DIAGNOSED!
How to Share the News

Steps to Successfully Filing for Disability

Choosing an IG Infusion Environment

Pros and Cons of Ports
Understanding Out-of-Pocket Expenses
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About IG Living

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Dealing with Post-Diagnosis Issues

ACCORDING TO PATIENT surveys conducted by the Immune Deficiency Foundation, it takes 12.4 years, on average, to diagnose a primary immunodeficiency (PI) disease. For autoimmune disorders (ADs), the diagnosis time is improved but still lengthy, according to Internet sources: On average, an AD patient will see six doctors over a period of four years before receiving a diagnosis. After so much time and energy is spent searching for answers, you’d think the road ahead would be clear and well defined. But, in reality, many issues have to be dealt with to best live with these chronic illnesses.

How to share the news of a diagnosis with family and friends can be perhaps one of the most difficult issues. It may be a relief for some that there is a reason behind years of unexplained illness, but explaining the complexities of a PI or AD to others can be challenging. In our article “Telling It All: How to Share the News of Your Diagnosis,” we provide some tools for prioritizing whom to tell and in what order, learning how to empathize with yourself as the person who is most affected, and soliciting the support needed. Patience is of utmost necessity because others will likely ask for repeated clarification, which is why preparing an “elevator pitch” might help them to better understand.

Whether an individual with a chronic illness is able to work becomes a very serious issue, especially since the toll of work can often exacerbate the condition and the expense of the medications can be financially draining. Fortunately, claiming disability is an option for many, which Cynthia Perry learned firsthand. In her article “How to Successfully File for Disability,” she provides tips that greatly increase the odds of avoiding the appeals process.

Post-diagnosis, a great majority of PI and AD patients are treated with intravenous immune globulin (IVIG). Enter the world of infusion environments. While most patients will be treated in the hospital and/or doctor’s office when beginning IVIG therapy, eventually, there will be other choices. As our article “Choosing an Infusion Environment” explains, all infusion settings have positives and negatives, so understanding the types, cost considerations and advantages and disadvantages can help in the decision process.

Many PI patients who remain on IVIG long-term may experience difficulty with venous access. When this happens, a port can sometimes be an alternative to uncomfortable, repeated needle sticks. But, ports subject patients to substantial risk, mainly due to the increased susceptibility for infection. Therefore, while it’s ultimately an individual decision, our article titled “The Pros and Cons of Ports” can help patients better understand how ports work and their benefits and drawbacks.

Finally, it’s no secret that IG therapy is a treatment that is often subject to high out-of-pocket copayments and co-insurance. While the Affordable Care Act placed maximums on these expenses, it hasn’t solved the issue of specialty tiers, as explained in our patient advocate’s column, Abbie’s Corner. Fortunately, legislators and patient groups are calling for the current tier policy to be changed.

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

Ronale Tucker Rhodes, MS
Understanding Out-of-Pocket Expenses

By Abbie Cornett

SINCE THE BEGINNING of the year, I’ve been asked many times about an issue that greatly affects many patients’ access to care: out-of-pocket expenses. In many instances, patients are unable to pay these expenses because they are so high. Understanding why out-of-pocket expenses have increased significantly requires a closer look at how the current system evolved.

In 2014, the Affordable Care Act (ACA) required all healthcare plans that were not grandfathered to implement an out-of-pocket expenses maximum. For the plan year beginning Jan. 1, 2014, these maximums were set at $6,350 for individuals and $12,700 for families. Going forward, out-of-pocket maximums increase every year based on increases in the average per capita premium for health insurance coverage. In 2015, the maximums increased to $6,600 per individual and $13,200 for families.

Under the ACA, people who previously were uninsured can now have insurance coverage. However, the law has not necessarily helped patients with chronic or expensive diseases gain access to care. This is because many insurance plans place most medications prescribed for the chronically ill in specialty tiers. These tiers require patients pay high out-of-pocket costs, with co-insurance rates as high as 30 percent to 40 percent.

And, while the ACA places a maximum on the out-of-pocket expenses a patient must pay, that maximum is still a very heavy burden for most chronically ill people. Further, many patients don’t fully understand their out-of-pocket obligation until it’s too late, when they realize they are unable to afford their medication under their chosen healthcare plan. Recent research shows that many patients either postpone getting their medications or forgo treatment altogether because they are unable to pay.

High out-of-pocket expenses affect the chronically ill more than the rest of the population — an issue that has come to national attention in a number of ways. Some states, including New York and Maryland, already have laws that either set caps or monthly limits on out-of-pocket expenses. Other states are also beginning to address this issue. For instance, in Oregon, there are currently two such bills pending. One would cap copays at $100 per 30-day supply. Another would place a cap of $200 for specialty medicines. Similar bills have also been introduced in the Illinois Legislature.

Many chronically ill patients feel they are discriminated against by the practice of placing medication in specialty tiers, which creates a situation in which patients don’t have affordable access to medication. In fact, in 2014, two leading national organizations — the National Health Law Program and the AIDS Institute — filed a complaint with the Office of Civil Rights at the U.S. Department of Health and Human Services (HHS) requesting officials take action against four insurers in Florida to end discrimination against patients. In addition, 300 patient groups submitted protest letters to HHS to complain of similarly discriminatory practices against patients.

Hopefully, with all the attention being brought to this issue by legislators and patient groups, the current policy will be changed to be more equitable for the chronically ill. As a patient advocate and a patient, I understand what a hurdle high out-of-pocket expenses can be to accessing treatment. No patient should have to endure overwhelming financial hardship to receive their medication.

ABBIE CORNETT is the patient advocate for IG Living magazine.

Out-of-Pocket Maximum/Limit

- The most you pay during a policy period (usually one year) before your health insurance or plan starts to pay 100 percent for covered essential health benefits.

- The limit must include deductibles, co-insurance, copayments or similar charges and any other expenditure required of an individual that is a qualified medical expense for the essential health benefits.

- The limit does not have to count premiums, balance billing amounts for non-network providers and other out-of-network cost-sharing, or spending for non-essential health benefits.

- The maximum out-of-pocket cost limit for any individual Marketplace plan for 2015 can be no more than $6,600 for an individual plan and $13,200 for a family plan.

How do you stay active while living with chronic illness?

Before [I was diagnosed with] myasthenia gravis (MG), I worked out four to five days a week at the gym. I loved it. After MG, I rarely get any exercise. I’m going to try to start a short walk each day, but living in Las Vegas, the walking season is short due to excessive heat.

— Laurie L.J.

I work out four days a week. I feel so much better when I do.

— Taylor W.

When I am stable, I do 10 to 15 hours of high-intensity dancing a week plus a little stretching/weight training. If I am battling an infection or illness, I unfortunately don’t get much in at all because I feel too weak to do it.

— Jennifer W.

How much do you know about chronic inflammatory demyelinating polyneuropathy (CIDP)?

I have lived with CIDP for almost four years. I get intravenous immune globulin (IVIG) every three weeks spread over two days. It’s a tough disease. Hopefully, someday, they will find a cure.

— Lynn M.

I’m eager to learn anything and everything, especially any possible cures/treatments that may be coming down the pipeline. I subscribe to and read your magazine, and I truly appreciate the information provided! Since it’s so rare, I do feel “alone” when it comes to talking to others about it. I am very fortunate to have family and friend support.

— Karen E.S.

Do you remember to schedule routine annual health screenings?

I haven’t had a physical or anything like that since I was in the ninth grade.

— Taylor W.

My primary care physician (PCP) says I see her enough that I don’t need this. I disagree with her on many issues, but it’s hard to find a good PCP to see me. I just hope with all the doctors I see that nothing is missed.

— Deb K.

Seriously? If it weren’t for all my doctor appointments, I wouldn’t have a social life. No need for reminders, but thanks.

— Melanie S.
AS WE DISCUSSED in the last issue, there are two clinical findings (infections and autoimmune manifestations) and three laboratory findings (serum immunoglobulin [IgG] measurements, pre-/post-immunization antibody responses and B lymphocyte count) that help to diagnose an antibody deficiency. Based on these findings, many names are used to define specific subsets of antibody deficiencies. In this discussion, we will focus on common variable immunodeficiency (CVID), which is variable in presentation, symptoms and laboratory findings (see the table).

With CVID, the frequency and severity of infections can range from mild (infrequent and easy to treat) to severe (frequent, recurrent, difficult to treat and resulting in additional complications). Autoimmune symptoms may be absent in some patients, or they may be the most difficult part of the disease.

By definition, the serum IgG level should be low for age. In some patients, the level may be near the lower limit of the normal range for age; in others, the level may be undetectable. A second serum antibody, typically IgA, is also expected to be low. While the IgA levels can vary from near normal, most frequently, they are low to undetectable. Serum IgM can be puzzling. In some patients, it may be the second serum IgG to be low, but in most, it is not. Indeed, in many patients, especially earlier in their disease progression, the serum IgM level is normal and, not infrequently, elevated — possibly because the immune system is trying to compensate for the loss of IgG and IgA.

The diagnosis of an antibody deficiency requires a functional abnormality. The current practice is to test the immune system’s response to the pneumococcal polysaccharide vaccine to determine if it can appropriately make antibodies. Pre-immunization serum is obtained prior to vaccination and post-immunization serum is obtained typically about four weeks after vaccination. Antibody titers to specific pneumococcal antigens are then measured and compared. The current definition for normal is that more than 50 percent should have values greater than or equal to 1.3 µg/mL with greater than or equal to a two-times increase between the pre- and post-immunization sera in children up to 6 years of age; for those older than 6 years, there should be greater than 70 percent response rate. In the table, the left end of the moderate response indicates response rates near to but poorer than 50 percent and 70 percent, respectively, whereas the right end of

### Spectrum of Features of CVID

<table>
<thead>
<tr>
<th>Feature</th>
<th>Normal (lack of symptoms or features)</th>
<th>Mild</th>
<th>Mild to Moderate</th>
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<th>Severe</th>
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<td>Infection Pattern</td>
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<td>Autoimmunity Pattern</td>
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<td>Pre-/Post-Immunization Pneumococcal Antibody Responses</td>
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<td>Blood B Lymphocyte Patterns</td>
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the severe response indicates zero detection. Once again, this illustrates the variability in CVID. Some patients may have a barely subnormal result, while in others there is no response.

Lastly, the number and distributions of specific B lymphocyte subpopulations can be essentially normal or they can be undetectable.

In summary, the laboratory parameters for patients with CVID may range from having serum IgG levels just below the normal ranges for age, along with pneumococcal antibody responses just below normal and essentially normal B lymphocyte subpopulation counts, to having undetectable serum IgG levels, an absence of detectable anti-pneumococcal antibody titers, and an absence of B lymphocytes. What’s more, any intervening pattern may be present, and the patterns do not have to parallel. For example, low IgG levels do not have to have equally as low pneumococcal titers and equally as low B lymphocytes. Some patients may have barely low IgG levels with essentially absent pneumococcal responses and only moderately low B lymphocytes, and vice versa.

The patterns of infections and autoimmunity also vary among individuals. However, generally, the patterns of disease and laboratory values do not vary greatly within an individual, although over time things can worsen. Usually, with appropriate treatment, the deleterious processes are mitigated.

Another, but not final, issue of variability is that the clinical features may not parallel the laboratory findings. There are patients who present with undetectable serum IgG levels, undetectable pneumococcal responses and undetectable B lymphocytes, which may have been more or less successfully treated with courses of antibiotics. (I am not advocating treating CVID with antibiotics; to the contrary, immune globulin replacement therapy is the indicated treatment.) And, then, there are patients who present with horrible autoimmunity and/or infectious complications, whose serum IgG and pneumococcal titer values may hover in the near-normal range, and B lymphocytes may actually be in the normal range.

In subsequent issues, we will focus on the features of specific antibody deficiency and how they contrast with the features of CVID.

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.
There are three things that come to mind for me when reviewing your question. First, has anyone tested you for IgA deficiency and/or IgA antibodies? If so, have you tried the low IgA product made by Baxter (Gammagard SD LIGA)? If you have tried Gammagard SD LIGA and did not tolerate it, then there are two other possible options. The first is to see whether your physician would feel safe trying a desensitization protocol. This would entail stimulating your immune system with gradually increasing doses of the IG in order to modify or stop the allergic response. There is some risk with this approach, so it would most likely be attempted in a very controlled setting. The second is for you to try subcutaneous administration of IG (SCIG) if you haven’t already tried it. There are two clinical articles that discuss the use of SCIG for patients with IgA deficiency or IgA antibodies. The first is titled “Substitution Therapy in Immunodeficient Patients with Anti-IgA Antibodies or Severe Adverse Reactions to Previous Immunoglobulin Therapy” published in the June 2003 (Vol. 61, No. 6) issue of The Netherlands Journal of Medicine. The second is titled “Anti-IgA Antibodies in Common Variable Immunodeficiency (CVID): Diagnostic Workup and Therapeutic Strategy” published in 2007 in Clinical Immunology (Vol. 122, 156–162). These might be of interest to you and your physician.
Introducing Flexible Dosing with Hizentra

With the freedom and flexibility of Hizentra, patients can work with their prescriber to create an individualized treatment plan that best fits their lifestyle.

“I choose daily.”  “I choose weekly.”  “I choose every 10 days.”  “I choose every 2 weeks.”

Melaine, Voice2Voice Advocate
Carl, Voice2Voice Advocate
Ben & Traci, Patient & Voice2Voice Advocate
Tim, Voice2Voice Advocate


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.

