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SELF-CARE IS is so important for maintaining mental, emotional and physical health. And, while overlooked by many, its necessity is paramount for those who suffer the effects of chronic illness. Aside from the basics of getting enough sleep, exercising regularly and eating a nutritious diet, chronically ill patients must contend with additional self-care issues such as unrelenting fatigue and pain while managing day-to-day life — which is the focus of helpful articles in this issue.

We begin with the topic of raising kids, normally challenging enough, but when parents are battling symptoms that limit the ability to perform daily activities, it takes a toll on the parent with the illness and can profoundly impact the children. Fortunately, as we explain in our article “Raising Kids While Coping with a Chronic Illness” (p.18), psychologists have created a roadmap to help parents help their kids, including being honest about their condition and talking to them on an age-appropriate level. Just as importantly, parents must deal with their own emotions by learning to live with their limitations and seeking support.

Unfortunately for many families, chronic illness can strike not only one person in the household, but multiple family members. This is the case for Denna McGrew who authored the article “Organizing the Chronically Ill Chaos” (p.22). With the experience of her family’s 17-year chronic illness journey, she devised a strategy to manage all the information needed by the multiple specialists her family members see. She likens her strategy to the game of football in which the patient is the quarterback and the specialists are the coaches. For those who have not yet developed a “go-to” document that accompanies them on every doctor visit, Denna’s template and description of what to include in each section may be extremely helpful for organizing the chaos.

Pain is one of the most common symptoms accompanying chronic illness. And, NSAIDs are a good nonnarcotic option to help relieve it. Although NSAIDs don’t require a prescription, they do pose dangers, especially when used long-term. That’s why it’s important for patients to understand their potential side effects and contraindications as we detail in our article “How to Properly Use NSAIDs” (p.34).

In many instances, NSAIDs won’t be enough to manage pain caused by illness. And, while controversial, in the past two decades, 33 states have approved medical marijuana laws to relieve pain and other symptoms. Our article “Managing Chronic Pain with Medical Marijuana” (p.28) takes a look at how cannabis works, the types available and where it can be purchased. And, to better assist patients in determining whether medical marijuana is something they want to explore, the article examines several studies that both support and refute its beneficial effects.

As always, we hope you enjoy these articles, as well as the many more educational and insightful topics presented in this issue of IG Living.
Gene Editing: The Good and Bad

By Abbie Cornett

A WORLD WITH no disease sounds like science fiction. Yet, while it is science, it may no longer be fiction. In recent years, methods of improving human health have made some amazing leaps, the most promising of which is a game-changing genetic engineering technique called CRISPR, or its more technical name CRISPR-Cas9.1

CRISPR, which stands for clustered regularly interspaced short palindromic repeats, is currently being used for both human and nonhuman applications. The technology holds the potential to cure many untreatable diseases, but it also poses tremendous risk to the human genome if it is pursued without ethical guidelines.

The most serious ethical concern is gene editing targeting germ cells. Any changes in germ cells can be potentially passed on to future generations, essentially introducing permanent changes in the human population.2 In 2015, the first attempt at editing human embryos yielded unsuccessful results. Out of 54 embryos, all but four failed. And, the four that had successful genetic changes all had potentially dangerous off-target mutations.

The ethical implications of CRISPR’s first off-target mutations were obvious. But, this clear line has faded now that CRISPR has evolved, bringing its error rate closer to zero.3 Currently, we are faced with ethical decisions about when editing the germ line to create inheritable changes is acceptable such as when curing disease, versus editing the germ line to enhance intelligence, increase longevity and create designer babies and hybrid humans.

In November, two genetically engineered girls were born in China. A scientific team at the Southern University of Science and Technology in Shenzhen, China, used CRISPR to alter the girls’ DNA to make them immune to the human immunodeficiency virus (HIV), a modification that can be passed on to future generations.4 Prior to this, scientists were using CRISPR only to cure diseases by editing somatic cells — changes that cannot be passed onto future generations.

On the surface, gene editing seems like an amazing accomplishment — but at what cost? The twins’ DNA was altered by editing a gene called CCR5, which allows HIV to enter human blood cells. But, research shows CCR5 also plays a role in memory and cognition. Not only does deleting this gene make mice smarter, it also improves human brain recovery after stroke, and it could be linked to greater success in school.4

The Chinese research team’s actions have been widely condemned by the world scientific community for a number of reasons. Most importantly: There is no way to predict the outcomes of this alteration on the twins’ development or what other repercussions may result.

For now, gene editing is experimental and is still associated with off-target mutations capable of causing genetic problems both early and later in life, including the development of cancer.2 And, while gene editing holds enormous possibilities such as developing therapeutic treatments for genetic disorders, HIV, cancer and myriad other conditions, it has many significant technical issues. While we now have the potential to cure disease, this technology can also be used to change what it means to be human. As such, gene editing of the human germ line should not be considered until a rigorous and thorough evaluation of the safety of the technology is undertaken. Further, its ethical implications need consensus by the global scientific community.

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References
More times than I can count. Doctor egos are often a cruel thing. So many refuse to admit or accept their limitations. I choose to be grateful for the physician I have now, but I still hope to never encounter any of those people again.

— Amy W

Oh, yes. It made my family members think it was all in my head. Later, I had proof it wasn’t, of course. What a horrible thing to experience and then have to experience the aftermath of a chronic infection, etc. At least with the primary immunodeficiency disease diagnosis, those doctors are afraid to say it’s in my head.

— Vicki DH

Have you ever been told it’s all in your head?

I should take up bowling. Did I have a husband? Oh, your divorce started these “issues.” I did not want to work, so I retired with this “illness”; maybe you should go back to work. And, [here are] my favorites after I was diagnosed. From my then-allergist: “I never thought you had that. You don’t look sick.” From my then-primary care physician: “Oh, you have that?” I was diagnosed at age 56. Years wasted.

— Betty FG

Are You Afraid of Going to the Emergency Room?

Absolutely! [Especially because of] the germ exposure with selective antibody immunodeficiency. I was literally told on more than one occasion: “You can’t have that.” “It’s too rare.” “I’m not treating you for something you can’t have.” Yeah, filing a complaint was a joke.

— Renee WL

Yes! The emergency room doctors don’t have a clue when it comes to common variable immunodeficiency! It’s so frustrating to have them call my doctor to confirm I regularly get pneumonia, and if I am not breathing well and coughing up blood with a fever, they should indeed look at me! It’s the same routine every time!

— Carly F

Have you been confronted with devastating delays due to needed prior authorization?

I find when an authorization is up for renewal, I try to prepare for any and all questions, responses, delays and, scariest of all, denials. It seems sometimes there is no such thing as being prepared because [of] the additional requests for more and more information [that] have never been asked [before] and, many times, I wonder what the relevance is of all that is requested. It’s during these difficult times, especially if I am already sick, that I want to throw in the towel. It takes a lot of patience and energy to enter the fight. Sometimes while I am fighting, I will just withdraw and temporarily throw in the towel. Luckily for me, I realize I have to keep fighting and never give in. I have too many reasons to fight, and I am not going to give in and let the other party win. My faith, doctors, husband, family and friends always seem to know the right things to say, they encourage me with just the right words and a hug never hurts the cause. I think what I am trying to say is never give up.

— Jenny G

I am a firm believer in checking: checking if referrals have been sent and received; checking doctor rating credentials; checking medication interactions — everything. If I don’t do it, I get lost in the system. Plus, checking doesn’t mean being bi–hy; It means being proactive and caring for myself.

— Birgit C
**Abbie** I spoke with Roger Kobayashi, MD, regarding your question. He said, in general, indwelling ports should never be used in PI patients since they are associated with thrombosis, which can result in stroke and renal infarction, and they can occasionally become colonized and discharge septic emboli. Further, IVIG is typically given every four weeks or occasionally every three weeks, rather than every two weeks. Is there a reason you are receiving IVIG every two weeks, and has your doctor measured your IgG trough levels?

Because of the issues you are experiencing, you should discuss with your doctor switching to subcutaneous IG (SCIG), since there are rarely associated medical contraindications. With SCIG, there is no need to access a vein since the IG is infused via small needle sets just below the skin. SCIG is typically infused every week, with the total monthly dose divided by four. For instance, if you are treated with 30 grams per month with IVIG, you would be treated with 8 grams per week (rounded up), 10 grams every 10 days or 15 grams every two weeks with SCIG.

**Question** Can a tumor marker be passed from a donor to a patient through an IVIG infusion?

My husband was recently told he has an autoimmune neuropathy, and he was prescribed 40 grams of intravenous immune globulin (IVIG) twice a week for two weeks and then once every other week for a total of 11 infusions. After his fourth infusion, his liver enzymes were elevating, so the treatment was stopped and he was sent to a liver specialist. The specialist performed blood work, and all tests came back normal except a tumor marker called CEA (carcinoembryonic antigen), which was super elevated at 19.4 (normal is 0 to 5, and anything over 10 is almost positive for metastasized cancer). Because of the normal blood tests, the specialist felt it was certain IVIG was the cause of his elevated liver enzymes. But, due to his elevated CEA, a chest CT scan, an MRI with and without contrast of the abdomen and pelvis, a PET scan and a colonoscopy were performed to look for cancer, but all were normal. The liver specialist said it’s possible a CEA level of 19.4 could be normal for my husband, but he had never seen that. Therefore, it was hypothesized that a CEA tumor marker may have been passed on by a plasma donor through the IVIG infusion. My questions are: Have you ever heard of a tumor marker being passed through a plasma transfusion? And, if so, is there any possibility that this may develop into cancer in my husband?

**Dr. Kobayashi** The answer is unequivocally no that CEA in high levels could be passed through IVIG or subcutaneous IG infusions, and there are no studies reporting this. It also would not be possible for the infusion of IVIG to cause cells to secrete CEA (which is an immunoglobulin superclass). Also, if a donor had an elevated CEA, it would be vastly diluted among the 5,000 to 50,000 other donors included in the batch that made up the IG solution. Further, noncancerous patients do not have a markedly elevated CEA [it can be positive in infections, rheumatoid diseases and pulmonary diseases, but not typically elevated].

CEA has poor sensitivity and is not recommended for cancer screening. This, then, raises the question of whether there was a reason for ordering the CEA test. The CEA tumor marker test is an old test that has been around since 1965 and is not used for screening. In my opinion, it would be prudent to follow the patient now that there is a positive result from a nonspecific test.
Blood Testing Issues for PI Patients, Part 2

By Terry O. Harville, MD, PhD

**NUCLEIC ACID TESTING** (NAT) is a modern method of testing blood for infectious microorganisms. Nucleic acid is DNA and RNA. DNA is required by all organisms for them to develop, grow and reproduce, except for some viruses that use RNA as the genetic information, and prions that do not appear to use nucleic acid for propagation. Since each organism has some specific DNA sequences (or specific RNA sequences), targeting these regions can be used to identify which microorganisms are present during an infection. Depending on the testing platform, NAT results may be obtained within four hours, therefore greatly accelerating treatment with the correct antibiotics.

Traditional testing methods grow microorganisms in culture media, which can take several days (possibly weeks for tuberculosis bacteria). And, unfortunately, there may be microorganisms that do not grow well, and for many viruses, there are no culture techniques. When growth is detected, microorganisms undergo staining to help identify which microorganisms may be present. Then, secondary culturing with specific media that support the growth of particular microorganisms is performed for further identification. This process generally works well and has been the standard for decades. The downside to this traditional process is it can take several days, which may delay the most appropriate therapy.

The advantage of NAT over traditional testing is results can be obtained within a few hours. Another plus is the specificity of the nucleic acid target sequences to better identify the microorganism, which again can result in a more definitive and faster therapy.

NAT can be used in many circumstances. For instance, it can be used to diagnose meningitis in infants and young children (or any age) who present with fever and symptoms indicative of the disease. Using NAT to identify a common virus in cerebral spinal fluid known to cause viral meningitis, and not finding the bacteria associated with bacterial meningitis, can allow for symptomatic treatment rather than the use of antibacterial antibiotics — all within a few hours. Further, this testing allows a patient to go home from the emergency room rather than be admitted to the hospital, thereby providing better care at a lower cost. NAT can also be used for patients undergoing bone marrow or organ transplantation, in whom reactivation of cytomegalovirus can create problems. With NAT, it is easier to detect reactivation of the virus, at times even before symptoms present, allowing therapy to begin before problems arise.

As discussed in the last column, laboratory tests that use antibodies made by the patient to identify the presence of microorganisms is unreliable in those with immunodeficiencies and in those receiving immune globulin (IG) replacement therapy. Hepatitis B infection is one example of this. Anti-hepatitis B surface antibody (HBsAb) and anti-hepatitis core antibody (HBeAb) are commonly tested for in patients with clinical features of hepatitis, or may be followed in those at risk for acquiring hepatitis B. HBsAb can occur in persons immunized against hepatitis B, whereas HBeAb should only be present in those infected with hepatitis B who can make antibodies (thus, HBeAb may not be present in those with immunodeficiencies, and may not be expected to occur). If a patient has been tested previously, has been found to be negative for these antibodies and then receives IG replacement therapy, they are likely to test positive for HBsAb (as a result of plasma donors who have been immunized for hepatitis B). However, they should not test positive to HBeAb since donors positive for this are excluded from donating plasma. Again, patients who cannot make antibodies are not expected to make HBeAb. There have been numerous patients who have undergone further extensive testing for hepatitis B as a consequence of previously testing negatively and then testing positive, causing unneeded psychological stress. But, NAT can detect whether hepatitis B DNA is present, thus avoiding additional unnecessary testing.

Even though NAT is very sensitive for detecting the nucleic acid of microorganisms, detection is limited. For instance, if someone has a virus present that has become quiescent (dormant), it is possible it will not be detected. When NAT is negative but symptoms persist, it should be performed at least three times on separate specimens to better confirm that microorganisms are not present.

In addition, there are significant drawbacks to NAT, which we will discuss in the next issue.

**TERRY O. HARVILLE**, MD, PhD, is medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences and a consultant for immunodeficiencies, autoimmunities and transplantation.
“I’LL DO THAT when I feel better.”

“When my doctors get my symptoms under control, I’ll start making changes.”

Do you ever feel as if you’re waiting for your pain, anxiety or depression to subside so your life can begin? Many of us approach life that way. It makes sense. How can we exercise when we’re physically exhausted all the time? How can we go out with friends and be good company when we’re depressed? How can we get any work done when we haven’t had a good night’s sleep in weeks? Thinking this way, living our lives this way, is common. It’s human. And, sometimes, it means we wind up spending years waiting to start living our lives.

So, what’s the alternative? Start taking steps toward living your life while those feelings or physical sensations are there. The depression, anxiety or pain may still be there, but you can still do something that matters to you today.

How do you motivate yourself to do this? One strategy is to focus on your values. What matters to you? Maybe you are thinking about domains of your life such as family, friends, work or health. Maybe you are thinking about qualities such as honesty, loyalty, kindness or respect. Whatever your values are, they are yours alone. You do not need to defend or explain them.

Here is an activity you can try to get in touch with your values: Imagine it is 20 years from now, and you are at a birthday party in your honor. Close your eyes, and visualize it. Where is it being held? Is it outside or inside? How is the space decorated? Is it a small gathering or a large party? How is everyone dressed? Formally or casually? What food is being served? Anyone you want can be there, living or dead. People can be the same age they are now and can look the same as they do now. Also, what do you see? What sounds do you hear? What smells do you smell? Move around the party and greet people, or have them come over to you.

Now, imagine that the people who are most important to you start giving toasts in your honor. These people might include partners, parents, children, friends, coworkers or others close to you. Different people may give different toasts, focusing on different aspects of who you are. What do you want these people to say about you 20 years from now? Not what they say, but what do you want them to say? Take a moment and write it down. These are your values. This is what matters to you.

Knowing what our values are helps to empower us, not make us feel bad about ourselves. Values guide the direction we want to move in and help us stay focused. Values can provide a stable anchor when we don’t know what to do in a situation or when our emotions are all over the place. Values help us decide what matters in the moment. Most important, values are to be lived every day. We will never accomplish our values and be done with them. For example, if you value relationships, then you can choose to work on your relationships every day. We are never done working on them.

So let’s circle back to the challenge I put forth at the beginning of this column: Start taking steps toward living your life while those feelings or physical sensations are there. Then, let’s look at the question I posed: How do you motivate yourself to do this?

Try focusing on your values. What are your top three values? Now, if I were a fly on the wall, what would I see you doing that would let me know those three values are important to you? How would I know based on your actions? For example, if family is important to you, I might see you making a phone call or texting a family member who is important to you. I might see you sending an email to a family member you haven’t spoken to in a while. I might see you show up at a family event even though you’re not feeling well, because it is important to the host that you be there.

The point is you want to identify a specific behavior you can do that is consistent with that value. (Try not to choose something you will not do or that you will do less of such as “I will not yell at my kids when I get frustrated with them.”) Instead, choose something positive you can do more of just once.

That is how we begin living our lives while experiencing physical and/or emotional pain. We can choose each day to do one thing that is consistent with our values, instead of waiting to do that when the pain goes away. It may be one of the hardest things to motivate yourself to do. And yet, you will no longer be waiting to start living your life. You will be living it.

ERIKA LAWRENCE, PhD, LCP, is director of translational science at The Family Institute at Northwestern University, Evanston, Ill.
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Administration of Immune Globulin: Where We Are Today

By Michelle Greer, RN

METHODS FOR administration of immune globulin (IG) therapy have expanded over the last decade. Beginning with intramuscular IG (IMIG) therapy of years past, it evolved to intravenous IG (IVIG) and then subcutaneous IG (SCIG) therapy, with the latter utilizing the pump or push method today. How the route of administration is determined primarily includes diagnosis, dose, patient and physician preference, site of administration and patient response.

IMIG Therapy

IMIG was initially prescribed for post-exposure to hepatitis A, measles, varicella and rubella. Today, some physicians also prescribe it for off-label (non-U.S. Food and Drug Administration [FDA]-approved) uses.

IVIG Therapy

IVIG is commonly prescribed for FDA-approved indications, including immune deficiency disorders and autoimmune diseases. It is also effectively administered off-label for a variety of diseases. Because IVIG is administered directly into a vein, the patient experiences an initial spike in immunoglobulin (IgG) levels, known as the peak trough level. After three to four weeks, this level drops, returning IgG levels to their baseline, which requires readministration of IVIG. When IgG levels drop to their baseline, symptoms of the disease typically return and, in some cases, dosing and/or frequency may need to be adjusted or another route of administration may need to be considered to lessen these symptoms. Additionally, initial peak IgG trough levels might result in systemic side effects such as headache, nausea, vomiting and blood pressure changes. Because of the way IVIG is infused and the potential for these systemic side effects, these infusions are almost always administered and monitored by a trained nurse.

SCIG Therapy

SCIG is administered into the subcutaneous layer of the skin. The most common sites are the abdomen and tops of the thighs, while less-common sites are the backs of the upper arms. Because SCIG is administered once or twice weekly rather than every three to four weeks, this results in an IG steady state rather than high and low trough levels, which contributes to better disease symptom control. Indeed, in one recent study, pooled analyses showed the incidence of infection increases as IgG levels falls toward the end of each three- or four-week IVIG dosing cycle.1

With SCIG, the potential and overall incidence of systemic side effects is greatly reduced because the absorption

<table>
<thead>
<tr>
<th>Visit number</th>
<th>Rapid push</th>
<th>Pump</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Visit 1</strong></td>
<td></td>
<td></td>
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<tr>
<td>Patients with ≥ 1 adverse event</td>
<td>n = 72</td>
<td>n = 29</td>
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<tr>
<td>Local reaction</td>
<td>22 (31%)</td>
<td>9 (31%)</td>
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<tr>
<td>Headache</td>
<td>20 (28%)</td>
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<tr>
<td>Gastrointestinal (nausea, vomiting, diarrhea)</td>
<td>1 (1%)</td>
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</tr>
<tr>
<td>Fever</td>
<td>1 (1%)</td>
<td>0</td>
</tr>
<tr>
<td>Rash</td>
<td>0</td>
<td>1 (3%)</td>
</tr>
<tr>
<td>Other</td>
<td>2 (3%)</td>
<td>0</td>
</tr>
<tr>
<td><strong>Visit 2</strong></td>
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<td></td>
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<tr>
<td>Patients with ≥ 1 adverse event</td>
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<td>n = 23</td>
</tr>
<tr>
<td>Local reaction</td>
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<td>5 (22%)</td>
</tr>
<tr>
<td>Headache</td>
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<tr>
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<tr>
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</table>

Table. Adverse Events Following Subcutaneous Immune Globulin (SCIG) Administration by Pump or Rapid Push Reported at Three Visits 1
of IG is much slower. Patients can also enjoy flexibility with their infusions and become independent and self-administer without the oversight of a healthcare practitioner.

Facilitated SCIG (fSCIG) combines traditional SCIG with hyaluronidase. This requires the additional step of administering hyaluronidase into the subcutaneous tissue prior to administering SCIG, which allows larger volumes of IG to be dispersed into the subcutaneous space, resulting in less-frequent dosing (usually monthly). Takeda’s HYQVIA is currently the only available FDA-approved product for fSCIG administration, and it is primarily used to treat immune deficiency diseases.

SCIG is volume-dependent and, therefore, dose-dependent since the subcutaneous space can only accommodate so much fluid at one time. While SCIG products were originally studied and approved only to treat primary immune deficiencies with lower dosing, CSL Behring’s Hizentra was recently approved to treat chronic inflammatory demyelinating polyneuropathy, an acquired immune-mediated inflammatory disorder of the peripheral nervous system.

SCIG: Pump vs. Rapid Push

SCIG was originally administered only via a pump that can be tailored for frequency, number of subcutaneous sites and length of infusion to accommodate the dose, total volume and patient preference. However, a newer administration method known as rapid push is now an option. With rapid push, instead of infusing into subcutaneous sites for an hour or longer, the IG can be administered subcutaneously more quickly with a needle, avoiding the longer setup and total infusion time. Rapid push does, however, require more frequent infusions.

Rapid push infusions typically last anywhere from five minutes to 20 minutes. The process includes locating a subcutaneous site, drawing the IG into a small syringe, connecting the small tubing on the subcutaneous needle and, depending on the volume, administering the IG over a predetermined time. The push rate can be increased or decreased depending on how the volume is tolerated. Since site reactions are the major side effect of SCIG, the goal is to infuse at a speed that results in minimal to no irritation.

Rapid push infusions can be administered as often as daily. It’s up to the physician and patient to determine how often is best to maintain an adequate response to treatment. A pharmacist and/or nurse can also provide input about the length of infusion for each administration, as well as other equipment and site selection to maximize success.

To date, rapid push has only been studied in immune deficiency patients. In one retrospective study, results suggest primary immune deficiency patients prefer SCIG administered via rapid push than with a conventional pump. And, while serum IgG levels were comparable between methods, safety was similar if not slightly better with rapid push. The researchers concluded rapid push offers the potential for even greater convenience than the pump infusion technique, but they recommend the results be confirmed in prospective studies.3

Another study comparing routes of infusion found rapid push provides the same steady state levels of IgG as SCIG infused with a pump, but the side effect profile in some cases was better. In addition, mean serum IgG levels did not differ significantly between administration methods of SCIG. According to the authors, these results suggest rapid push is an effective administration method of SCIG delivery in immune deficiency patients and presents a valid alternative to pump administration.3

Some patients prefer to infuse less frequently and don’t mind the pre-infusion setup and use of a pump. However, others may want to infuse more frequently, but tailor infusions to their schedule, making rapid push infusions more preferable. For instance, some people may prefer to infuse “on the 5s” — the fifth, 10th, 15th, 20th, 25th and 30th days of the month, whereas others may prefer a Monday-Wednesday-Friday schedule. The schedule should be determined by what is clinically sound combined with what fits into a patient’s lifestyle.

Matching Options to the Patient

Many administration options are available to people who require IG replacement therapy. Patients and their physicians should select the administration option that best suits their lifestyle, while providing them the maximum clinical benefit.

MICHICLE GREER, RN, is senior vice president of sales for NuFactor Specialty Pharmacy.

References
ADMA Biologics ASCENIV (immune globulin intravenous, human [IgA 10% liquid]), formerly referred to as RI-002, has been approved by the U.S. Food and Drug Administration (FDA) to treat primary humoral immunodeficiency disease (PI) in adults and adolescents 12 years to 17 years old. The company anticipates having the product available for commercial launch during the second half of 2019.

Approval was based on a Phase III clinical study that enrolled 59 patients with PI at nine sites across the U.S. in which study patients received regular infusions of ASCENIV over the course of one year. The trial’s primary endpoint evaluated the rate of serious bacterial infections (SBIs), and secondary endpoints included time to first SBI and to first serious infection, days on antibiotics, days off school or work due to infections, all confirmed infections of any kind and hospitalizations due to infection. There were no SBIs during the 12-month study period. The approved labeling will include a boxed warning about potential thrombosis and renal dysfunction or failure, as well as the most common adverse events observed in the pivotal study, which were headache, sinusitis, diarrhea, gastroenteritis viral, nasopharyngitis, upper respiratory tract infection, bronchitis and nausea.

“We are excited about this significant achievement in receiving FDA approval for ASCENIV, a novel, patented IVIG product that we feel is a necessary addition to existing available therapies for patients who suffer from PI. We hope availability of ASCENIV will help ameliorate a portion of the current shortages facing U.S. IVIG supply,” stated Adam Grossman, president and CEO of ADMA Biologics. “There are approximately 250,000 PI patients diagnosed and living in the U.S., and we believe there is an opportunity to treat meaningful segments of this patient population with ASCENIV. As previously disclosed, ASCENIV is manufactured using our unique, patented plasma donor screening methodology and tailored plasma pooling design, which blends normal source plasma and plasma from donors tested using our proprietary microneutralization assay. Going forward, we believe this FDA approval better positions ADMA to further its mission to evaluate ASCENIV in immune-compromised patients infected with or at risk for respiratory syncytial virus (RSV) infection. We look forward to working with the FDA and the immunology and infectious disease community on developing a clinical investigation to evaluate use of ASCENIV in this patient population in the near future.”


Medicines
New IVIG 10% Approved by FDA to Treat PI

A recent study has found dysfunction of B cells is the cause of interstitial lung disease (ILD) in common variable immunodeficiency (CVID) patients. In this largest study conducted for treatment of CVID ILD, researchers examined 73 CVID patients and found their levels of immunoglobulin (Ig) M in the blood increased when their ILD worsened, which reflected the extent of IgM production locally by B cells in the lungs. They then found by depleting B cells with rituximab (Rituxan), they could effectively treat CVID ILD. In addition, they identified a protein that activates B cells known as B cell activating factor that contributes to lung disease recurrence in CVID, which they believe may serve as an additional therapeutic target in the future. The ability to identify elevation of IgM as a marker of CVID ILD progression, say the researchers, allows physicians to know precisely when their patients need treatment without waiting for ILD to decline.

“Because we are able to treat these patients using a precision medicine approach, we can spare them the side effects of more broadly immunosuppressive therapies that have been previously tried in these patients with mixed results,” explained Paul J. Maglione, MD, PhD, assistant professor of medicine at Boston University School of Medicine. Dr. Maglione hopes this study will lead to safer and more potent treatment of lung disease in CVID.

Research
Researchers Find Cause and Treatment for CVID Lung Disease

Research

Warning Signs of PI Differ Among Adult and Pediatric Patients

A study has found using the Jeffrey Modell Foundation (JM F) warning signs for immunodeficiency is not as effective for adults as it is in children. In the study, researchers performed a retrospective chart review in a two-center North American cohort of patients with primary immunodeficiency disease (PI). Charts of 137 pediatric and 400 adult patients with PI were evaluated for the presence of JM F warning signs and, compared to controls, with normal preliminary biochemical immune evaluation. They found fewer than 45 percent of adults with PI presented with two or more of the warning signs, compared with 64 percent of children. The warning signs found in a significantly increased proportion compared to controls differed for pediatric PI patients (recurrent pneumonia, failure to thrive, need for IV antibiotics, serious bacterial infection and recurrent otitis media) versus adult PI patients (recurrent otitis media, recurrent sinusitis, diarrhea with weight loss and recurrent viral infection). When evaluating additional criteria, there was slightly improved diagnostic accuracy of the warning signs when autoimmunity was present in the pediatric PI cohort. Adult PI patients demonstrated atopy more frequently than controls, while atopy (pre-disposition toward developing certain allergic hypersensitivity reactions) was found to have a negative association with the presence of PI in pediatric patients. Also, no improvement in diagnostic accuracy of the warning signs was found with the addition of allergic disease, autoimmunity or malignant and benign proliferative disease in the adult PI cohort.

Researchers suggest physicians should be educated about differing presentations of possible immunodeficiency between age groups, as well as expansion of the warning signs to include noninfectious comorbidities such as autoimmunity in pediatric patients.


Research

New Study Will Evaluate hsIgG vs. IVIG in ITP Patients

In January, Momenta Pharmaceuticals performed dosing of the first subject in the Phase I/II clinical trial of M254, hypersialylated immunoglobulin G (hsIgG). The four-part study will evaluate the safety, efficacy, pharmacokinetics and pharmacodynamics of intravenous (IV) M254 in approximately 65 subjects, including healthy volunteers and patients with immune thrombocytopenic purpura (ITP). Parts A and B are double-blind, placebo-controlled, single-ascending dose cohort studies in healthy volunteers and ITP patients, respectively. In Part C, ITP patients will receive M254 or IV immune globulin (IVIG) in a crossover dosing design, while in Part D, ITP patients will receive multiple doses of M254. The primary efficacy endpoint is an assessment of platelet response.

ITP is a rare autoimmune disease that leads to excessive bruising and bleeding, which can affect children and adults, and can be acute or chronic. M254 is a hypersialylated human immunoglobulin G engineered with significantly enhanced tetra-sialylation. In preclinical studies of ITP and other inflammatory diseases, M254 has shown an increase in potency of up to 10 times that of conventional IVIG.

“We believe M254 has the potential to be a significantly better option for patients than conventional IVIG. Our aim for this study is to show clinically what we have observed in extensive preclinical models, which is that hypersialylated IgG is substantially more potent than intravenous immunoglobulin G (IVIG) in ITP and other inflammatory disorders,” said Santiago Arroyo, MD, PhD, senior vice president of development and chief medical officer of Momenta Pharmaceuticals. “We look forward to obtaining initial clinical data in the first half of 2020.”

The U.S. Food and Drug Administration (FDA) has approved Firdapse (amifampridine) tablets to treat adults with Lambert-Eaton myasthenic syndrome (LEMS). The drug is the first agency-sanctioned treatment for the autoimmune disease.

Firdapse was assessed for its safety and effectiveness in clinical trials of 64 adults that measured the Quantitative Myasthenia Gravis score (a 13-item physician-rated categorical scale assessing muscle weakness) and the Subject Global Impression (a seven-point scale on which patients rated their overall impression of the effects of the study treatment on their physical well-being).

For both measures, patients receiving Firdapse experienced a greater benefit than those on placebo. The drug’s most common side effects included paresthesia, upper-respiratory tract infection, abdominal pain, nausea, diarrhea, headache and elevated liver enzymes.

In people with LEMS, the body’s own immune system attacks the neuromuscular junction (the connection between nerves and muscles) and disrupts the ability of nerve cells to send signals to muscle cells. LEMS may be associated with other autoimmune diseases, but more commonly occurs in patients with cancer such as small cell lung cancer, where its onset precedes or coincides with the diagnosis of cancer. The prevalence of LEMS is estimated to be three per million individuals worldwide.

“There has been a long-standing need for a treatment for this rare disorder,” said Billy Dunn, MD, director of the Division of Neurology Products in FDA’s Center for Drug Evaluation and Research. “Patients with LEMS have significant weakness and fatigue that can often cause great difficulties with daily activities.”

A number of case reports have documented the efficacy of intravenous immune globulin (IVIG) in children with autoimmune encephalopathy (AIE), and several studies have described improvements in autism-related characteristics following IVIG therapy. Now, in the largest case series of children with autism spectrum disorder (ASD) treated with IVIG, researchers identified brain-targeted autoantibodies in children with ASD.

In the study, 82 children were screened for specific blood autoantibodies or other markers associated with AIE of which 49 of those whose autism-related behavioral and other symptoms were recommended a trial of IVIG.

Thirty-one children received varying monthly doses of IVIG, with the majority receiving 2.0 grams per kilogram of body weight or more. The majority of parents (90 percent) reported some improvement, with 71 percent reporting improvements in two or more symptoms. Statistically significant improvement was documented for the subset of patients whose caregivers completed the Aberrant Behavior Checklist (ABC) and/or Social Responsiveness Scale (SRS) questionnaires. The antidopamine D2L receptor antibody, anti-tubulin antibody and ratio of antidopamine D2L to D1 receptor antibodies were related to changes in the ABC. Changes in the Cunningham autoantibody panel predicted SRS, ABC and parent survey-based treatment responses with good accuracy. Adverse effects of IVIG administration were common (62 percent) but mostly limited to the treatment period; only two patients (6 percent) discontinued IVIG because of adverse effects.

The investigators believe findings from this open-label case series provides evidence supporting a neuroimmune subgroup in patients with ASD.

FDA Approves Firdapse for Rare Autoimmune Disorder

Improved Social and Behavioral Scores Following IVIG Therapy Suggest a Neuroimmune Etiology in Some Children with Autism

Researchers have found Privigen (intravenous immune globulin; IVIG) is a safe and effective therapy for pretreated chronic inflammatory demyelinating polyneuropathy (CIDP) patients. In the study, the researchers compared results from the PRIMA (a prospective, open-label, single-arm study of IVIG in Ig-naive or IVIG-pretreated subjects) and PATH (a double-blind, randomized study, including an open-label, single-arm IVIG phase in IVIG-pretreated subjects) studies both separately and together. Efficacy assessments included change in adjusted Inflammatory Neuropathy Cause and Treatment (INCAT) score, grip strength and Medical Research Council (MRC) sum score. Adverse drug reactions (ADRs) and ADRs/infusion were also recorded.

Results showed the adjusted INCAT response rate was 60.7 percent in all PRIMA subjects at week 25 (76.9 percent in IVIG-pretreated subjects) and 72.9 percent in PATH patients. In the pooled cohort, the INCAT response rate was 71.9 percent and median time to INCAT improvement was 4.3 weeks. No clear demographic differences were noticed between early (responding before week seven) and late responders. In the pooled cohort, median change from baseline to last observation was -1.0 points for the INCAT score; +8.0 kPa (kilopascal) for maximum grip strength; and +3.0 points for MRC sum score. In the pooled cohort, 271 ADRs were reported in 105 subjects (44.7 percent), a rate of about 0.144 ADRs per infusion.


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Raising Kids While Coping with a Chronic Illness

Juggling family demands while navigating the unpredictable challenges of a chronic illness can feel overwhelming. The good news is support is available, and parents don’t have to do it alone.

By Trudie Mitschang

SHORTLY AFTER CATHY Roll and her husband, David, adopted their young daughters, they got news that would forever alter their family dynamic: Cathy was diagnosed with multiple sclerosis (MS). “I had been struggling with pain and fatigue even before we went to Ukraine for the adoption,” she recalls, “but after we returned, it was just relentless. At first, I thought, ‘Well, maybe this is what motherhood is all about.’ But, chasing around after two children made me so incredibly tired, like I had fallen into a dark hole.”

Cathy’s experience mirrors that of countless other parents who suddenly find themselves faced with the unexpected challenges presented by chronic disease, the term used to describe any long-term illness that can last or recur over a lifetime. Such conditions affect tens of millions of Americans, many of them parents of young children, and they include diagnoses like MS and other autoimmune diseases, primary immune deficiencies, as well as diabetes and depression. Regardless of the specific diagnosis, parents...
tasked with managing the daily demands of their medical care may be at a loss when it comes to learning how to also care for their children.

“Handling chronic illness is about learning to live in balance,” says Rosalind Dorlen, PsyD, a psychologist at Overlook Hospital in Summit, N.J., who specializes in treating the depression and anxiety that often accompany long-term health problems. “You can’t dwell on questions like, ‘Why is this happening to me?’ or ‘What if it gets worse?’ But you do have to be constantly conscious of your health status, and take the time to rest, exercise and have fun. It’s important to focus on feeling well and to maintain a positive outlook.”

Learning to Live with Limitations

Parents who have been diagnosed with chronic illness sometimes battle symptoms that can severely limit their ability to perform the daily physical tasks involved with raising kids. Simple things like picking up a crying child, preparing school lunches or playing a backyard game of catch are just some of the everyday activities that can become difficult or even impossible. Accompanying feelings of guilt and the concern about disappointing their children only adds to the emotional pressure.

“In the last year, I have had three surgeries, two other extended hospital stays, one ambulance ride, five emergency room visits and countless doctor visits, illnesses, injuries and bad health days. These keep me from doing my job as a college professor, from hanging out with friends, from family events and, most importantly, from taking good care of my kids all of the time,” says J. Ann Marie, a mother of three who was diagnosed with a primary immune deficiency at age 35. “Oh, I take care of them with every ounce of energy I can manage, but sometimes I physically cannot move. Those days, those days hurt me inside. I have to hear, ‘Mommy is sick again?’”

Mental health professionals stress that as difficult as it may be, it’s vital for parents to remember that having a chronic illness isn’t their fault. Postponing or missing events and activities will be inevitable because of the unpredictability of symptoms. The truth is, parents can’t protect their children from disappointment, but it is possible to ease their feelings of being let down. Parents can let their kids know how proud they are of their accomplishments and find creative ways to share in them. For example, if they have to miss an athletic event, another family member can record the event, and they can set aside time to view the recorded event together as a family. The key, say experts, is open and honest communication and a willingness to address children’s fears, disappointments and concerns.

In a recent study published in the peer-reviewed Journal of Nursing, researchers noted that with the obvious exception of infants, children share a need to understand the cause of a parent’s illness. They want to know things like whether they are responsible, whether it’s contagious and who will take care of them. The study stressed that in order for communication to be effective, it must be tailored to the children’s age and developmental level to help them develop needed coping skills.

Additional methods for helping kids develop coping skills include establishing a fixed family schedule and routine as much as possible, and including a schedule for regular sleep times, mealtimes, quiet time and activities such a family movie or game night. Although chronic illness is a life-altering event for a family, it is possible to turn it into an opportunity to strengthen family bonds and create resiliency. Parents can set the emotional tone of the household and help their children deal with the uncertainty of illness by being honest, setting realistic expectations and maintaining a positive attitude.

Talking It Out

One of the most difficult skills facing anyone diagnosed with a chronic illness is learning to talk about it with others. This challenge is compounded when the proposed dialog is between parents and their children. Yet, experts agree parents shouldn’t try to shield their children by hiding the illness,
Practical Tips for Talking to Your Kids About Your Illness

- Set aside as much uninterrupted time as possible. Children react in different ways, and you want ample time to deal with whatever emotions may surface. After dinner is a good time, since they are often calmer after eating and are winding down for the evening.

- Children pick up on your energy. If possible, wait until you have had time to digest and process the news so you can remain calm.

- Speak in terms they can understand. Try not to interject too much medical terminology. You can also use this time to introduce and explain some terms they may be hearing in relation to your illness.

- Allow them to ask questions. Answer with age-appropriate responses, staying focused on the specific question at hand. Including more details than necessary can be overwhelming for kids.

- Ask them questions. How do you think this will change things at home? At school? Ask for their suggestions about how to make things easier so they feel like they’re part of the conversation and the process. Inclusion is important and helps keep dialog flowing.

- Don’t hide your feelings. While you certainly don’t want to frighten your children, you do want them to know there are times when you may feel sad or particularly tired, and there are times you may even cry. Let them comfort you. It’s important for them to feel they are active participants in your life, and nothing warms a child’s heart more than knowing he or she has helped you feel better.

Practical Tips for Talking to Your Kids About Your Illness

because it is impossible to keep such a secret over time. