that MBL deficiency affects children, namely those between ages 6 months and 18 months. Early research revealed that failure to thrive and recurrent infections characterized by severe local inflammation were associated with the deficiency. Current research supports that MBL deficiency is especially hard on children, whose immune systems are immature and who therefore have an increased susceptibility to infections.

**Since the 1970s, researchers have recognized that MBL deficiency affects children, namely those between ages 6 months and 18 months.**

Pneumonia is common in children, and recent studies suggest impairments of the innate immune system may account for these infections. “It is crucial to identify patients with such impairments to better manage and prevent future complications,” say immunologists Michelle Halbrich, MD, Moshe Ben-Shoshan, MD, and Christine McCusker, MD, who add that even in cases with mild clinical presentations, a high level of suspicion of an underlying immune deficiency is needed.

For some children, issues with MBL deficiency can persist beyond 18 months old. One parent describes her struggle with her two daughters, both of whom have MBL deficiency: “My girls are now 9 and 10, and after years of chasing this down, both are doing much better.” She credits early detection and treatment of infections with her children’s improvement. “I am more aware, as well as doctors, so antibiotics are immediate — no waiting until they are really sick,” she says. One child is also on long-term low-dose antibiotics to help combat her infections, the same measure that is sometimes also taken with other forms of immunodeficiency.

**MBL Deficiency in Adults**

Initially, MBL deficiency was believed to exclusively affect children, not adults whose immune systems have matured. Before discussing the potential issues MBL deficiency can have in the adult population, it’s important to contextualize that conversation. For many people, a low MBL level, in and of itself, causes no health issues. In an online question-and-answer about the condition, Phil Lieberman, MD, clinical professor of medicine and pediatrics in the Departments of Internal Medicine and Pediatrics at the University of Tennessee College of Medicine, underscores this point. “First, I think it is important to understand that many individuals who have mannose binding lectin (MBL) deficiency do not suffer any adverse symptoms related to the diminished levels of MBL,” he writes.

MBL deficiency can, however, cause issues for some adults. The view that the condition only affects children was first challenged in 1995. A group of researchers showed that adults with the condition could have severe and unusual infections in which MBL gene mutations were the only identified cause of immunodeficiency. These infections included skin abscesses, chronic diarrhea associated with a type of parasite, meningococcal meningitis with recurrent herpes simplex, and a form of fatal necrotizing pneumonia. The patients ranged in age from 15 years to 56 years and were both male and female.

Since that study, MBL deficiency in adults has also been associated with recurrent bacterial sinusitis, more severe forms of recurrent community-acquired pneumonia, pneumococcal sepsis, E. coli-induced pyelonephritis, fallopian tube infections and persistent hepatitis B.

A 2013 study paints a clear picture of what it can be like to suffer myriad health issues at significantly higher rates than those who don’t have the condition. These include higher rates of:

- Bacterial infections of the vagina
- Bronchitis
- Common cold
- Conjunctivitis
- Cystitis
- Gastritis
- Gingivitis
- Esophagitis
- Onychia
• Otitis media
• Pharyngitis
• Pneumonia
• Prostatitis
• Sinusitis
• Stomatitis
• Tracheitis
• Urethritis

The study also showed that those with MBL deficiency had a 43 percent chance of having suffered from recurrent and severe episodes of infections, compared with only 16 percent in the control group. And, antibiotic treatment was significantly more common among MBL deficiency patients.12

Certain groups of adults are at greater risk when they have MBL deficiency. These include patients with suppressed immune systems such as those who have another form of immunodeficiency in addition to the MBL deficiency. Dr. Liebermann recommends people with low MBL deficiency look for associated immunological defects such as hypogammaglobulinemia.11 Also at greater risk are those who have had an organ transplant, and those who are undergoing chemotherapy.1,11

Adults with autoimmune diseases can also be negatively affected by MBL deficiency. Diseases such as systemic lupus erythematosus and rheumatoid arthritis have a more severe course in the presence of MBL deficiency. In cases of cystic fibrosis, MBL deficiency leads to a poorer prognosis.5

Therapies for MBL

Treating MBL deficiency isn’t a one-size-fits-all approach. For those who don’t have health issues associated with the condition, treatment is unnecessary.1,11 For those with associated health issues, a three-pronged approach is employed. First, antibiotics, including prophylactic antibiotics, are used for infection control. Second, vaccinations are recommended. And third, patients are encouraged to have their antibodies checked to determine if an additional form of immunodeficiency is present.13,14

“Both recombinant (synthetically made) and plasma-derived MBL products are commercially available, but are still currently under research investigation as a form of replacement therapy,” says Dr. Geng. “The most likely indication for this investigational therapy may be patients diagnosed with MBL deficiency plus another known defect of the immune system who are suffering from severe acute bacterial infections.” However, according to Terry O. Harville, MD, PhD, medical director in the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, “because these therapies have a very short half-life, they are not practical except perhaps in acute illnesses.”

In the future, because a certain gene has been found to cause this condition, gene therapy may be a viable treatment.15

CERTAIN GROUPS OF ADULTS ARE AT GREATER RISK WHEN THEY HAVE MBL DEFICIENCY.

Living with MBL

“MBL deficiency and treatment remains controversial,” explains Dr. Harville. “Data indicate that 30 percent of the population in Denmark are MBL deficient without excessive complications, so many are not convinced it is a problem.” Nevertheless, it is one of the most common human immuno-deficiencies, and several studies have shown that deficiency of MBL increases the overall susceptibility of an individual to infectious disease. As such, more research is needed to better treat those diagnosed who do suffer from recurring infections.

DANA HENRY is a writer and editor in the Kansas City area who specializes in science, medicine and health.

References
Trudie: Tell us about your diagnosis and health history.

Chelsey: I was diagnosed with CVID 15 years ago, and with hereditary angioedema (HAE), a rare inherited blood disorder, five years ago. Like many who have complications from CVID, I have a history of cancer, hemolytic-uremic syndrome, numerous surgeries and so many infections that even if I tried, I would never be able to remember them all. With HAE, I sometimes have extreme flares that seem to go through cycles, while other times, I can go for months without any swelling episodes.

Trudie: When did you begin being treated with intravenous immune globulin (IVIG)?

Chelsey: I started IVIG infusions when I was 12 years old. Because I was young, I chose the denial route for a lot of years. I wanted to be able to keep up and do everything that a typical healthy person is able to do. Around age 18, I decided that I wanted to take a break from my IVIG infusions; I was doing well, so I thought maybe I didn’t need them anymore. I have a wise doctor who let me take a break so I could see for myself how much it was helping me. He explained to my mother that no one can make someone do infusions — they must want to — and he was right. After completing my sophomore year of college, I became so sick that I begged to start the infusions again.

Trudie: What considerations did you have when starting a family?

Chelsey: Because of the type of chemotherapy I received when I was young, my oncologist told me that if I wanted to start a family, I had to have all my kids before I turned 30. That was hard because it didn’t give me a lot of time to decide what I should do. My husband and I love kids and always wanted to have a large family, but there was a huge concern about passing on my immune deficiency to our children. We were still debating about what to do when we got the unexpected news that I was pregnant. It was a very difficult pregnancy (it was during the pregnancy when I was diagnosed with HAE). My son Trace, who is now 5 years old, was born on our three-year wedding anniversary. My daughter Alana is 3-and-a-half years old, and my youngest daughter Lilia is 22 months old. I feel very blessed because, so far, all of my children are incredibly healthy. My children are the greatest blessing I have in my life. If I had to do it over again, I would still have them because I honestly can’t imagine life without them.

Trudie: Where do you have your infusions?

Chelsey: For about 10 years, I

PROFILE: Chelsey Safken

By Trudie Mitschang

CHELSEY SAFKEN was just shy of her 16th birthday, when she was diagnosed with osteosarcoma (bone cancer). Since then, this resilient young wife and mother of three children aged 5 years and younger has battled multiple setbacks and illnesses, including a diagnosis of common variable immune deficiency (CVID). While many might feel discouraged by such a deluge of health challenges, Chelsey’s positive attitude and strength have helped her overcome adversity and live life to the fullest — no matter what.
received my infusions at an infusion center. When I had my son, I switched to home care. I absolutely love having my infusions at home, and my nurse has become part of the family. I receive Cinryze infusions for HAE twice a week, and my IVIG infusions every two weeks.

**Trudie:** Have you had any challenges with insurance coverage?

**Chelsey:** I think the hardest part of dealing with insurance companies is when I have to switch policies. Sometimes trying to get authorization takes time and, on occasion, I’ve had to miss infusions while waiting for approval. No matter what, I always know within the first couple weeks of January, I will hit my out-of-pocket maximum for the year. Knowing medical bills are a major part of our budget, my husband and I always have to factor that into our expenses. Thankfully, there have been some amazing programs that have helped cover some of the medical expenses, which I am so grateful for.

**Trudie:** How does chronic illness impact your marriage?

**Chelsey:** Sometimes my husband has to take on extra responsibilities around the house and with the kids due to my health. He’s usually really good about it, because he knows if I rest, I typically rebound a lot quicker. I’ve never been one to sit on the sidelines, so it’s really hard for me to rest. He is always the one telling me I need to take it easy and not push myself too hard. My kids are now starting to ask a lot more questions about why I always have to go to the doctor. I try to answer them openly and honestly because I don’t want them to be afraid, but I also shield them because they are young.

**Trudie:** Do you belong to any support groups?

**Chelsey:** I don’t belong to a support group, but I have an incredibly supportive family. I have a large extended family and am really close to my parents and siblings. They are always there for me, and I lean on them a lot.

**Trudie:** Do you have a diet or exercise regimen to manage your health?

**Chelsey:** I don’t follow an exact exercise routine, but I am incredibly active. I love to go for walks and hikes with my family. I feel staying physically active is incredibly important to staying healthy. It helps boost my mood and body. As far as diet goes, I try to eat only fresh foods and avoid any processed foods. I eat a large variety of fresh fruits and vegetables, especially those that I know have anti-inflammatory properties.

**Trudie:** You’re a regular contributor for the IMMUNOe blog. How did that come about?

**Chelsey:** I started writing because IMMUNOe approached me. They wanted to have a blog from a patient’s perspective, and I began writing it to help others.

**Trudie:** What’s been the most difficult part of living with chronic illness?

**Chelsey:** I spent a lot of years living with a real fear that if I admitted being sick, illness would define me. I now realize that my disease isn’t who I am, but it’s definitely a part of me. I’ve now learned how to set boundaries for myself that help me to stay well.

**Trudie:** What frustrates you most?

**Chelsey:** I think people really misunderstand what a genetic immune deficiency is. Most people have only heard of acquired immune deficiency, and they are very different things. I also wish people understood it’s not a laziness issue or an excuse when I need rest. It’s a real condition, and just because I look totally normal doesn’t mean that I am. I feel like it’s a blessing and a curse because with these diseases, I usually look completely healthy.

**Trudie:** What has living with illness taught you?

**Chelsey:** I don’t want my life to be limited by the things I missed out on or by my health conditions. I have learned the importance of maintaining a healthy body both physically and emotionally; wellness is the whole person. Even though having rare diseases brings a lot of complications with everyday life, it has also taught me a lot of good things. The most important lessons I’ve learned are that every moment is precious and life needs to be lived to the fullest!

**The most important lessons I’ve learned are that every moment is precious and life needs to be lived to the fullest!**

**Trudie Mitschang** is a contributing writer for *IG Living* magazine.
SOMETIMES WHILE waiting in line at the deli in the grocery store, I notice the person serving customers has minimal enthusiasm. No facial expression. Definitely no conversation. Just doing his or her job. Then, when it’s my turn, there’s a shift. The person perks up, and we talk about the day and what’s going on. We wish each other a good day, and we mean it. That happens to me with the teller at the bank, too. A nonanimated face suddenly becomes alive. We connect and feel good when we say goodbye. In fact, the same is true for most of my interactions, whether it’s with a nice waitress, a helpful salesperson, etc. Truthfully, I never really thought about it until I heard other people talking about how rude or unfriendly someone was to them, when my experience at the same place was pleasant and kind. But, recently, I figured out why.

I was patiently sitting in an administrative office waiting to settle some paperwork as the staff passed by, getting on with their day. As one passed by, she said: “You’re happy.” I thought to myself: “Not especially. I wonder why she’d say that?” A few minutes passed by and the papers were handed to me. The supervisor said: “It is always such a pleasure to have you come by. Every time I see you, you’re smiling. You can stop by anytime.”

Aha! Smiling! Really? I didn’t even know I was doing it. That must be my resting face. Then I thought: “Doesn’t everyone smile?” It can’t be that unusual. So, I started looking at the faces around me. I never realized how many people don’t smile. I don’t mean you have to go around brimming with happiness even if you don’t have a legitimate reason not to smile. But the majority of people I encounter seem to be expressionless or look angry. Not only do they seem unapproachable, but it’s like they want to pick a fight or don’t want to bother being civil.

So much of our day is made up of small interactions. We can start them off on a positive note, without saying a word, by just smiling. From the musical “Annie,” I remember a line from a song: “You’re never fully dressed without a smile.” No one knows I have an autoimmune disease; actually, three of them. Or that I get infusions several times a month. Or that I’ve been poked and prodded so much I should be walking around with a permanent scowl on my face. But the smile is a choice. Maybe it is one I’ve made so deep inside my psyche I don’t even realize it; it’s just part of my nature now. My day-to-day relationships are brighter, more engaging, fun and surprising. I’m not saying they are like that all the time, but most are.

Here’s the real secret: It makes me feel happy. Who doesn’t want to feel good? We are all in this mess of a life together trying the best we can. We can learn from each other, compliment each other, have moments of kindness with each other and giggle with each other. Please think of that the next time you get coffee or pick up your meds from the pharmacy. So, look up and out from this magazine and smile. Like Louis Armstrong sang in his famous song: “When You’re Smiling”: “When you’re smiling, when you’re smiling, the whole world smiles with you.”

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.
An Accessible Apartment

By Ilana Jacqueline

WE ARE MOVING. Again. This brings me such intense joy. In my mind, I am already living in our new place. I think about it every time I have to let my dog out to go to the bathroom on our one square foot of backyard between our patio wall and the hedges. I think about it every time I lug myself, my laptop, laundry or infusion supplies up our wobbly spiral staircase.

We have known we needed to move for almost three years, but we could not settle on the right place (and I’m sure many of you have had your own sticker shock at the cost of rent these days). I’ve had plenty of time to think about all the things in my current apartment that make my life harder than it has to be. I’ve also had plenty of time to think about what I want in an apartment that will make my life easier for someone with fatigue, immune deficiency and allergies.

A sad state of the stairs. I’m making a vow right here and right now: I will never live in another two-story dwelling again. I don’t care if I win the lottery and can afford a five-story house! Stairs are the worst. Stairs are the worst. And I’ve really tried my best. I have two infusion kits — one on the first floor, one on the second. The only problem is when I’m mid-infusion upstairs and the doorbell rings, or dinner is ready or the dog needs to be let out, I end up carrying the pole with me downstairs anyway (on a spiral staircase). I also have to carry clothes baskets up and down the stairs, and when my dog is sick or injured, I have to carry him up and down the stairs. So many surgeries this year resulted in me either sleeping on our couch or just going to my mother’s to recover, since I couldn’t get up or down the stairs. What is the point of having your own apartment if it’s completely inaccessible when you need it most?

Bathroom blues. When we first moved into this apartment and I saw the giant — and I mean enormous — bathtub in the master bath, I was floored. This was my first place, and I had never imagined I’d find an apartment with a bathtub as big as a jacuzzi. I was so excited. It took me about an hour to fill it. The water was pretty cold by then, but that was OK! This tub had jets! What luxury. I couldn’t believe I was able to access this tub every single night! And then I turned on those jets and watched with slow horror as the water turned to mud.

It had not occurred to me that the apartment was 20 years old, or that the previous tenants had never used the jets or cleaned them. I spent most of the night bleaching the bathtub and wondering if there was a place that sold jet covers to keep those pathways from making contact with the bath water. I finally got some instructions on how to flood the inside tunnels with cleaning solution, but I just didn’t know how clean I could really get it in such a state. And with a port and immune deficiency — well, it just wasn’t worth the risk. I stuck to showers in my big, empty bathtub for five years. I’m ready to have a small bathtub again. One with no hidden corners where bacteria can collect out of sight and will take only five minutes to clean!

A wish for windows. This might sound silly. What kind of apartment doesn’t have windows? Apparently, the kind that comes with a bathtub the size of a walk-in closet. Yes, the layout of my apartment is bizarre. We have a loft with a skylight and sliding glass back doors, but we don’t have any other windows. We can’t open the skylight and let the breeze through, or air out the house after cooking something on the stovetop. As a result, the apartment is often stuffy, and we limit what we cook at home since we don’t want to breathe in the smoke with no way to vent the scent or heat.

Adoring your flooring. While they are inarguably more labor intensive to sweep and scrub than carpet, tile or wood floors make a huge difference for patients with allergies. This was definitely one of the advantages of this apartment, which had zero carpeting. Even though the floors were hard to take care of and constantly needed to be swept, vacuumed or mopped, I at least always knew when there was a spill or dust. Keep these factors in mind next time you go apartment hunting.

Remember as well that under the Fair Housing Act of 1998, you are permitted to make modifications to an apartment that doesn’t fit your disability needs. For example, you can install grab bars in the showers, add wheelchair access for ramps or widen doorways. You may have to pay for these modifications, but you are legally entitled to make them (hud.gov/offices/hudo/disabilities/reasonable_modifications_mar08.pdf).

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Helping Children Understand Long-Term Illness

By Jessica Leigh Johnson

AFTER TWO YEARS substitute teaching in elementary schools, I’ve learned one thing: All kids get sick — not just mine. There isn’t a single month in the school year when some child isn’t coughing, running a fever or going to the nurse’s office with a tummy ache. There’s always something going around, and eventually, the majority of kids in school will catch it. But these common ailments are minor. They may hit hard and fast, but they go away just as quickly, and the children bounce back and return to school as if nothing happened.

That’s not the case with chronic illness. Chronic health conditions don’t come and go. There may be medications to make symptoms more bearable or to prolong damage to organs and tissue, but there is no easy cure. Even though children who suffer from chronic illness may not feel “sick” every day, the illness still affects daily life and development, even into adulthood.

What Makes an Illness “Chronic”?

A chronic health condition is defined as a health problem that lasts longer than three months, affects normal activities and requires extensive medical care, including hospitalizations and/or the need for home healthcare. Chronic illness is a general term referring to conditions such as asthma, diabetes, primary immunodeficiency disease, epilepsy, cerebral palsy, cystic fibrosis and many others. Any condition people must live with every day of their life is considered chronic.1

Based on this definition of chronic illness, researchers at the University of Michigan estimate about 15 percent to 18 percent of children in the United States suffer from a chronic health condition.

How Do Parents Explain Illness to their Child?

It’s hard on both children and their families when a chronic illness is diagnosed. With frequent doctor appointments, lonely hospital stays and scary and sometimes painful medical treatments, children may wonder what is happening to them. And while it’s typically up to parents to explain, they may be wondering the same thing. With all the complicated, technical terminology attached to chronic medical conditions, how do parents explain the details of the illness to their children in a way they will understand?

When explaining chronic illness, several factors need to be considered, including the children’s age, personality and the specific illness in question. Kids’ understandings of illness, as well as their coping strategies, change as they grow older.1

Infants and toddlers. Because of their young age, infants and toddlers will have very little understanding of their illness, so they don’t require a lengthy explanation with big, fancy words. What they need is to feel safe and secure. When they experience pain, whether from the condition itself or from a treatment, that feeling of safety is threatened. When their freedom of motion is restricted (think of a nurse trying to insert an IV into a toddler) or they’re separated from parents, it’s hard for them to develop a strong sense of trust and security. The best thing parents can do to communicate a sense of comfort to their young children is to be present during hospitalizations, stay close for painful or frightening procedures and hold, sooth and interact with their babies as much as possible.1 The more detailed explanations of the illness will come later.

Preschool children. Children in preschool or just starting school are beginning to develop a sense of independence. Being in the hospital or having to comply with a new medication regimen can challenge this budding independence, so it’s important for parents to convey the purpose and necessity of hospital stays and medical treatments using age-appropriate terms. Otherwise, children may try to regain a sense of control by testing the limits set by their parents. They may refuse to take medicine or sit still for a necessary blood test.

While children in this age group are old enough to understand they’re sick or that something in their bodies isn’t working quite right, they probably don’t understand why or how exactly their illness will impact their life long-term. Also, at this age, children are afraid of pain and of being hurt, so it’s best for parents to be upfront and honest about tests and procedures that may cause pain, and explain that the treatment is being done to help them, not harm them.2 Before each treatment or procedure, children should know why it’s being done, who will be doing it, what equipment will be used and whether it will be painful or uncomfortable. All information should be relayed in the context of the children’s developmental level. If parents feel underprepared for this task,
most pediatric hospitals have child life specialists who help prepare children for hospitalization, surgery and various medical procedures.1

School-aged children. School-aged children may try to figure out what caused their illness, and they may think it’s their fault. Maybe they were mean to a younger sibling or disobeyed their parents, and they think the illness is their punishment. Parents must reassure school-aged children that the disease is in no way a result of bad behavior.

It’s common for parents to struggle with how much to share with their children regarding their illness. They may worry explaining the details will cause their children extra worry or stress. But, it’s usually best to be upfront and honest with kids, even when talking about something that may cause them concern. While parents shouldn’t give too many details for the children to process, parents also must not withhold facts or information. Otherwise, children may overhear a doctor or nurse talking about their condition and, without complete understanding, may begin to imagine the worst.2 Besides, children are perceptive and can usually tell when a parent isn’t being completely honest or is hiding something. Withholding the truth damages the bond of trust between parents and children that is so important when dealing with a chronic illness. If children have questions, it’s best for parents to answer them in an honest and direct manner. If parents are unsure how to explain the illness or don’t know quite what to say, the children’s doctor or a social worker may be able to offer suggestions.3

Adolescents. Teenagers and young adults can understand a more complex explanation of their illness and may have many detailed questions. As they are developing a sense of independence from their families, they may want to be more involved in making decisions regarding their treatment. Parents should encourage their adolescents to monitor their illness and manage their own treatment as much as possible.2 This will help them as adults when they have to take control of their health.

When explaining chronic illness, several factors need to be considered, including the children’s age, personality and the specific illness in question.

Adolescents will still have plenty of questions regarding their condition, and parents can encourage them to share those concerns with healthcare professionals as they further develop their own identity separate from their families.1

Another important thing to keep in mind is adolescents’ bodies are rapidly changing, so parents need to keep an eye on any unusual symptoms or side effects of medication that would require dosage changes. Some medical conditions are hard to manage during the turbulent teen years, so it is imperative that parents and teens keep an open line of communication about the possibility of any side effects due to the illness or treatment. Rebellion can also affect teens’ health in a negative way. If adolescents choose not to adhere to their treatment regimen, the chronic illness could reach an unstable state. And, if that happens, it is most helpful if parents encourage discussion about what is happening rather than reprimand.1

Needed Conversations

No parent wishes a chronic illness upon their children, and the topic can often be difficult and uncomfortable to address. But if children live with a chronic condition, these conversations need to take place. Regardless of their age, children with long-term illness should be free to discuss any feelings or concerns about their illness with their parents. As children grow, their thoughts and feelings about their illness may change. By cultivating a habit of open and honest communication, parents will gain a better understanding of how to best support their children through this chapter of their lives.

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

PRODUCT GUIDE

Itching for Relief

By Trudie Mitschang

SKIN CONDITIONS like rashes are certainly not unique to individuals with primary immunodeficiency diseases (PI). While troublesome complaints of eczema or psoriasis frequently occur in people with normal immune systems, skin diseases are often one of the earliest visible symptoms of PI. And, as many PI patients have learned, certain types of rashes and skin conditions are also a side effect of intravenous immune globulin (IVIG) treatments. “A rash is a very common side effect of IVIG, and some patients develop worse rashes than others,” says Abbie Cornett, IG Living’s patient advocate. “If a certain brand of IG is triggering a reaction, it’s possible that switching to another brand could help. Taking premedications such as steroids or Benadryl before treatments can also sometimes reduce or eliminate some of the side effects.”

For PI patients who are bothered by recurring rashes, it may be helpful to keep a health diary that tracks any common patterns or triggers to devise the best treatment plan and minimize discomfort. Getting to the root cause is always the first step toward healing.

What Are Rashes?

A rash refers to any sort of inflammation and/or discoloration that distorts the skin’s normal texture or appearance. A skin rash may be characterized by redness, blisters, bumps, irritation, itching or a scaly appearance. Medically speaking, there are numerous types of rashes:

• IVIG-triggered rashes, which may appear as a blistering type of eczema, often begin approximately eight to 10 days after exposure to the medication. These skin lesions often resolve within a period of one to four weeks.¹

• Eczema or atopic dermatitis, a rash that primarily occurs in people with asthma or allergies, is often reddish and itchy with a scaly texture.

• Psoriasis is a common skin condition that can appear scaly, itchy and red, and is frequently found on the scalp, elbows and joints.

• Seborrheic eczema is a type of eczema that most often affects the scalp and causes redness, scaly patches and dandruff. It can also occur on the ears, mouth or nose. In babies, this rash is known as “crib cap.”

• Lupus erythematosus is an autoimmune disease that triggers a rash on the cheeks and nose. It is sometimes called “butterfly,” or malar, rash.

• Rheumatoid arthritis is an autoimmune disease that can cause a rash to form on various parts of the body.

• Contact dermatitis, a common rash that causes redness, itching and, sometimes, small bumps, occurs when a person comes in contact with an irritant such as poison ivy.

Diagnosing and Treating Rashes

Rashes can be tricky to diagnose since some develop right away, while others appear several days after the trigger event. In addition, many rashes clear up on their own and can be treated with over-the-counter products. At-home treatments may include moisturizers, lotions, baths, cortisone creams that relieve swelling, and antihistamines that relieve itching.

Mild rashes and skin conditions that don’t respond to at-home treatments can be diagnosed and treated by a primary care provider or an immunologist. More severe skin conditions may require treatment by a dermatologist who specializes in skin diseases. Treatment may include local application of moisturizing lotions and steroid ointments directly to the rash. If symptoms don’t improve, the provider may prescribe topical ointments containing more potent steroids or other immunosuppressant medications. Rarely, oral or intravenous immunosuppressant medications may be needed to treat severe skin rashes and relieve symptoms.

Don’t Take Skin Rashes Lightly

Skin is the largest organ in the body, and when battling infection or responding to the stress of IVIG, it’s natural for occasional visible external reactions to appear. Because skin plays an important role as a barrier to bacteria and other organisms in the environment, severe rashes that ooze or bleed can serve as an entry point for bacteria into the bloodstream, and should not be taken lightly. If a rash does not respond to home care, is reoccurring and seems to be chronic, or is extremely itchy, painful or disfiguring, it’s wise to see a doctor who can help identify its cause and develop an effective treatment plan.

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.

Reference

Calm Inflammation
Topical corticosteroids (or just steroids) treat rashes by easing redness and reducing inflammation and itching so that the skin can begin to heal. Cortizone-10 Intensive Healing Formula Maximum Strength Anti-Itch Crème is formulated to offer the highest-strength rash relief without a prescription.
$6.46; walmart.com

Ease the Itch
Benadryl Itch Stopping Cream formulated with 2% diphenhydramine hydrochloride topical analgesic and 0.1% zinc acetate skin protectant can soothe irritated itchy skin. The formula is a popular over-the-counter favorite to temporarily relieve itching and discomfort.
$4.49; target.com

Reduce Redness
Skin that is red and scaly can leave a person feeling extremely self-conscious. Unsightly eczema and similar conditions can be treated with the Exederm Flare Control Cream for Eczema and Dermatitis. Topical application reduces redness, rashes, itchiness and inflammation.
$9.99; bedbathandbeyond.com

Ice, Baby, Ice
Ice or cold packs can provide soothing, cooling relief for inflamed skin. An antimicrobial wrap like the Arctic Ease Reusable Cold Compression Therapy wrap can be used on-the-go, and works without the mess of ice or the need for refrigeration.
$14.49; jet.com

Natural Comfort
Aloe vera is considered a go-to natural remedy for soothing rashes and skin irritations. Aloe is rich in natural vitamins and minerals that can help assist with healing and soothing the skin. Fresh aloe vera is best, but an aloe vera gel from the health food store can work equally as well. Jason Aloe Vera Gel:
$8.63 amazon.com

Shopping Guide to Rash Treatments
The Everything Guide to the Autoimmune Diet
Author: Jeffrey McCombs
Publisher: Simon and Schuster Digital Sales

In *The Everything Guide to the Autoimmune Diet*, readers will learn what foods can help improve their conditions and how to avoid the ones that exacerbate problems. This gluten-free diet focuses on healing the gut, boosting immunity and restoring wellness. Included are 150 nutritious recipes such as turkey breakfast sausages, farmers' egg casserole, breakfast fried rice, coconut cream of broccoli soup, harvest chicken soup, Mediterranean turkey burger, herbs de Provence-crusted bison sirloin tip, Ojai ginger-orange salmon, casa blanca chicken skewers, beet and peach salad and pumpkin spice applesauce. The book also features meal plans and a variety of detoxifying juice cleanses to help heal the body naturally.

Hashimoto’s Protocol:
A 90-Day Plan for Reversing Thyroid Symptoms and Getting Your Life Back
Author: Izabella Wentz, PharmD
Publisher: HarperOne

Dr. Izabella Wentz, the author of the *New York Times* bestseller *Hashimoto's Thyroiditis*, returns with a prescription to reverse the symptoms of this serious autoimmune condition that is becoming one of the country’s fastest growing diseases. Diagnosed with Hashimoto’s at 27, Dr. Wentz draws on her own personal experience, as well as her work consulting with thousands of patients, to offer a practical pathway for healing and reversing the autoimmune damage at the root of the disease. The first step is a quick-start two-week detox that includes foods to eat and inflammatory foods to avoid, advice on supplements to support the liver and an adrenal recovery plan. Next, readers create a personalized plan with foods, supplements and other lifestyle interventions tailored to their body’s own unique Hashimoto’s triggers, which they can identify using self-tests included in the book. Also included are original recipes.

Autoimmune Disease Journal: Daily Chronic Illness Journal
Author: JC Grace
Publisher: CreateSpace Independent Publishing Platform

This three-month chronic illness journal allows individuals to record daily experiences and track changes over time. Areas for entry include the daily mood tracker, daily positive and negative tracker, daily symptom tracker, test results record, daily entry area, inspirational bookmarks and journal tags.

New and useful reading

The Complete Anti Inflammatory Diet Cookbook: Stop Auto-Immune Diseases with Anti Inflammatory Herbs — Anti Inflammatory Smoothie, Breakfast, Lunch and Dinner Recipes
Author: Martha Stone
Publisher: Amazon Digital Services

Within the pages of this anti-inflammatory diet cookbook, readers will find 25 recipes that contain anti-inflammatory herbs to help reduce, prevent and eliminate inflammation. Each recipe has been tested to ensure its accuracy and tastiness for maximum effectiveness. In addition, all recipes include the complete time it will take to prepare the dish, a description of the dish, serving size that can easily be adjusted by altering the amount of ingredients, a complete list of ingredients required and step-by-step instructions written in an easy-to-follow format. Also included is a chapter dedicated to anti-inflammatory crockpot recipes.
Download the *IG Living* eBook today—now available for iPad, Nook and Kindle!

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

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

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

— Jenny Gardner

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