*Voice2Voice advocates are not healthcare professionals or medical experts. For medical questions, patients should contact their physicians. Voice2Voice advocates are compensated by CSL Behring LLC for their time and/or expenses.

Please see additional Important Safety Information on reverse side and brief summary of full prescribing information for Hizentra, including boxed warning, on adjacent page.
Important Safety Information (continued)

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 symptoms that could indicate hemolysis (destruction of red blood cells), and other potentially serious reactions that have been seen with Ig treatment, including aseptic meningitis syndrome (brain swelling); kidney problems; and transfusion-related acute lung injury.

The most common drug-related adverse reactions in the clinical trial for Hizentra were swelling, pain, redness, heat or itching at the site of injection; headache; back pain; diarrhea; tiredness; cough; rash; itching; nausea and vomiting.

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

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, including boxed warning, on adjacent page.

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

Hizentra®, Immune Globulin Subcutaneous (Human), 20% Liquid
Initial U.S. Approval: 2010

BRIEF SUMMARY OF PRESCRIBING INFORMATION

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

WARNING: THROMBOSIS

See full prescribing information for complete boxed warning.

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

- For patients at risk of thrombosis, administer Hizentra at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.

-----------------------------------INDICATIONS AND USAGE-----------------------------------

Hizentra is 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.

-----------------------------DOSAGE AND ADMINISTRATION---------------------------------

For subcutaneous infusion only. Do not inject into a blood vessel.

Administer at regular intervals from daily up to every two weeks (biweekly).

Dosage

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) \( \times \) 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/IGSC 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.

<table>
<thead>
<tr>
<th>Infusion Number</th>
<th>1st</th>
<th>2nd to 4th</th>
<th>5th</th>
<th>6th and above</th>
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<tr>
<td>Volume (mL/site)</td>
<td>( \leq 15 )</td>
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<tr>
<td>Rate (mL/hr/site)</td>
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* As tolerated

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-------------------------------

- Pregnancy: No human or animal data. Use only if clearly needed.
- Pediatric: No specific dose requirements are necessary to achieve the desired serum IgG levels.

Based on January 2015 revision

0.2 g per mL (20%) protein solution for subcutaneous injection

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

- IgA-deficient patients with anti-IgA antibodies are at greater risk of severe hyperviscosity and anaphylactic reactions.
- Thrombosis may occur following treatment with immune globulin products, including Hizentra.
- Aseptic meningitis syndrome has been reported with IGIV or IGSC treatment.
- Monitor renal function, including blood urea nitrogen, serum creatinine, and urine output in patients at risk of acute renal failure.
- Monitor for clinical signs and symptoms of hemolysis.
- Monitor for pulmonary adverse reactions (transfusion-related acute lung injury [TRALI])
- Hizentra is made from human plasma and may contain infectious agents, e.g., viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.
**Guillain-Barré Syndrome and the Influenza Vaccine: To Vaccinate or Not to Vaccinate?**

By Elissa Ritt, MAS

**GUILLAIN-BARRÉ SYNDROME** (GBS) is considered an aggregate of syndromes that manifest as an autoimmune inflammatory polyneuropathy with an acute onset. Patients generally present with the weakness and diminished reflexes associated with a demyelinating polyneuropathy that may quickly progress over a period of hours or days. GBS often affects respiratory muscles, and the shortness of breath, difficulty swallowing and distressed breathing that can result require respiratory support in approximately one-third of patients. While 80 percent of patients recover the ability to walk on their own after six months, and 60 percent completely recover their strength in one year, approximately 20 percent of patients do not recover their strength and live with severe disability. Even when treated with immunotherapy such as plasma exchange or intravenous immune globulin, up to 5 percent die from the disease.

Many GBS patients recall a brief,
seemingly innocuous gastrointestinal or respiratory infection prior to the acute onset of neuropathy symptoms. These infections are often referred to as “antecedent infections” and are thought to trigger the immune response that is responsible for the development of GBS. However, to complicate the matter further, there is evidence that GBS can also be triggered by the vaccinations, specifically the seasonal influenza vaccine, that can protect against these infections. Which is correct? Does the influenza vaccine cause GBS, or does it protect patients from its onset by preventing antecedent infections? A review of the literature shows mixed results, but recent research demonstrates that the seasonal influenza vaccine might not be such a culprit.

A Look at the Evidence

The relationship between the influenza vaccine and GBS was first questioned when an inordinate number of patients who had received a swine influenza vaccine in 1976 developed GBS. As a result, the vaccination program was suspended, and a study assessed the national incidence of GBS from 1976 to 1977, which determined that many of the 1,098 GBS cases were indeed related to the vaccination. In addition, the study found an elevated risk for GBS in every adult age group among those who had been vaccinated.

Since then, subsequent studies linking the influenza vaccine to GBS have shown conflicting results. In 1994, suspicions were aroused once again when the number of GBS cases during the 1993-1994 influenza season rose to 74, up from 37 during the 1992-1993 season. Unlike the 1976 study, however, this study failed to show a significant increase in GBS cases among vaccinated patients.

In 2009, investigators from the United Kingdom used their General Practice Research Database to draw conclusions regarding the relationship between those vaccinated with the influenza vaccine between 1990 and 2005 and the onset of GBS. While they found no increased risk of GBS among those who were vaccinated, they found a greatly increased risk of GBS among those who reported an influenza-like illness. This increase was found to be most pronounced in the first two months following the report of an influenza-like illness, which is consistent with the pattern of antecedent infections previously described. GBS is often noted for its seasonality, with an increased incidence during the influenza season. What this study didn’t show is that the influenza vaccine actually protected patients from GBS onset.

Recently, investigators used simulation models to study the effect of the seasonal influenza vaccine and the influenza virus itself on the incidence of GBS. Published in Emerging Infectious Diseases in February, the study showed a very slight increase in GBS incidence in the case of low influenza rates coupled with low vaccine effectiveness; however, most simulated scenarios showed no increased risk of acquiring GBS after influenza vaccination. Furthermore, this study actually showed a small reduction in GBS cases following influenza vaccination under the typical, real-world conditions of average influenza rates and average vaccine effectiveness. In other words, the influenza vaccine had a slight protective effect against the development of GBS.

Does the influenza vaccine cause GBS, or does it protect patients from its onset by preventing antecedent infections?

What to Do?

With evidence both supporting and refuting the link between the seasonal influenza vaccine, the influenza virus and GBS, what is an individual who is considering the influenza vaccine to do? GBS can be a devastating condition, but so can influenza, especially for the elderly and other populations considered high-risk. Evidence-based reviews of the literature can be helpful, but each individual should make his or her own healthcare decisions, and risk-benefit scenarios must be taken into consideration.

ELISSA RITT, MAS, is medical science liaison for NuFACTOR Specialty Pharmacy.

References

The Immune Deficiency Foundation has launched LivingwithCGD.org, a new website for those living with chronic granulomatous disease (CGD). The site will provide the latest news and information regarding diagnosis and treatment and serve as a platform to connect individuals and families living with CGD. The site is being launched at a pivotal time for CGD with more treatment options available today enabling patients to lead healthier and more active lives.

CGD is a rare type of primary immunodeficiency disease in which one group of the body’s white blood cells, called neutrophils, fails to make the hydrogen peroxide, bleach and other chemicals needed to fight bacterial and fungal infections. It affects an estimated 1,200 people in the U.S. and approximately 25,000 people worldwide.

Legislation
Patients’ Access to Treatments Act Is Reintroduced

On March 25, Representatives David McKinley (R-WV) and Lois Capps (D-CA) reintroduced the Patients’ Access to Treatments Act (PATA), H.R. 1600, in the U.S. House of Representatives. The Act, if passed, will provide thousands of people with primary immunodeficiency diseases access to life-saving treatments by lowering patient out-of-pocket costs in commercial health plans. PATA would require commercial health insurers to impose the same copayment obligations for specialty drugs as they do for other medications.

“The Immune Deficiency Foundation enthusiastically supports the Patients’ Access to Treatments Act to ensure our patients with primary immunodeficiency diseases receive necessary and appropriate treatment,” said Marcia Boyle, president and founder of the Immune Deficiency Foundation (IDF). “IDF believes that all patients, regardless of income, should have access to life-saving medications. By enacting PATA into law, people with primary immunodeficiencies and other chronic or rare diseases will have access to their full range of treatments without the worry of high out-of-pocket costs.”

Medicines
ADMA Biologics’ IVIG RI-002 Receives Positive Phase III Results

ADMA Biologics has received positive results on the primary and secondary endpoint evaluations from the Phase III trial for its intravenous immune globulin (IVIG) product RI-002 to treat primary immunodeficiency disease (PI). The multi-site study treated 59 patients diagnosed with PI with RI-002, which resulted in a total of 93 days (1.66 days per patient per year) lost from work or school due to infection; only one hospitalization due to an infection of only five days; and IgG trough levels above those required by the U.S. Food and Drug Administration (FDA) for IVIG products. In addition, there was a marked increase in all of the measured specific anti-pathogen antibodies in PI subjects, with the greatest increase (5.3-fold) seen in the level of neutralizing antibody titers to respiratory syncytial virus (RSV). The safety profile was comparable to that of other IG products.

“These Phase III results suggest that RI-002 and its unique antibody profile containing standardized high levels of anti-RSV neutralizing antibodies may demonstrate an improvement in certain clinical outcomes,” said James Mond, MD, PhD, ADMA Biologics’ chief medical and scientific officer. “We believe that the data from the primary and secondary outcomes analyses will enable ADMA to differentiate RI-002 from other IVIG products and offer clinicians and patients a promising alternative to current therapies for the immune deficient population.” ADMA is currently assembling its biologics license application for planned submission to FDA during the first half of 2015.

Websites
IDF Launches Website for Chronic Granulomatous Disease

The Immune Deficiency Foundation launches LivingwithCGD.org, a new website for those living with chronic granulomatous disease (CGD). The site will provide the latest news and information regarding diagnosis and treatment and serve as a platform to connect individuals and families living with CGD. The site is being launched at a pivotal time for CGD with more treatment options available today enabling patients to lead healthier and more active lives.

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Resource

IDF Publishes New Book for PI Children

The Immune Deficiency Foundation (IDF) has published *A Zebra Tale*, an illustrated storybook written for children of all ages with primary immunodeficiency diseases (PIs) and their families. Intended to provide hope and encouragement, the book chronicles the journey of a young horse who faces recurrent infections, struggles to understand why he gets sick often and ultimately finds contentment and acceptance when he is diagnosed with a PI, discovering that he is a zebra.

The identity of a zebra for PI patients is based on an old saying in medical school that doctors learn: “When you hear hoof beats, think horses, not zebras.” The saying teaches doctors to focus on the likeliest possibilities when making a diagnosis, not the unusual ones like PIs. “Members of the primary immunodeficiency community have long awaited the publication of *A Zebra Tale*,” explains Marcia Boyle, IDF president and founder. “With a touching story created by a mother of a child with primary immunodeficiency and beautiful illustrations developed by a patient, this storybook was truly a labor of love. It will be a meaningful resource for zebras of all ages, helping them to cope with the emotional impact of living with primary immunodeficiency and to understand that they are not alone.”

The book can be downloaded or ordered in hard copy at www.primaryimmune.org/idf-publications.

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Take Charge of Your Health with IDF ePHR

The Immune Deficiency Foundation (IDF) knows that keeping track of medical records for patients with primary immunodeficiency (PI) can be challenging. Therefore, we developed IDF ePHR, the electronic personal health record designed for those who live with PI.

**IDF ePHR offers:**

- Easy-to-use tools to help you improve your health and organize your life.
- A safe, secure and private system to store your medical information at no cost to you.
- Convenient access from anywhere through your computer, smartphone or tablet.
- Simple methods to track symptoms, conditions, and infections that help identify health patterns.
- Reports to take to your healthcare providers, enhancing the information you share with them.

**Take charge of your health and create an account today!**

www.primaryimmune.org/takecharge

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For more information, e-mail info@idfephr.org or call 800-296-4433.
Autoimmune Corner

Research

Breakthrough Tool Developed for MS

Researchers at Case Western Reserve University School of Medicine have developed a first-of-its-kind imaging tool to examine myelin damage in multiple sclerosis (MS). The new molecular marker, MeDAS, offers the first noninvasive visualization of myelin integrity of the entire spinal cord at the same time. Currently, a long lag exists between the onset of disease, physical symptoms and diagnosis via behavioral testing and magnetic resonance imaging. It is hoped that the tool will help physicians diagnose patients earlier, monitor the disease’s progression and evaluate therapy efficacy. “While MS originates in the immune system, the damage occurs to the myelin structure of the central nervous system. Our discovery brings new hope to clinicians who may be able to make an accurate diagnosis and prognosis in as little as a few hours compared to months or even years,” said Yanming Wang, PhD, senior author of the study and associate professor of radiology at Case Western. “Because of its shape and size, it is particularly difficult to directly detect myelin damage in the spinal cord; this is the first time we have been able to image its function at the molecular level.” Created by Wang’s laboratory, the MeDAS molecular probe works like a homing device. Injected into the body intravenously, it is programmed to seek out and bind only to myelin in the central nervous system. A positron-emitting radioisotope label on the molecule allows a PET scanner to detect the targets and quantify their intensity and location. The data can then be reconstructed as an image.

Research

Study Suggests Six Autoimmune Disorders Caused by Weak Common Genetic Variants

In the largest sequencing study of human disease to date, researchers investigated the genetic basis of six autoimmune diseases. The exact cause of these diseases — autoimmune thyroid disease, celiac disease, Crohn’s disease, psoriasis, multiple sclerosis and type 1 diabetes — is unknown, but it is believed to be a complex combination of genetic and environmental factors. In each disease, only a proportion of the heritability is explained by the identified genetic variants, and the techniques used to date have generally identified common variants of weak effect.

In this study, which used high-throughput sequencing techniques, researchers sought to identify new variants, including rare and potentially high-risk ones, in 25 previously identified risk genes in a sample of nearly 42,000 individuals. Whereas the rare-variant synthetic genome-wide association hypothesis suggests that a small number of rare variants in risk genes are likely to be a major cause of the heritability of these conditions, this study found that the genetic risk of these diseases more likely involves a complex combination of hundreds of weak-effect variants that are each common in the population. Researchers estimate that rare variants in these risk genes account for only around 3 percent of the heritability of these conditions that can be explained by common variants.

“These results suggest that risk for these autoimmune diseases is not due to a few high-risk genetic variants, but seems, rather, due to a random selection from many common genetic variants which each have a weak effect,” said David van Heel, lead author of the study. “For each disease, there are probably hundreds such variants, and the genetic risk is likely to come from inheriting a large number of these variants from both parents.” If this is the case, it may never be possible to accurately predict an individual’s genetic risk of these common autoimmune diseases. “However, the results do provide important information about the biological basis of these conditions and the pathways involved, which could lead to the identification of new drug targets,” adds van Heel.

The study was published in the journal Nature.
9th Annual Neuropathy Action Foundation Awareness Day Is June 18th

The 9th Annual Neuropathy Action Awareness Day is an opportunity for patients to interact with other patients, providers and exhibitors, as well as to learn about neuropathy and how to cope with it, policy issues and patient advocacy. The day consists of educational sessions, an exhibit area, a sit-down luncheon and a silent auction. Educational sessions include:

- Treatable Neuropathies: CIDP and MMN — A Neurologist’s Approach to Diagnosis and Treatment
- Understanding a Neurologist’s Approach to the Diagnosis and Treatment of Peripheral Neuropathy
- East-West Medicine Approach to Neuropathy
- Neuropathic Pain and Pain Treatments
- Occupational Therapy: Living a Full and Complete Life
- Unusual Neuropathies
- Food as Medicine: Improve Health, Increase Vitality and Live Longer!

Other highlights include an update on the Neuropathy Action Foundation (NAF) by founder and president Dominic Spatafora, the “You Are Our Hero Award” and celebrity and elected officials.

The event is scheduled for June 18th at the Intercontinental Hotel in Los Angeles, from 9:30 a.m. to 4:00 p.m. Individuals who are unable to attend the event in person can participate via video livestreaming technology free of charge. A computer and Internet access are required, and individuals will be able to ask speakers questions and receive answers in real time. The day will also be recorded so it can be watched on the NAF website after the event has ended.

The event is free; however, NAF requests a $20 donation from patients and caregivers and $100 from others who attend the luncheon. Register online at www.neuropathyaction.org or call (877) 512-7262.
After a diagnosis, a patient's to-do list is a mile long. You might have to schedule follow-up appointments or procedures, find resources and groups that can offer support, and keep track of all the new medicines you need to take. But that’s not all. You also need to tell the people in your life about your diagnosis, and that might just be the hardest item on the list to check off.
**FOR MANY PEOPLE**  living with a chronic illness, life can be divided into two parts: before diagnosis and after diagnosis. Even when symptoms have been present for months or even years, the point of diagnosis is a significant milestone. It’s as if you’ve been cleaved in half: You now stand squarely in the “after-diagnosis” half of your life, while the “before-diagnosis” life drifts into the distance. And, as that old life is carried away, you may fear it will take many of your relationships with it.

Your immediate family and closest friends will most likely be the first to know about your condition. But how do you break the news? And how do you tell others such as friends with whom you have less contact, classmates, extended family members, congregation members and other community members?

It might be tempting to keep silent in the hope that those who aren’t in the know won’t have an altered sense of who you are. This is a way of shielding others from your after-diagnosis life, as well as preserving those relationships in a past that might feel safer and more secure than your present or future. But reaching out to those you have shared aspects of your life with might be one way to help bring your past and present together. This, in turn, may help you alleviate the feelings of isolation that so many people living with chronic illnesses experience. Reaching out even more by being public with your condition might also be a therapeutic approach to processing your diagnosis and living with your illness.

Below are some tools to consider when sharing your diagnosis with those who are part of your life and with the larger community.

**Stay Centered**

Staying centered works on two levels. The first is to share your diagnosis with others by placing yourself at the center of the process and envisioning working your way out. On a piece of paper, mark yourself at the center. Then draw consecutive rings around you. Each ring represents a different group of people in your life that you want to tell. Loved ones will most likely be in the ring that’s closest to the center. Close friends might be in the next ring. Distant family members might be in a ring that’s farther from the center. Add enough rings to represent everyone you want to talk with about your condition, and make sure their proximity represents how close they are to you. Now you have a guide for working your way out from the center. Depending on your situation, you might have to make some adjustments in terms of the order in which you share your news, but the diagram gives you a basic blueprint to work from.

The second way of staying centered is exactly what it sounds like: As you go through the process of informing other people about your condition, remember that you are at the center of this news. Patricia Anderson, an emerging technologies librarian for the Health Sciences at the University of Michigan, is a patient advocate who is also very public about her own health issues. She points out that many people who have chronic illnesses spend a long time hiding the manifestations of their conditions before they are finally diagnosed correctly. During that time, they might ask themselves how much they can cover up so they don’t lose their jobs or place their relationships at risk. Anderson says a common theme among those with chronic illnesses is: “How far can I fake it?”

Making the transition from faking it to talking openly about your condition is an accomplishment in itself. As you make this shift, remember that your life is the one most affected by the diagnosis, and you are the person who needs support and understanding during this time. Keep this in mind and honor what you need, including how much energy you can give to any one person and what pace you can adopt when informing others about your condition. People will naturally have questions about the news you are sharing. They might even have misconceptions about what you are trying to relate. The act of communicating and clarifying can be exhausting on top of a condition that may already be draining your energy. Think of each act of communication as a sprint. Warm up for it. Don’t go longer than you are able. Honor the signs your body sends that tell you you’ve done all you can do in one sitting or with one person. Even if you haven’t crossed the finish line in a given conversation, know when you need to pause and say, “I think we’ve talked about this enough for now. Let’s come back to it again another day,” or something to that effect.

Another way to stay centered is to make sure you have all the support you need before you embark on telling people about your diagnosis. Solicit the support of those closest to you first. Find support groups that can help, including those in your area and those available online through organizations that serve people with your condition. You might even consider talking with a therapist for a period of time to help you process the diagnosis and the steps you are taking to communicate that diagnosis to others.

**Tell It, and Tell It Again**

Be prepared to tell your story more than once to the same person. Medical conditions can be complicated and difficult to understand. As you are sharing your news with someone, he or she is likely trying to process it on an emotional and intellectual level. There’s so much to take in and contextualize that it’s almost impossible to be the recipient of such news without having follow-up questions or needing further clarification.

Be as patient as possible. Be as understanding as possible. If you can, have information in print or online that discusses your
condition. Make those reference materials available during or shortly after each conversation. If you know of support groups for loved ones dealing with your condition, share that information as well. If you seem to be sharing more information than someone is comfortable with or able to understand, reduce the message to its core. If someone wants to know more and you are comfortable with that level of sharing, feel free to go into more detail. Talking through the diagnosis in depth can help you process it, which can in turn promote your own healing.

In addition, keep in mind that you might not be the only one telling your story. Anderson points out that loved ones and others advocating for your health may also need to communicate information about your illness, and most likely more than once. Like you, they should be prepared to tell your story as well as possible and as often as needed.

Create Your Elevator Pitch

There will be situations in which you and those advocating for your health will need to convey the essence of your diagnosis quickly and succinctly. This is where having an elevator pitch comes in handy. In marketing, an elevator pitch is a short summary used to quickly define a profession, product or service. It’s supposed to be no longer than an elevator ride, hence the name.

For a medical condition, your elevator speech might be: “I have a rare, genetic condition that affects my immune system” or “I have pain all over my body that limits my activities.” Everybody’s elevator speech will be unique, and you can use variations in different settings. For instance, if you choose to share your diagnosis during an interview, you could add a second sentence that speaks to your ability to perform the functions of your job. Melanie, who was diagnosed with common variable immunodeficiency (CVID) in 2010, recommends using words that other people are likely to understand when giving a summary of your condition. Rather than saying she has a “primary immunodeficiency,” she tells people she has a “genetic immune deficiency” because people are more likely to understand the term genetic as opposed to primary. “Telling people my condition is genetic helps them understand that it isn’t contagious,” explains Anderson, “which can be one of the first questions people ask when I tell them I have CVID.” Anderson also chooses her words carefully when crafting her elevator pitches. For her methylenetetrahydrofolate reductase (MTHFR) deficiency, rather than going into the detailed complexities of the condition, she tells people she has a genetic enzyme deficiency that means her body can’t properly get nutrients from green leafy vegetables. (For more on elevator pitches, see Ilana Jacqueline’s Life as a 20-Something column in the April-May 2014 issue of IG Living.)

Choose Your Scope

It’s up to each person to decide how open they want to be about their condition. Some people might want to tell only their immediate family members. Others might want to tell the world. Many will want to aim for something in between. There’s no right or wrong in this regard. Some people who receive life-changing diagnoses go on to become community educators and health advocates. Others keep their diagnosis more private and
share it selectively.

Though you will most likely experience a great deal of love and support during the process of sharing your diagnosis, you should also be prepared for those who don’t understand what you’re going through. The more open you choose to be and the greater the number of people you include in your communications about your condition, the more likely you are to run into a few naysayers. Anderson cautions: “You can’t assume that everyone will just be kind. Some folks will think you’re crazy, or that your diagnosis is wrong, or your doctors are stupid. Sometimes, being public means you have to defend yourself, your treatment choices and your lifestyle.”

Melanie was reticent at first to be open about her condition. Over time, however, she became more comfortable sharing information in the hope that it would help educate others. She now posts about CVID openly on Facebook, and she maintains a blog called CVID Awareness (cvidawareness.org), which she took over anonymously in February and still maintains semi-anonymously. She recognizes that many people with CVID are afraid of the ramifications of revealing it publicly, but she also sees the need to educate the public and be visible to others who also have the condition.

When you consider scope, you might also want to think about whether to discuss your condition with people from the past. Before diagnosis, many people with chronic illnesses spend years being misunderstood or treated as hypochondriacs. Finally having a diagnosis could help those from your past have a better understanding of what they couldn’t make sense of at the time. When Melanie was diagnosed, she chose to reach out to an ex-boyfriend who hadn’t understood why she was always ill. She says he appreciated knowing about the diagnosis, and the conversation provided a sense of closure that had been lacking before. To give another example, if you missed work and people believed you were making excuses for your absences, it might be helpful to get in touch with the supervisor at that position to update him or her about your health and provide context for your missed workdays. This can be especially helpful if you rely on that supervisor for a reference. The downside of reaching out to those from the past, whether they are former friends, partners or employers, is that they may or may not be open to what you are communicating.

Harness Social Media (and Avoid Its Pitfalls)

Social media can be part of your communication strategy, but be careful. The allure of social media sites is that we can broadcast messages to a large group of people. This can be a tempting way to announce a medical condition because it removes the pressure of a one-on-one conversation and requires less energy than more traditional ways of communicating personal news.

Melanie says she especially appreciated Facebook at the time of her diagnosis because it gave her a convenient way to message acquaintances that she had passing or infrequent contact with. This prepared those acquaintances to see her posts on the subject without being confused about their content. Depending on how comfortable you are with social media, Melanie’s approach might work for you. But for family members and close friends, discussing the diagnosis privately is important. You don’t want someone you know well and care for deeply to learn about your medical condition by reading a post on your Facebook timeline. That could be hurtful and confusing, as well as lead to feelings of resentment that linger.

Social media can also be used effectively to continually educate people about your condition. You can link to research articles, news from organizations that specialize in your condition, and other information that, over time, will give those who follow you a deeper understanding of your condition. At the same time, you should consider the potential effect such information could have. If you aren’t careful with privacy settings, for example, your posts could be seen by a current or potential employer, and you might not want that level of sharing. You also can’t assume your friends are seeing all your posts about your condition. Most posts on social media are seen by a fraction of those they are shared with. Don’t be surprised if you post an update and it seems like people missed it. They probably did. If you don’t continue to touch base with people individually about your condition, they might lose touch of where the diagnosis and treatment stand.

Anderson has made a personal commitment to transparency and openness in order to educate and support others. Her “strategy,” she says, is by necessity a combination of “can’t tell a lie, can’t keep a secret, and don’t ask, don’t tell.” Her communications on social media allow her to share with and learn from others, which is part of her healing process. “I’ve noticed my friends who keep their conditions secret tend to suffer the greatest emotional trauma from it,” Anderson says. In her case, being open about her health, online and offline, has led to more support in all areas of her life.

Whatever approach you take and however widely you decide to cast your net, know that sharing your diagnosis is an act of bravery and strength. Soon, you won’t be in it alone. With a considered approach to communicating about your illness with others, you can engage a band of supporters to help you make your way through the process and help you bridge your life before and after your diagnosis.

DANA MARTIN is a writer and editor in the Kansas City area who specializes in science, medicine and health.
How to Successfully File for Disability

The odds of being approved the first time when filing for disability will vastly improve if you plan ahead and follow some simple steps.

By Cynthia Perry

THERE USUALLY ISN’T a single day or symptom that leads a patient with a chronic illness and his or her medical team to decide when work is no longer an option. Instead, you may suffer a gradual decline in your ability to juggle all of life’s demands: family, work and social obligations. At some point, your boss may ask why you have so many doctor appointments, or why you miss so much work. Co-workers may wonder why you are sick so often. Over time, you may find that almost every hour you aren’t at work you spend asleep: work, sleep, repeat. These are all good indicators that filing for disability may be necessary.

Planning for Disability

Follow these steps to plan for disability:

Review private disability coverage. It’s a good idea to hire an employment lawyer to review your company’s disability policy; a reputable lawyer will normally do this for a few hundred dollars, and this is money well spent. A lawyer will be able to tell you about any pre-existing condition clauses, how much coverage you have, cost of living accelerators (rare in private insurance), if and how your benefits would be taxed, whether you have to apply for Social Security disability (SSDI), and what monies will be deducted from your long-term disability payments. He or she can also explain under what circumstances your coverage will end (usually your last day of employment), and what happens if you are fired, laid off or quit. Perhaps most importantly, a lawyer can explain any clauses about “own profession” vs. “any profession” in your policy, and give you some guidance relating to your particular medical condition; however, keep in mind that a lawyer is not a medical professional.

Review SSDI requirements and benefits. Social Security’s website has a wealth of information about eligibility for disability benefits and the application process. There are two ways to speed up the Social Security application process. The first is “compassionate allowance” for people with amyotrophic lateral sclerosis and certain types of cancer; you have to be terminal to qualify this way. The second is to qualify under “List A.” This is a list of conditions so severe that Social Security grants “automatic” disability (assuming you meet the specific criteria under the listings). Study the List A criteria closely, and see if you meet any of them.

You can also find out from Social Security what your benefits would be, and if and how they would be taxed. You can do this online or in person at your Social Security office.

Ask doctors for support. As you meet with your various doctors for other issues, talk to them about what is going on at work. Ask them very directly if they would support your filing for disability. Talk to them about anything that you have that meets the SSDI List A criteria. Ask them if they would complete disability paperwork.

If you do file for disability, list only the doctors who said they would support your disability claim. You are in control of which doctors your private insurance and SSDI contact.

Document everything at work. Document everything that happens at work, just in case you ever need that information. Keep this in your briefcase or purse in case you are ever escorted out of the building with no warning. Also, keep all of your private disability benefits paperwork in your briefcase, car or home for the same reason.

Track symptoms. Track every one of your symptoms every day in a spreadsheet. You will find it enlightening, and perhaps frightening, to see how much your chronic illness is impacting your life. When you file for disability, you will need to provide this information to both private insurance and SSDI.

Filing for Disability

Once you and your doctors decide it is time to file for disability, stop work (either as forced by your employer or as recommended by an employment lawyer); you can’t be working when you file for disability. Make filing for disability your new full-time job.
When filling out your disability forms, your odds of winning your case will be improved by doing the following:

• Get copies of your medical records. If any do not appear to support your disability claim, don’t list that doctor as part of any disability filing.

• Contact your patient advocacy group(s) to see if they provide any free legal advice for disability applications. Any lawyers they have on staff can offer great insight into how to fill out paperwork to win disability benefits for your specific medical condition(s).

• File for SSDI as soon as you file for private insurance. This indicates to your private insurance that you are confident in your ability to win your case.

• Summarize your symptoms from the symptom tracker you have been keeping (e.g., average per month for six months: one shingles flare lasting three days, one urinary tract infection, one migraine).

• Summarize your doctor appointments for the last year (e.g., average of one doctor appointment per week for the last year).

• Provide a prescription summary for the last year from your pharmacy; indicate any side effects the drugs you take cause on the provided forms.

• Submit any peer-reviewed medical journal articles you can find that support your case (e.g., poor prognosis, no known cure, etc.). Provide your specific medical records that support how your case relates to the medical journal articles (symptoms you have as documented in the articles).

• Be sure to explain how dealing with your chronic illness(es) has impacted your life in detail; explain everything you used to be able to accomplish and what your life has been reduced to now by way of comparison. There will be space on forms to do this, and you may be able to do this in personal interviews, too. Have your “elevator speeches” prepared; don’t be afraid to pour out your emotions about how this affects your family and loved ones.

• Finally, request in writing that your private and SSDI applications be reviewed by a specialist that is board-certified in a specialty appropriate for the most important condition you are claiming on your applications.

After you have submitted your applications, check their status about every two weeks (this goes for both private insurance and SSDI). Particularly with SSDI, it is not uncommon to have things fall through the cracks at any stage in the review process. In addition, be sure to follow up with the doctors you have put on your SSDI forms to see if they have been contacted by Social Security and if they provided the requested information to the agency. If any doctors have not been contacted, call your case worker and ask that they be contacted immediately.

The approval process won’t be stress-free and certainly won’t be automatic, even if you have conditions on Social Security’s List A. But by following these steps, you will improve the odds that you are approved in the first review, avoiding the appeal process entirely.

CYNTHIA PERRY is a wife, mother and advocate. She started her career as a technical writer, later transitioning into marketing analytics and strategic planning. Her life and work have taken her many unexpected places at the forefront of medical and genetic research, and she now shares those experiences with others through her writings.
Choosing an Infusion Environment

Patients need to consider a number of factors, including cost, convenience and safety, when determining which environment is best for them.

By Heather Claverie

LAURA ROHE KNOWS a thing or two about infusion options. At age 14, the Omaha, Neb., resident was diagnosed with common variable immunodeficiency. She counts herself as one of the lucky few to receive such a quick diagnosis since, on average, a patient will spend 12 years waiting for one. Now 40, Rohe has spent the vast majority of her life receiving intravenous immune globulin (IVIG) infusions to maintain adequate antibodies to prevent infections. For 22 of those years, she first received IVIG in a hospital, then at home and then in a doctor’s office. Four years ago, she switched over to subcutaneous IG (SCIG) therapy, and she infuses in the comfort of her own home. “I feel like my health is better because I’m getting the steady infusion every week, with the convenience of doing it at home and less side effects,” says Rohe, who is also a registered nurse for Allergy, Asthma and Immunology Associates in Omaha.

Types of Infusion Environments

Home, hospital, outpatient infusion clinic or doctor’s office? Those are the choices facing patients who are treated with IG.

Until the 1980s, patients were required to remain in a hospital setting for infusions. But when expenses became a major factor for insurance companies, they began looking for alternatives to the costlier hospital infusions. That’s the main reason for such significant growth in home-based infusions, says Leslie Vaughan, senior vice president of clinical programs for NuFACTOR Specialty Pharmacy.

Individuals may receive infusions in their homes, an option that, unlike clinical settings, offers convenience, privacy and flexibility. For example, home infusions eliminate the need to drive long distances for treatment for those living in remote locations. In addition, they allow patients to schedule their infusions at their convenience.

Most SCIG patients can receive infusions at home without supervision because the risk of serious adverse reactions is reduced compared with IVIG infusions. However, medical status will play a role in the site-of-care decision. Some physicians prefer the hospital or clinical setting for those who receive IVIG for proximity to medical supervision in the rare case of adverse events such as anaphylactic reaction. In addition, some IVIG patients have a higher risk of thrombosis because the infusion is administered through the vein. That risk is further increased for those with a health history of diabetes, age (65 or older), coronary artery disease, hypertension, hyperviscosity disorder (including multiple myeloma, macroglobulinemia and polycythemia), thrombotic events and peripheral vascular disease.

In clinical settings, doctors and nurses are able to interact with patients on a monthly basis, while supervising and monitoring their health and response to treatment. This can be especially important for patients living with a chronic disease who may be so accustomed to being sick that they may fail to notice red flags signaling possible infections or worsened disease states. With the monthly contact inherent in a clinical setting, a physician or an experienced infusion nurse will be more likely to notice a change in a patient’s health.

Cost Considerations of Environments

Since the price tag for hospital infusions may be higher than infusing at home, many insurance companies are now trying to identify the best clinical and financial site of care, which in many instances is the home setting, says Vaughan. “It’s a matter of economics for insurance companies,” she explains.

Costs can be cut significantly with home-based infusions due to lower contracted reimbursement and fewer ancillary charges.
such as facility fees. Patients who are taught to become independent with self-administration of SCIG have significant reduction in nursing expense for a payer. They receive several training visits, and once independent, are typically monitored via monthly telephone interaction with the pharmacy dispensing their medication. Most patients receiving IVIG require a nurse. In the case of home infusion, industry standard is for the nurse to remain in the home for the duration of the infusion. In an infusion center or physician office setting, nursing cost may be leveraged by having one nurse attend to several infusions concurrently.

Unfortunately, reimbursement may prohibit some patients from being treated at home. Medicare Part B has limited coverage for IG in the home setting. Medicare Part B only covers five primary immune deficiency (PI) ICD-9 codes under the IVIG benefit, and there is no additional payment for a pump or supplies. Nursing services are only covered if a patient is certified as homebound, and a Medicare-certified agency provides the nursing care. For SCIG patients, these five PI codes have a more favorable reimbursement rate than IVIG, with Medicare even covering the cost of the mechanical pump that is needed for SCIG therapy under the durable medical equipment benefit. With SCIG, the patient is taught how to self-administer and typically becomes independent with care.

Patients with diseases other than the five PI diagnoses may receive coverage under Medicare Part D; however, once again, supplies and nursing are still not covered and become the patient’s responsibility.

Advantages and Disadvantages of Environments

The trend toward home-based infusions, like all infusion environment options, comes with both pros and cons. While the site of care is often a personal decision, it is made on the basis of a variety of factors, including convenience, privacy, safety and reimbursement.

For many patients, the home setting is “a more comfortable environment,” Vaughan says. Patients don’t have to travel and are able to receive the treatments in the comfort of their own home with all the privacy that allows. In addition, eliminating the need for the individual to drive after treatments is much safer, since they may receive premedications that cause drowsiness prior to and during the infusions. “Safe driving after infusions should be a consideration of the infusion suite or prescriber,” says Vaughan.

Some patients prefer the privacy of their home to a hospital or infusion clinic, where infusions typically occur in one large room screened off by curtains. On the other hand, others prefer the clinical setting because they enjoy the camaraderie and the ability to connect with others with their same condition. However, in clinical settings, there are more people. And more people translates to more germs — not the best situation for individuals with compromised immune systems.

When Donna Hobson caught a cold from one of the tots at her home-based preschool, she assumed it was just that — a common cold. Yet, after spending more than a month in the hospital and losing 25 pounds, she knew something was seriously wrong. “From then on, it was just a downhill battle; the staph just spread,” she says. Hobson was eventually diagnosed with PI. She initially received IVIG at home with a nurse. She was then trained to do the infusions herself, a process Hobson describes as “kind of scary, when you’re not medically trained.”

Unfortunately, reimbursement may prohibit some patients from being treated at home.

Then the insurance factor fell into place. Insurance was covering the infusions at $3,000 a month. But when she switched over to Medicare, Hobson was told to head to the hospital. Her first infusion bill from the hospital was $8,700 — nearly triple that of the home infusion price tag. “It was all a matter of insurance,” she says. Hobson switched to SCIG and now administers her treatments herself at home. “Sub-q is wonderful because it gives me the freedom to do it at 9 a.m. or noon,” she adds. “I actually did it on Christmas Eve.”

Making the Choice

When choosing an infusion environment, all factors need to be considered — from cost to convenience to safety. All infusion environments have positives and negatives, and only patients and their doctors can decide which is the best for them.

HEATHER CLAVERIE is a contributing writer for IG Living magazine.

References
The Pros and Cons of Ports

Ports can be controversial due to the danger they pose for immune deficient patients, but in some cases, they may be warranted.

By Abbie Cornett

IMMUNE DEFICIENT PATIENTS and others who are treated with immune globulin (IG) receive this therapy in one of two ways: intravenously or subcutaneously. IV infusions can be from temporary peripheral IVs (the type typically performed when the IV catheter is inserted at each visit and removed after use) or permanent central lines.

A Picc line is a type of temporary central line inserted through a peripheral vein and may be used for several days and up to several weeks when properly maintained. Another temporary central line is a larger catheter venous line inserted directly into a large vein under the collarbone or in the neck and placed all the way to the heart. This type of central line and the Picc line can be inserted at the bedside. There are two types of permanent central lines, both of which are surgically implanted. One is a large catheter that exits the skin with the end of the catheter available to access for taking blood or administering IV medications. The other, also surgically placed, is a port.

Traditionally, IG therapy is begun intravenously (IVIG), and if difficulty is experienced with accessing the vein, subcutaneous IG (SCIG) infusions are often recommended. More recently, though, immunologists recommend beginning SCIG from the onset of therapy. In some instances, instead of SCIG, there is an option of having a port installed, which can be controversial. Patients considering a port should be familiar with its advantages and downsides.

What Is a Port?

A port is a small device that is surgically implanted under the skin of a patient. The port has a “hollow space inside that is sealed by a soft top,” which is connected to a small catheter that is inserted inside a vein leading to the heart.1 There are many different types of ports, so a patient’s physician will suggest the appropriate one.

Placement of a Port

The port is usually placed on either side of the upper chest below the collarbone. In general, the preferred veins for central access are the right internal jugular, left internal jugular, right subclavian and left subclavian — in that order.2 However, the doctor will determine where the best placement of the port is depending on what is best suited for a patient’s treatment.

How Does a Port Work?

A port works like an IV, but instead of inserting a needle into a vein, a special needle is inserted through the skin into the port so medications and fluids can be given. A port can also be used to draw blood samples and may reduce the time needed to infuse some medications.
Advantages of a Port

Ports are popular with patients for many reasons. Many times, a patient’s peripheral veins have been damaged due to repeated needle sticks and the medications that they receive. A port takes away the need for multiple sticks and can greatly reduce the fear of treatment, especially in children.

Diane Galbavy is an example of a patient whose needle anxiety was affecting the quality of her treatments. Diane was diagnosed with leukemia and a primary immune deficiency in June 2013. When she began her treatment, she was unaware of what a port was; she just knew she hated the repeated needle sticks. At the end of the first year of treatment, her anxiety level had reached a point where she just couldn’t imagine going through the stress of getting an IV every 28 days for the rest of her life. Her physician agreed that she should have a port installed. Since getting her port, she doesn’t dread her treatment and feels more positive about her condition. She describes her port as “a beautiful present in a little box.”

The Disadvantages of a Port

While ports may be looked upon favorably by many patients because of easy venous access and comfort, they do not come without substantial risks. According to pediatric immunologist Terry Harville, “A port can be a double-edged sword for patients with a primary immune deficiency.” He believes this question should be asked: “Is a port for convenience or necessity?” Dr. Harville doesn’t recommend a port for his immune deficient patients unless they have no other venous access or they have a co-diagnosis that requires them to receive other forms of IV medication on a regular basis. If there is no co-diagnosis, he recommends they switch from IVIG infusions to SCIG infusions.

Indeed, many physicians believe ports represent a significant concern for immune deficient patients. The American Academy of Allergy Asthma and Immunology’s practice guidelines state: “The placement of permanent central venous access solely for the purpose of IVIG administration should be discouraged. Permanent central venous catheters may be associated with thrombotic and infectious complications.”

A port provides a direct conduit for organisms into the bloodstream of a patient, which creates the risk of a serious infection. To reduce the risk of infection, sterile techniques must be used when accessing the port. And, this can be a problem because not all medical personnel have been trained in the proper method of accessing ports. Should an immune deficient patient show any sign of infection such as fever after a port has been accessed, Dr. Harville recommends performing blood cultures from the port and from a different vein and starting IV antibiotics through the port. This would typically require hospitalization until the culture information can be sorted out.

Besides the risk of infection, ports have other disadvantages. They require surgery to place in the vein, and they can cause considerable scarring particularly if they have to be replaced or become infected. Further, the vein into which the port is placed is "sacrificed" in order to use it. This means that if the port is ever removed from that vein, the vein can no longer be used again. As noted above, there are a limited number of veins suitable for port placement, and with each port placement, there will be one less available.

While ports don’t interfere with normal activity, if a patient plays contact sports that could result in the port being hit, padding over the site may be recommended.

An Individual Decision

While the placement of a port in immune deficient patients raises many concerns, there are many patients like Diane who find them a wonderful solution. Ports significantly reduce needle anxiety, particularly for children, and provide easier administration of medication when venous access is compromised. In the end, however, the decision to have a port installed is one that must be discussed with a physician who can help a patient decide the best course of treatment.

ABBIE CORNETT is the patient advocate for IG Living magazine.

Source
A form of severe combined immunodeficiency, DGS is likely underdiagnosed due to the variability in its characteristics and symptoms, and researchers are still trying to unravel the mystery of why it occurs.

By Ronale Tucker Rhodes, MS

SOME 50 YEARS AGO, Dr. Angelo DiGeorge, an endocrinologist, observed that a subset of patients had similar clinical features, including hypoparathyroidism, an underactive parathyroid gland that results in hypocalcem ia (low blood calcium levels), an underdeveloped or absent thymus that results in problems with the immune system, conotruncal heart defects and cleft lip and/or palate. The disorder was coined “DiGeorge syndrome,” or DGS, until the 1970s, when a speech pathologist named Robert Shprintzen, PhD, described a group of patients with similar clinical features and coined the term velo-cardio-facial syndrome (VCFS); others also referred to it as Shprintzen syndrome. Interestingly enough, other children with similar clinical features were diagnosed with the autosomal dominant form of Otipz G/BBB syndrome and Cayler cardio-facial syndrome. It wasn’t until technology was developed in the 1980s that identified the underlying chromosome, 22q11, in over 90 percent of these patients. Thus, it was discovered that different groupings of features were described as separate conditions even though they were part of a single syndrome — 22q11 deletion syndrome — with many possible signs and symptoms. Today, many physicians refer to DGS as 22q11 deletion because it describes the underlying chromosome problem or as VCFS because it describes the main body systems involved.

What Is DGS?

DGS is one of 11 forms of severe combined immunodeficiency (SCID) classifications. It is a common syndrome, occurring in an estimated one in 4,000 to 6,395 newborns; however, researchers and doctors suspect it is more common and is undiagnosed due to its variable features. For instance, DGS may not be identified in people with mild symptoms, and it may be mistaken for other disorders with overlapping features.

DGS is caused by abnormal cell and tissue development during fetal growth. Individuals with DGS are susceptible to infections due to poor T cell production and function and often have altered facial characteristics, abnormal gland development or defects in organs such as the heart. It is a lifelong condition that mostly affects infants and children, who differ in the organs and tissues affected, as well as in the severity of the disease.
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ANSWERS AND ADVOCATES

Tony
Golf enthusiast
living with PI
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What Causes DGS?

In humans, DNA is organized as 23 pairs of chromosomes. One pair, the sex chromosomes, consists of either two X chromosomes (XX), resulting in a girl, or one X and one Y chromosome (XY), resulting in a boy. The other 22 pairs of chromosomes, referred to as autosomes, are numbered 1 through 22. Each chromosome has two or three parts: a short arm (which is not present in some), a central portion and a long arm. The long arm is called by the number of the autosome and “q.” Therefore, the long arm of chromosome 22 is called 22q. The arms also have sections that are numbered and appear as light and dark bands. 22q11 is the 11 band (pronounced one-one) on the long arm of chromosome 22.5

Symptoms of DGS vary greatly from patient to patient depending on what body systems are affected and how severe the defects are.

Most cases of DGS result from a deletion of chromosome 22, but a small number of cases of DGS have defects in another chromosome, notably 10p13.6 Every person has two copies of chromosome 22, one inherited from each parent. In persons with DGS, one copy of chromosome 22 is missing a segment that includes an estimated 30 to 40 genes, which haven’t been clearly identified and aren’t well understood. The region of chromosome 22 that’s deleted in DGS is known as 22q11.2. This deletion usually occurs as a random event in the father’s sperm or in the mother’s egg, or it may occur early during fetal development. It is rarely an inherited condition passed to a child from a parent who also has deletions in chromosome 22 but may or may not have symptoms.7

Symptoms of DGS

Symptoms of DGS vary greatly from patient to patient depending on what body systems are affected and how severe the defects are. And, while some symptoms may be apparent at birth, others may not be apparent until later in infancy or early childhood.6 Characteristics of DGS include underdeveloped facial characteristics, parathyroid gland abnormalities, heart defects and thymus gland abnormalities. These characteristics can lead to a number of symptoms, including cleft palate, poor function of the palate, delayed acquisition of speech, difficulty feeding and swallowing, bluish skin due to poor circulation of oxygen-rich blood as a result of a heart defect, breathing problems, twitching or spasms around the mouth, hands or throat, frequent infections, delayed growth, failure to gain weight, poor muscle tone, delayed development, learning disabilities, behavioral problems and hyperactivity.8,9

Facial characteristics include an underdeveloped chin, eyes with heavy eyelids, ears that are rotated back and defective upper portions of the carilobes. With hypoparathyroidism, DGS patients may have trouble maintaining normal calcium levels, which can cause them to have seizures. Heart defects all involve the aorta and the part of the heart from which the aorta develops. The thymus controls the development and maturation of T lymphocytes, as well as helps B lymphocytes develop into plasma cells and produce immunoglobulins (antibodies). The smaller the thymus, the fewer T lymphocytes will be produced. T lymphocytes are essential for resistance to certain viral and fungal infections. Therefore, DGS patients are at increased susceptibility to viral, fungal and bacterial infections.7 Recurrent infections tend to decrease in late childhood and adulthood, with approximately one-third of affected adults having mild recurrent infections.5

Diagnosing DGS

Typically, DGS is diagnosed at birth or in infancy based on clinical observation. Historically, the diagnosis of DGS was made when at least three of the characteristics described previously were present, which caused many mild cases of DGS to be missed.6

Today, a variety of tests can help to diagnose DGS. Lab tests include a complete blood cell count and serum calcium and parathyroid hormone studies. Tests that evaluate T-cell count and function include flow cytometry, reverse-transcriptase polymerase chain reaction assay to assess thymic T-cell count for detection of TCR excision circles and antibody response studies. Imaging studies to diagnose thymic and cardiovascular abnormalities in 22q11.2 include radiography, magnetic resonance imaging, computed tomography scanning, echocardiography, and angiography and magnetic resonance angiography. Genetic studies can also be conducted, including the chromosomal microarray analysis or array comparative genomic hybridization, TBX1 gene study, multiplex ligation-dependent probe amplification and fluorescent in situ hybridization (FISH).10 The FISH test is the technology (previously mentioned) developed in the 1980s that can identify
deletions of 22q11 that are too small to be seen under the microscope. Today, FISH is the most definitive of the diagnostic genetic tests. It is not routinely conducted for every amniocentesis or from every blood sample from patients; instead, it is performed only when physicians suspect a 22q11 deletion in a person or a fetus. If a 22q11 deletion is detected in a child, both parents are offered the FISH test to determine if the child’s deletion is inherited. In approximately 10 percent of families, the deletion is inherited. And, an individual with a 22q11 deletion has a 50 percent chance with each pregnancy of passing it on to their child.

It should be noted, however, that a child may still have DGS even if the FISH test is negative. As noted previously, this technology only tests positive in approximately 90 percent of DGS patients. A small percentage of DGS patients have a deletion affecting the short arm of chromosome 10 that can be tested with a different FISH. But, most DGS patients who have a negative FISH test have no chromosomal abnormality that can be found currently. Therefore, if a child is diagnosed with DGS on the basis of certain characteristics, the diagnosis remains true even if the FISH test is negative.

## Treating DGS

Treatment of DGS patients differs depending on their specific symptoms, and many physicians will likely be involved. For instance, heart defects will be evaluated by cardiologists, cleft lips or palates will be evaluated by plastic surgeons and speech pathologists, feeding difficulties will be evaluated by speech and gastrointestinal specialists, and T-cell disorders and recurrent infections will be evaluated by immunologists.

Critical problems of DGS can usually be corrected with treatment. Hypoparathyroidism can typically be managed with calcium and vitamin D supplements. Cleft palate can be repaired with surgery. Surgery is also required to repair heart defects and to improve the supply of oxygen-rich blood.

If there is limited thymic function, infections are treated as they would be for all children, and the normal vaccine schedule is followed. The immune system function normally improves with age for those with moderate thymic impairment. If there is severe thymus impairment, treatment requires a transplant of thymus tissue, specialized cells from bone marrow or specialized disease-fighting blood cells. In rare cases in which the T-lymphocyte defect of the thymus is significant enough to cause the B lymphocytes to fail to make sufficient antibodies, immune globulin (IG) replacement therapy is required.

In 2012, researchers on behalf of the International DiGeorge Syndrome Immunodeficiency Consortium conducted an evaluation of the records of 1,023 DGS patients with a mean age of 5.5 years, 885 of which had immunoglobulin data available. The researchers examined immunoglobulin levels according to age, and found that low levels of immunoglobulin are present in a significant minority of patients and, overall, between 2 percent and 3 percent of those patients were receiving immune globulin replacement therapy. From that study, the researchers concluded that DGS is associated with significant humoral immune deficiency.

Many DGS patients also experience developmental, mental health or behavioral problems that can be treated with speech therapy, occupational therapy and developmental therapy.

## DGS Research

Much more needs to be understood about DGS to treat these patients and improve their outlook. Researchers are trying to identify the 30 to 40 missing genes on chromosome 22, many of which have not been well-characterized, that contribute to the variability in DGS characteristics and symptoms. For instance, they have found that the loss of a particular gene on chromosome 22, TBX1, is likely responsible for many of the DGS characteristic signs such as heart defects, cleft palate, distinctive facial features, hearing loss and low calcium levels, as well as behavioral problems. And, the loss of the COMT gene may also help to explain the increased risk of behavioral problems.

In 2013, an international team of researchers described a new mechanism by which most human cells can avoid being bombarded by DNA fragments. DGS is characterized by absence of the “microprocessor” protein complex, which means patients lack a “vigilante” gene to watch out for repeated sequences and, therefore, are potentially susceptible to being bombarded by DNA fragments. These researchers are now conducting studies, using an embryonic model of induced pluripotent stem cells donated by patients with DGS, to determine the impact of the repeated sequences during the embryonic stage. By examining...
the deletion that causes this pathology, they believe these studies will clarify the molecular base for DGS, as well as permit the long-term development of new therapies for its treatment.13

**Much more needs to be understood about DGS to treat these patients and improve their outlook.**

In 2014, researchers discovered information about the pathogenesis of feeding and swallowing difficulties often found in children with neurodevelopmental disorders. Using an animal model of DGS, the researchers found clear signs of early feeding and swallowing disruption and underlying changes in brain development, known as pediatric dysphagia. “A lot of children with pediatric dysphagia tend to be sicker from birth onward. Making the health of these kids as stable as possible from birth onward would allow clinicians to pick up on developmental signs sooner, which are often masked by more immediate problems like having ear or respiratory infections, not sleeping or not gaining weight,” said Anthony-Samuel LaManitia, PhD, professor of pharmacology and physiology at the GW School of Medicine and Health Sciences and director of the GW Institute for Neuroscience. “The physical stress caused by the complications of dysphagia early on likely exacerbates the fundamental behavior issues that will emerge later. A happy, healthy baby is often able to focus on observing and gathering information to drive important experience-dependent changes in the brain. A sick baby has less time to do so, possibly making cognitive outcomes even worse.”14

Researchers at the University of California, Davis, have found that for children with DGS, anxiety (but not intelligence) is linked to poorer adaptive behaviors such as self-care and communications skills that affect daily life. The study evaluated 78 children with DGS, ages 7 years to 15 years, with a battery of standardized tests related to behavior, anxiety, adaptive functioning and intelligence. Thirty-six typically developing children with no known genetic syndromes were also evaluated for comparison. Many anxiety scores were found to be significantly higher in children with DGS than in typically developing children. Fifty-eight percent of children with DGS were found to have at least one elevated anxiety score, although only 19 percent had previously been diagnosed with an anxiety disorder. In addition, higher anxiety scores correlated with lower adaptive function among children with DGS. The study findings suggest that helping children cope with fear-based symptoms may be the best strategy for increasing independence and protecting against psychiatric problems later in life.15

**DGS Outlook**

Until more is known, the outlook for DGS patients depends on the degree to which the organ systems are affected. The most important determining factor is the severity of heart disease. The deficit in T-cell production is also important, although the infection pattern appears optimistic since most patients don’t suffer from recurrent infections in adulthood.9

Tragically, a small percentage of children with DGS with severe heart defects and immune system problems won’t survive the first year of life. Those with less acute problems, who receive proper treatment, will survive into adulthood. Many will need extra help and long-term care for their individual health needs, as well as for their behavioral conditions.16 But with continued research, there is potential for better understanding and treatments.

And when the mystery of DGS is better understood, a cure cannot be far behind.

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**RONALE TUCKER RHODES, MS, is the editor of IG Living magazine.**

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**References**

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**PROFILE:**

Jessica King

By Trudie Mitschang

Jessica King, who was diagnosed with CVID in 2013, adopted Horus, a greyhound rescue, after feeling a strong connection with him.

**AFTER BATTLING AN** autoimmune disease as a teen and young adult, Jessica King celebrated a period of remission by living life to the fullest, even landing a job working for the Central Intelligence Agency (CIA). But, life threw her another curve when a series of health setbacks led to mid-career retirement and a diagnosis of common variable immunodeficiency (CVID).

Trudie: Tell us about your life before CVID.

Jessica: I suffered from and battled dermatomyositis (DMS), an autoimmune disease, from the age of 15 until I was 22. The disease absolutely wreaked havoc on my body, and the side effects from the years of numerous and various toxic cocktails of treatment regimens were horrific. When the disease finally went into remission, I transitioned from the mental state of “being sick” to “was sick.” Finally feeling free of the health baggage I had carried for so many years, I dove deep into anything and everything that came my way. As my first tattoo states, “Life without regret,” I strove to live my life to the fullest. Eventually, I found myself living in the outskirts of Washington, D.C., working for the CIA.

Trudie: How did you get involved with the CIA?

Jessica: I studied criminal justice in both undergraduate and graduate school with the goal of obtaining a job in the security field. A career as a security officer with the CIA offered vast opportunities applicable to my educational background and fit my criteria to have a job that both excited and interested me. My subsequent career with the CIA was filled with incredible experiences, opportunities and rewards that I thrived on until my medical retirement in November 2014.

Trudie: How were you diagnosed?

Jessica: I had pneumonia in 2008. The following year, while serving a yearlong assignment in Iraq, I became extremely ill. Eventually, I had to be medevacked back to the U.S., where I underwent surgery and received nearly two months of medical treatment. Concerned with the steady diet of powerful antibiotic medications she was prescribing combined with my seven-year history of being on immunosuppressant drugs, my primary care doctor recommended that I consult with an ear, nose and throat (ENT) doctor, who reviewed my medical history and ordered a CT scan of my sinuses.
that revealed abnormalities. The doctor recommended a tonsillectomy and sinus surgery in hopes that it would help alleviate the burden on my likely compromised immune system. I had my tonsils removed in February 2013 and sinus surgery in April 2013. My health spiraled downward in the months following the last surgery. Finally, in the beginning of August, the ENT threw up his hands and said that there was a problem greater than he could understand and referred me to an infectious disease specialist. It probably took me close to an hour to explain my entire medical history to the doctor, but within minutes, he hypothesized that I had an immune deficiency. He ordered a bunch of blood work, and the results confirmed a diagnosis of CVID.

**Trudie:** What was your treatment regimen?

**Jessica:** I received my first intravenous immune globulin (IVIG) treatment in October 2013 in the comfort of my own apartment, surrounded by loved ones who flew into town to be with me, the watchful eye of my dog, Horus, and the careful monitoring by the nurse. My nurse returned to my apartment every three weeks for the next year, caring for me as she would her own daughter while politely cursing my always vanishing veins.

**Trudie:** You have an athletic background. How has that kept you motivated?

**Jessica:** Being cognizant of how much better I feel within when I am active encourages me to take advantage of that precious time when I am not limited by my physical weakness. My innate craving for activity does not diminish when my physical strength diminishes. Instead, as it grows stronger, so do my dreams and aspirations. I dream of once again horseback riding through a foreign country as I did just a few years ago. I know this is a fathomable dream, a realistic goal to aspire, something I have hope in and thus something that provides me with tangible cause to take good physical care of myself.

**Trudie:** What has your illness taught you about resilience?

**Jessica:** I have learned that being resilient in my faith and hope is actually more important than focusing on my physical battles. I have come to accept that life with DMS and CVID means that I do not have control over all the ways my body can and will fail me. Although originally I thought losing such control of my health was the greatest possible loss in living a fulfilling life, I eventually learned that the strength of my mind is far more empowering than the strength of my body. I have directed my mind to keep the fight of my spirit alive and strong so that I focus on my faith in living a rewarding life despite my ailments.

**Trudie:** What would you like to accomplish with the rest of your life?

**Jessica:** I have been writing chapters of what I hope to someday be my published memoir off and on through the years, never really having the necessary dedication, time and/or desire to focus on it. I would consider it a success if I can provide inspiration, hope or encouragement to even just one person by telling and sharing my life’s journey. It is now my time in life to make a difference.

**TRUDIE MITSCHANG** is a contributing writer for *IG Living* magazine.
The Other Side of the Gown

By Stacy Oliver

FOR THE PAST eight years, I’ve been the one getting most of the major medical attention. Before being diagnosed with several autoimmune diseases, I saw one doctor once a year for a checkup. Now, I have a team of specialists. I have had several procedures, a few emergency room visits and one major surgery. I’m the one usually wearing the ever-fashionable hospital gown and blathering away on a high dose of painkillers.

Throughout it all, my husband has been there by my side guiding me through doctor appointments and hospital stays and keeping tabs on all my medical needs. That’s why when he had to have outpatient knee surgery, I was startled to see him in the gown. I felt like Alice on the other side of the looking glass. Here he was lying on the gurney in the gown. I couldn’t take my eyes off of him wearing it, the most ill-fitting, uncomfortable piece of clothing. Where was I? What’s happening?

For the first time in a long time, I was on the other side of the gown. My husband had an IV in his hand. This was all wrong. Then he was wheeled away, and I was left in the dreaded waiting room. I had brought magazines and a book, but I couldn’t focus. So I did what I usually do when I’m faced with stressful situations: I went to sleep.

Yep, when the nurse called me to finally go back to see him when he was out of surgery, I awoke startled. I became aware of this sleep reaction in college. There are two forms of it. The first is my sleep reaction to stress. For instance, when faced with a stressful writing assignment, I’d find the need to go back to my room and rest. Lying down and shutting my eyes helped me think. Maybe it’s the letting go of the physical and letting my mind drift that helps. I always came up with my best creative ideas this way. The second is my sleep reaction when faced with the emotional stresses of anxiety or fear. I find that letting go this way helps me escape traumatic situations. Even after my husband’s surgery and I was awake, I was still “zoned out” because it was so painful seeing the one I love so vulnerable.

It was then that I realized how much I trust the medical professionals I’ve chosen to be the guardians of my health. I listen and process what they have to say, but maybe with only one ear. I just go with the flow because I know my husband has my back and that he’s fully paying attention to everything involving my care. He’s wide awake and ready to be my biggest advocate. I am no doubt his, but I haven’t been in action for a while; I’ve been on the bench for most of his medical issues. So when called into the game, I was rusty.

Seeing him in that gown was a wake-up call to be more vigilant of his medical needs and mine. He’s the kind of person who had his surgery taped so he could watch the procedure afterward. That will never be me (blech!). But I can be his MVP on our health team in different ways. I can listen more carefully and take notes, do research on our ailments and make full use of the doctors while I have their attention. It still doesn’t mean I won’t drift off, but not when it comes to our care. When it comes to creative endeavors, that’s a different story. Like what am I going to write about in my next column? Zzzz…

STACY OLIVER was diagnosed in 2008 with multifocal motor neuropathy (MMN). She is the assistant director of the Center for the Writing Arts at Northwestern University, and she is working on her supersecret identity as Neuropathy Girl, who will one day save the world after her infusion and a nap.
Making a Plan

By Ilana Jacqueline

I’VE FOUND THAT the first step in pulling yourself up after a flare-up knocks you facedown on the pavement is to realize that you must have a plan. This epiphany should happen while you are still on the ground. Maybe you haven’t been discharged from the hospital yet. Maybe you’re home but are still in the throes of whatever pain or infection that started all of this. Maybe you’re asking: “Why should I even bother to make a plan when I can’t even get out of bed?”

When your life feels directionless and you’re in the midst of a post-illness depression, you have to turn your attention not to what you have missed out on or let fall through the cracks, but to what steps you’re going to take — not to get back to where you were, but to an even better place.

Recently, I found myself in a two-month-long in-and-out-of-the-hospital on heavy doses of painkillers kind of flare. And everything became unglued. I couldn’t work. I couldn’t plan for my wedding or my fiancé’s graduation party. I couldn’t keep up with my friends or care for my house, my appearance or relationships. And when the heavy waves that kept knocking me off my feet finally receded, I was left with the crumbled, disorganized pieces of my life with no clue how I was going to put it all back together and move forward.

You’re never going to walk out of a major flare, snap your fingers and watch your life magically put itself back together. It doesn’t happen overnight. It will take work and time. But, you’ll get there.

Making a plan saved me from the gut-punch of anxiety that hit me every time I looked at my overloaded inbox or piles upon piles of laundry. To start, I made a list of the sections of my life that needed help:

- My work
- My relationship
- My strength
- My organization
- My goals
- My wellness

1. **My strength.** Two months of what was practically bed rest left me in a feeble state. I wasn’t going to lift weights the next day, but I could slowly start taking supplements, get massages and adjustments to wake up my muscles, and begin walking around the neighborhood each day to increase my endurance. One small step at a time.

2. **My relationship.** This flare was difficult for me, but it was just as difficult (if not more) for the people I love. And with a chronic disease, this kind of pressure can weigh down on your relationships. What I needed was to make plans: going out to dinner, going to the movies and maybe even looking into going on a vacation in the near future. Most importantly: We needed to laugh and put a priority on having fun.

3. **My work.** I had scaled back on my hours considerably while dealing with this flare. But the moment I stepped out of the hospital, I went to my calendar and scheduled out my workweek. Slowly, over two weeks, I built back up my hours. And even when I had moments of panic about getting it all done, I felt reassured to know that I had every intention of being back to my regular schedule by the end of the month.

4. **My organization.** As soon as I’m able to make it to the grocery store, my first errand is to refill my hospital kit with things like toothpaste, deodorant, shampoo and cleaning wipes. I will plan a day to do all the laundry. I will call to reschedule all the appointments I missed and return all the voicemails I’ve barely had a moment to listen to. Slashing these simple errands off my list is an easy way to feel like I’m having a direct hand in the improvement of my life.

5. **My goals.** New Year’s is a great time for resolutions, but going through a particularly awful illness and coming out on the other side of it is an even better time! Having short- and long-term goals can be both motivating and stabilizing.

Once you’ve made your big list of must-dos, it’s time to break it down into smaller segments. Maybe on Monday you’ll focus on work and laundry, on Wednesday you’ll plan a date night for you and your partner, and Friday might be a good time to reschedule those missed appointments for the next week.

If your intention is to push the big red reset button on your life, you must reconnect with those you used to be in contact before you got sick, and meet the expectations of your friends, family, doctors and yourself. You have to try. And making a plan is what trying looks like.

**ILANA JACQUELINE** is a 24-year-old dysautonomia and primary immune deficiency disease patient from South Florida. She’s been writing professionally since 2004 on everything from health and wellness to celebrities and beauty. Her blog www.letsfeelbetter.com is both a personal collection of anecdotes about life with chronic illness, as well as a resource for patients of all ages.
OUR LIVING ROOM is engulfed with the spicy-sweet fragrance of cinnamon, nutmeg and allspice; it must be dessert time! The sound of Nana’s mixer whirring and whipping up sweet cream is the signal we are one step closer to the main event. Once the “cowboy coffee” is finished percolating and we’re in our places with bright shiny faces, we all know what happens next: presents! Handcrafted red velvet bows fly through the air while shiny slivers of wrapping paper begin to accumulate around ankles. Shouts of joy and squeals of delight compete for air space as “wish lists” are realized. The look on my husband Mark’s face confirmed my worst fears: I went a little overboard, again.

As the post-celebratory “hangover” begins to settle into my pores and pocketbook, the most painful side effect unfolds before my very bloodshot eyes: The gifts lay untouched while the cardboard boxes from which they were delivered morph into airplanes, playhouses, robots and the perfect shelter for hide-and-seek. Then, to add insult to injury, the kids beg me for the craft stuff (it’s under lock and key because glitter glue doesn’t come off hardwood floors) just so they can decorate the boxes. All the while, the brandnew, costly toys and electronic gadgets they begged us for — not to mention the monies we scrimped, saved and sacrificed “date night” for — lay among ancillary gifts like underwear and tube socks.

“That’s some expensive free cardboard,” Grandpop whispers, breaking the silence among the adults. This is where I saunter into the kitchen and drown my sorrows in various pies, fudge and a hefty dollop of sweet whipped cream.

Despite being in their teens, our primary immunodeficiency disease (PI) kids still become giddy over packages, especially when their medical supplies arrive at our door (Caleb and Molly are treated with subcutaneous immune globulin [SCIG] at home). You’d think the boxes were fabricated with 24-karat gold the way they revel in their arrival! No one would ever believe how excited our kids can get over a box of needles, syringes and alcohol wipes! And, I’ve learned a very important lesson not to open their boxes; that is a big no-no. There is a certain procedure when opening the boxes. You must be mindful of tearing the tape with great care, not be hostile with scissors and make sure you are in the right mind-set. Cranky, uptight and moody persons need not apply. It’s not just what’s in the boxes, it’s the great potential with and within the boxes that delights Caleb and Molly. Allow me a few paragraphs to describe their magical powers.

My kids attend a science, technology, engineering and mathematics (STEM) academy. Science and engineering equals lots and lots of experiments and projects. Because experiments and projects require sturdy boxes, packing peanuts, syringes, tubing and the occasional chuck pad, our kids become some of the most desired teammates on group assignments; Calvin, Caleb and Molly often hold high ranks in the hierarchy of middle school politics and social pyramids all because of their well-stocked coffers. Most recent was an assignment Molly received in English: Create a 3-D diorama book report for Charlotte’s Web. The rubric challenged the students to use recyclables of various shapes and sizes, and, of course, the more detailed, the higher the grade.

It’s What’s Inside That Counts

By Cheryl L. Haggard
“How do they expect you to pull off this book report using recyclables, Molly? Sheesh!!” I complained.

“No problem, Mom!” Molly announced with all the strength and confidence an almost-fourth-grader could muster.

I unleashed the glue gun, glitter and construction paper on my PI kid. Molly created a barn from her infusion supplies box, spray-painted tubing (from her most recent infusion) and used it for Charlotte’s web, and painted spent bottles of IG pink to assume the character Wilbur the pig! Even Wilbur’s tail, a mini-spike, impressed. And now Molly’s recycled, repurposed and reimagined barn from Charlotte’s Web is displayed in the school’s library. To some, the box and its contents represent a job well done, but for us it means so much more.

Just when I thought the projects and experiments of middle school were a thing of the past for Caleb, I find him in the garage, a can of gold spray paint in his left hand and box cutters in his right.

“What, pray tell, are you up to?” I questioned.

“Well, um…,” Caleb stammered, “a history project.”

“Since when does history use gold spray paint and box cutters?” I asked.

Now we know. Caleb’s assignment was to create a breastplate, spear, helmet and shield out of cardboard, duct tape and the like (they were studying Greek and Roman battle strategies). Then, they were going to live-action role-play (better known as LARP) with their cardboard-rigged gear when the unit was completed. Caleb worked for weeks, using as much of his supplies box and contents as creatively as he could, even using the mini-spikes as arrowheads! He was just about finished with his ensemble when he asked me for any ideas I might have for his breastplate.

“It’s the hardest part of this whole thing and the most important,” Caleb explained. “I’m really stumped!”

We were beyond the glue gun and glitter; we needed a creative intervention.

“Didn’t we get your box of supplies delivered today?” I asked my exasperated son.

“Yeah, but there isn’t anything new,” Caleb countered.

“Well, let’s take a look-see,” I said, with my glass half full.

Caleb carefully opened his box with ritualistic caution. Once we got past the first few layers of packaging materials (which makes for great wrapping paper! Very chic!), we saw something shiny, squishy and newish looking. Caleb managed to unfold the layers of bright, silver-y packaging material that was obviously new and perfect for a high school Roman breastplate!

“The box delivers again!” I said, high-fiving Caleb. We were able to cut a hole for his head to fit through, then tie the sides under his arms with tubing painted to look like leather. His breastplate was recycled perfection. We couldn’t wait to hear about his first battle, even though his immune system battles for his life every day.

“Well, how did it go?” I asked Caleb when he got home that afternoon from school.

“It was awesome!” Caleb announced, heading to our backyard. “We had so much fun practicing trench warfare and taking on the upper classmen in hand-to-hand combat. It was the best day of school ever!”

“Where you going now?” I asked, watching Caleb make his way to the backyard where he had about three of his supply boxes set up with bull’s-eyes drawn on them.

“Target practice!” he replied.

For my PI kids, what’s inside the box and the boxes themselves are equally important, especially when it comes to their SCIG supplies. But, for me, what’s on the inside of those boxes is what saves a life, and that’s what really matters most! ☀️

CHERYL L. HAGGARD is a stay-at-home mom and has three children with PI, two of whom have CVID.
Is It Time to Homeschool Your Chronically Ill Child?

By Jessica Leigh Johnson

IT’S RARE TO turn on the TV these days without hearing about pandemic bird flu or a brand-new virus sickening the nation in “unprecedented” proportions. One month, enterovirus is causing widespread panic among parents; the next, it’s Ebola or measles. As a mother with three children who suffer from a primary immunodeficiency (PI) disease and are vulnerable to infection, headlines like these frazzle my nerves and keep me awake at night. When my protective instincts kick in, I consider pulling my boys from the biggest germ factory of all — public school — and teaching them at home, where they’ll be safe.

While removing a child from school every time a new superbug makes headlines is a bit of an overreaction, there are times when teaching a chronically ill child at home is preferable to sending him or her to school. But how do parents know when it makes more sense to educate their chronically ill child at home rather than in a traditional school setting?

While there’s no magic checklist that will give parents a definitive answer, taking these factors into consideration may aid them in making this difficult decision.

Frequent absence. Surprisingly, most truancy specialists do not consider frequent absence reason enough to remove a child from a traditional school setting on a permanent basis. If parents are determined to keep their chronically absent child enrolled in public school, there are ways to work around the issue of multiple missed school days.

Chronic absence is defined as missing 10 percent or more of school days. When a child is determined to be chronically ill, the parents can fill out a 504 plan specifying the modifications and accommodations needed for the student to perform at the same level as his or her peers. This can include a tutor who comes to the home so the child can keep up with schoolwork and not fall behind the other students in class.

According to children-with-special-needs expert Terri Mauro, “Section 504 of the Rehabilitation Act and the Americans with Disabilities Act specifies that no one with a disability can be excluded from participating in federally funded programs or activities, including elementary, secondary or postsecondary schooling.” Disability in this context can refer to a physical impairment such as an illness or chronic condition.

Unfortunately, even if a tutor comes to the home, sometimes the amount of time the child spends with that tutor is not enough to keep up with classmates. Tutors cost money, and because of lack of funding, some schools may be able to afford only one hour of tutoring per day for a homebound child. Despite the school system’s best efforts, the child may still fall behind. In this case, homeschooling would provide more instruction time and may improve the child’s chances of success.
More individualized attention. With upwards of 25 students in a classroom, teachers can’t possibly focus enough attention on a particular student in order to monitor rapid changes in his or her health. Many times, school nurses split their days between several schools and are not always available in a medical emergency. For peace of mind, some parents choose homeschooling so they can keep a closer eye on their child. The health status of a child with conditions like asthma, food allergies and diabetes can change quickly and unpredictably, and paying close attention to symptoms at their onset can be lifesaving.¹

The need for flexibility. A chronically ill child often has numerous medical appointments and procedures. Homeschooling offers the flexibility to schedule these appointments without having to work around school hours.

A chronically ill child will also have days when he or she is just not feeling 100 percent and may need more down time than his or her peers. After a day-long infusion, that child can be tired and lethargic. It may take a day or two to recover from a particularly draining procedure. Homeschooling allows the child to tackle schoolwork in smaller amounts while taking frequent breaks to rest and then resume studies when ready.

Less exposure to germs. While many chronic conditions such as PI can be well managed with immune globulin therapy, a chronically ill child can be extremely vulnerable to even the most common cold germs. For a child with complex immune deficiencies involving more than one part of the immune system, or a child on chemotherapy or undergoing bone marrow transplant, exposure to a classroom full of children poses a dangerous threat to their health. It doesn’t take a doctor to figure out that the fewer people a chronically ill child comes in contact with, the better chance he or she will have to stay healthy. For parents of an ill child more vulnerable to infection, homeschooling may be the safest course of action.

Less stress for both child and parents. The life of a chronically ill child can be complicated enough without adding the hassle of dealing with school personnel who don’t understand the child’s disease, and often wonder why a child who “looks” healthy is sick so many days. Some parents choose homeschooling simply to eliminate this one area of constant stress.

Sometimes a child with a rare and misunderstood condition is made fun of or picked on due to adverse symptoms. With homeschooling, the child is taught in a safe and comfortable environment, free from negative stressors that can detract from his or her ability to learn and focus on schooling.

Along with the benefits of homeschooling come drawbacks that must be taken into consideration, a major one being the potential lack of socialization in a homeschool setting. Only part of school is actual book learning; the other half is social skill learning, which the child will need throughout the rest of his life.² Making friends, dealing with or ignoring bullies and working with others are important social skills that cannot be taught but rather are learned through firsthand experience. Homeschooling parents can combat this issue by involving their child in sports, community organizations such as 4-H or local theater groups, and church youth programs, to name a few.

One last thing to consider when choosing homeschooling is the quality of the curriculum and the qualifications of the teacher. If parents don’t feel qualified, they can look to their local public schools to see what types of homeschooling options are available. The options, governed by each district’s local education agency, will differ depending on the state and region. In addition, there are many online homeschool programs with licensed instructors. In order to choose a high-quality curriculum, parents must wade through a vast array of options before finding the right one. They can start by looking for one that parallels their worldview, uses their preferred learning approach and fits their budget.³ Many resources are available online for parents just starting out on the homeschool journey, including www.k12.com, howtohomeschoolforfree.com and home school.com.

No matter what others choose, parents have to follow their instincts and do what they feel is best for their family. Just as no two children are alike, there is no one-size-fits-all approach to educating a child, especially if he or she suffers from chronic illness (and frequent absences). A child who is able to maintain a high level of attendance can thrive in public school. But when circumstances make that difficult, parents do have the option to homeschool, and, hopefully, they’ll find support for their decision. Parents, teachers and the school system can work together to find the solution that makes the most sense and leaves both child and parents feeling comfortable. ■

JESSICA 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
Understanding Allergies and How to Treat Them

By Trudie Mitschang

ALLERGY SYMPTOMS CAN make life miserable for anyone, but for the primary immune deficiency (PI) patient, these bothersome symptoms can be especially hard to deal with. Even more frustrating is the fact that it can be difficult to distinguish between symptoms caused by PI itself and those that are the byproduct of allergic reactions.

Understanding Allergies
Reactions to allergens occur when a person develops “allergic” antibodies, known as IgE, which are each specific for a particular allergen such as pet dander, pollen or dust mites. The IgE antibodies bind tightly to allergic cells, called mast cells, in the skin, airways, gastrointestinal tract and around blood vessels. The allergic cells are activated when the bound IgE recognizes an allergen, after which the cells release many mediators of the allergic reaction, including histamine, a chemical that can cause hives, swelling, runny nose, sneezing and itching. Depending upon where in the body the reaction between the IgE and the allergen happens, different symptoms can occur.

At the first sign of allergies, most people blame the allergens, but most allergens themselves are actually harmless. It’s the immune system’s misguided reaction to common allergens that causes the troublesome symptoms. We develop allergies from a critical part of the immune system that is responsible for dealing with parasites. All the mediators released in an allergic reaction are intended to make parasites become irritated in an inhospitable environment so they leave the body. Unfortunately, allergens such as pollen grains cannot be made to leave, so there is a continued attack with the release of the allergic mediators. And, for PI patients, specific changes to the immune system may increase the risk of developing allergies and make the symptoms even worse.

Diagnosing Allergies in Immune Deficient Patients
Diagnosing allergies can be tricky, especially in PI patients. A doctor may recommend a skin prick test, which involves taking a drop of allergen and poking the surface of the skin. If a positive reaction occurs, a rash that looks like hives will develop. Another common method for uncovering allergies is a blood test to detect and measure the amount of allergen-specific IgE antibodies in the blood. Allergy blood tests can screen any of the most common allergy triggers, including dust, pet dander, trees, grasses, weeds and molds, and they can also be used to detect food allergies. The test

Benadryl (diphenhydramine)
This antihistamine’s name has almost become synonymous with allergy relief. Diphenhydramine blocks the effects of the naturally occurring chemical histamine in the body. Benadryl is available as a liquid, tablet and topical medication, and is available in formulas for children and adults to treat sneezing, runny nose, itching and watery eyes, hives, rashes, itching and other symptoms of allergies.

Zyrtec-D (cetirizine and pseudoephedrine)
This combination medicine is used to treat cold or allergy symptoms such as nasal and sinus congestion, sneezing, itching, watery eyes or runny nose. Cetirizine is an antihistamine that reduces the natural chemical histamine in the body. Pseudoephedrine is a decongestant that shrinks blood vessels in the nasal passages.

Over-the-Counter Best Bets

Nasalcrom (cromolyn sodium)
This nasal spray is used to prevent allergy symptoms such as runny nose, stuffy nose, sneezing, itching and post-nasal drip. It is an anti-inflammatory medication that works to keep allergens from reaching the mast cells so fewer histamines are released.

Equate (phenylephrine)
Equate is used to relieve nasal discomfort caused by colds, allergies and hay fever. It is also used to relieve sinus congestion and pressure. Phenylephrine will relieve symptoms but will not treat the cause of the symptoms or speed recovery. It works by reducing swelling of the blood vessels in the nasal passages.
**Prescription Picks**

**Promethazine (Phenergan)**
This antihistamine is used to treat allergy symptoms such as itching, runny nose, sneezing, itchy or watery eyes, hives and itchy skin rashes. Phenergan also prevents motion sickness, and treats nausea and vomiting or pain after surgery. It is also used as a sedative or sleep aid.

**Pseudoephedrine (Sudafed)**
This medicine is used to relieve nasal congestion caused by colds, allergies and hay fever. It is also used to temporarily relieve sinus congestion and pressure. Pseudoephedrine will relieve symptoms but will not treat the cause of the symptoms or speed recovery. It is in a class of medications called nasal decongestants, and works by causing narrowing of the blood vessels in the nasal passages.

**Cromolyn Sodium Ophthalmic Solution**
This anti-inflammatory medication works by preventing the release of substances in the body that cause inflammation. Cromolyn sodium ophthalmic is used to treat allergy symptoms that affect the eyes such as itching, burning, watering, swelling, redness or sensitivity to light.

**Montelukast (Singulair)**
Montelukast is an inhibitor that works by blocking a substance called leukotriene, which helps to decrease certain asthma and allergy symptoms. Montelukast is also used before exercise to prevent breathing problems (bronchospasm) and to relieve symptoms of hay fever and allergic rhinitis (such as sneezing, stuffy/runny/itchy nose). It helps make breathing easier by reducing inflammation in the airways.

**Atrovent (Ipratropium)**
This prescription nasal spray treats a runny nose by stopping the production of mucus. It is available in two strengths to treat runny nose due to allergies and for symptoms of hay fever and cold in children and adults.

is known as a RAST test, and it can be further used to find many and rare items that a person may be allergic to.

One of the challenges faced by PI patients and their doctors is the overlap of symptoms between allergies and the disease itself. Overlapping symptoms may include runny nose, sneezing, stopped-up nose or head, frequent sinus and ear infections, loss of taste and smell, trouble concentrating, tiredness and/or trouble sleeping.

**Finding Relief: Medicine Cabinet Checklist**
The first line of approach for treating allergic disease is avoidance. Some recommendations include checking food packaging and not eating food items that one is allergic to, avoiding visiting homes with pets to which one is allergic, and using specific allergen-certified pillow and mattress encasings to reduce dust mite exposure. Many more recommendations can be found at the American Academy of Allergy, Asthma and Immunology and American College of Allergy, Asthma and Immunology websites (AAAAI.org and ACAAI.org).

If you suffer from frequent allergies, your doctor may prescribe a symptom-specific medication to relieve your suffering. Allergy medications fall into the following categories:

- **Sedating and non-sedating antihistamines.** Sedating antihistamines relieve allergy symptoms but may cause drowsiness and other side effects. Newer antihistamines are said to be non-sedating, although some users may still experience drowsiness. Antihistamines compete with histamine to prevent or reduce the signs and symptoms of an allergic reaction. They are available as oral medications, creams, lotions, nasal sprays and eye drops.
- **Corticosteroids.** These help reduce inflammation. They are available as nasal sprays, topical creams and ointments, tablets, injectables and eye preparations.
- **Mast cell stabilizers.** During an allergic reaction, mast cells release histamine and other substances. Mast cell stabilizers, such as cromolyn sodium, keep these cells intact.
- **Leukotriene inhibitors.** Leukotrienes are released during an allergic reaction and can aggravate allergic conditions and asthma. These drugs target leukotriene receptors.
- **Nasal anticholinergics.** These reduce discharge from the nose.
- **Decongestants.** Decongestants work by constricting blood vessels, which limits the amount of secretions coming from the inner lining of the nose.
- **Immunomodulators.** These topical medications are used to treat severe skin allergies such as severe eczema.
- **Autoinjectable epinephrine.** This medication is used to treat a life-threatening allergic reaction known anaphylaxis caused by severe allergic response to foods, drugs or insect stings.
BOOK CORNER

The Autoimmune Paleo Cookbook: An Allergen-Free Approach to Managing Chronic Illness
Author: Mickey Trescott
Publisher: Trescott LLC

This book is a resource for those looking to embark on a diet that focuses on removing potential food triggers and healing the gut. The first section of the book explains the autoimmune protocol — what it is, why it works and which foods to eat and avoid while on the elimination diet. Instructions are given on how to clear the pantry of questionable ingredients and replace them with healing, nutrient-dense whole foods. Also included are sections on food quality, a shopping guide, tips and tricks to make the protocol go more smoothly and ideas for batch-cooking and breakfast. Two four-week meal plans are included, as are 112 recipes suitable for anyone on the strictest phase of the autoimmune protocol: no grains, beans, dairy, eggs, nuts, seeds or nightshades.

How to Be a Friend to a Friend Who’s Sick
Author: Letty Cottin Pogrebin
Publisher: PublicAffairs

Throughout her recent bout with breast cancer, Letty Cottin Pogrebin became fascinated by her friends’ and family’s diverse reactions to her and her illness: how awkwardly some of them behaved; how some misspoke or misinterpreted her needs; and how wonderful it was when people read her right. After talking to her fellow patients and dozens of other veterans of serious illness, seeking to discover what sick people wished their friends knew about how best to comfort, help and even simply talk to them, Pogrebin distilled their collective stories and opinions into a compendium of practical guidance and usable wisdom. Infused with sensitivity, warmth and humor, the book shares candid stories from her own and others’ journeys and their sometimes imperfect interactions with well-meaning friends.

The Moth Eaten World
Author: Suzanne Edison
Publisher: Finishing Line Press

When Suzanne Edison’s child was diagnosed with an autoimmune disease, she “used writing as a way to understand and express the range of feelings and experiences” she was having. The Moth Eaten World is a translation of her fear, anger, grief and hope — a roller-coaster ride of emotions — into a book of poetry about raising a child with chronic illness. Poem by poem, she explores not just a failing body, but a daughter’s disease and a mother’s journey through this world. The poems about strength and struggle, beauty and fear, faith and doubt … offer the reader a clear view of a catastrophic situation made palatable by Edison’s writing skills.

new and useful reading

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You Don’t Look Sick! Living Well with Invisible Chronic Illness
Authors: Joy H. Selak and Steven S. Overman, MD, MPH
Publisher: Haworth Medical Press

You Don’t Look Sick! is a patient’s true-life accounts and her physician’s compassionate commentary on the three stages of chronic illness: getting sick, being sick and living well. The book is written for patients in all stages of the chronic illness journey. It addresses practical aspects of chronic illness such as hiring a doctor, managing chronic pain, coping with grief and the loss of function, winning battles with health and disability, recognizing the limitations of chronic illness care and charting a path for change. Included are stories, dialogue and humor, as well as suggested reading materials for learning to live well, medical Internet resources, illness-specific websites, names and addresses of national associations and a bibliography of medical books by topic.
### Ataxia Telangiectasia (A-T)
- **WEBSITES**
  - A-T Children’s Project: [www.atcp.org](http://www.atcp.org)

### Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
- **WEBSITES**
  - GBS/CIDP Foundation International: [www.gbs-cidp.org](http://www.gbs-cidp.org)
  - The Neuropathy Association: [www.neuropathy.org](http://www.neuropathy.org)

### Evans Syndrome
- **ONLINE PEER SUPPORT**
  - Evans Syndrome Research and Support Group: [www.evanssyndrome.org](http://www.evanssyndrome.org)

### Guillain-Barré Syndrome (GBS)
- **WEBSITES**
  - GBS/CIDP Foundation International: [www.gbs-cidp.org](http://www.gbs-cidp.org)
  - The Neuropathy Association: [www.neuropathy.org](http://www.neuropathy.org)
  - GBS Support Group: [www.gbs.org.uk](http://www.gbs.org.uk)
  - GBS/CIDP Foundation International Discussion Forums: [www.gbs-cidp.org/forums](http://www.gbs-cidp.org/forums)

### Idiopathic Thrombocytopenic Purpura (ITP)
- **WEBSITES**
  - ITP Support Association – UK: [www.itpsupport.org.uk](http://www.itpsupport.org.uk)
  - Platelet Disorder Support Association: [www.pdsa.org](http://www.pdsa.org)

### Kawasaki Disease
- **WEBSITES**
  - American Heart Association: [www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsOfChildhood/Kawasaki-Disease_UCM_306777_Article.jsp#.T1T2boePW E0](http://www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsOfChildhood/Kawasaki-Disease_UCM_306777_Article.jsp#.T1T2boePW E0)
  - Kawasaki Disease Foundation: [www.kdfoundation.org](http://www.kdfoundation.org)
  - KidsHealth: [kidshealth.org/parent/medical/heart/kawasaki.html](http://kidshealth.org/parent/medical/heart/kawasaki.html)

### Mitochondrial Disease
- **WEBSITES**
  - United Mitochondrial Disease Foundation: [www.umdf.org](http://www.umdf.org)
  - MitoAction: [www.mitoaction.org](http://www.mitoaction.org)

### Multifocal Motor Neuropathy (MMN)
- **WEBSITES**
  - The Neuropathy Association: [www.neuropathy.org](http://www.neuropathy.org)

### Multiple Sclerosis (MS)
- **WEBSITES**
  - All About Multiple Sclerosis: [www.mult-sclerosis.org/index.html](http://www.mult-sclerosis.org/index.html)
  - Multiple Sclerosis Association of America: [www.msaa.com](http://www.msaa.com)
  - National Multiple Sclerosis Society: [www.nationalmsociety.org](http://www.nationalmsociety.org)

### Peripheral Neuropathy (PN)
- **WEBSITES**
  - Neuropathy Action Foundation: [www.neuropathyaction.org](http://www.neuropathyaction.org)
  - The Neuropathy Association: [www.neuropathy.org](http://www.neuropathy.org)
  - Texas Chapter of the Neuropathy Association: [www.handsfeethroat.org](http://www.handsfeethroat.org)

### Primary Immune Deficiency Disease (PI)
- **WEBSITES**
  - Immune Deficiency Foundation: [www.primaryimmune.org](http://www.primaryimmune.org)
  - Jeffrey Modell Foundation: [www.info4pi.org](http://www.info4pi.org)
  - American Academy of Allergy, Asthma & Immunology: [www.aaaai.org](http://www.aaaai.org)
  - International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: [www.ipopi.org](http://www.ipopi.org)
  - New England Primary Immunodeficiency Network: [www.nepin.org](http://www.nepin.org)
  - Rainbow Allergy-Immunology: [www.uhhospitals.org/rainbow/services/allergy-immunology](http://www.uhhospitals.org/rainbow/services/allergy-immunology)

### Scleroderma
- **WEBSITES**
  - Scleroderma Foundation: [www.scleroderma.org](http://www.scleroderma.org)
  - Scleroderma Research Foundation: [www.srfcure.org](http://www.srfcure.org)
  - Scleroderma Center: [www.hopkinsmedicine.org/rheumatology/clinics/scleroderma_center.html](http://www.hopkinsmedicine.org/rheumatology/clinics/scleroderma_center.html)

### Stiff Person Syndrome (SPS)
- **WEBSITES**
  - American Autoimmune Related Diseases Association Inc.: [www.aarda.org](http://www.aarda.org)
  - Genetic Alliance: [www.geneticaIlalliance.org](http://www.geneticaIlalliance.org)
  - Living with Stiff Person Syndrome (personal account): [www.livingwithsp.com](http://www.livingwithsp.com)
  - Stiff Person Syndrome: [www.stiffpersonsyndrome.net](http://www.stiffpersonsyndrome.net)
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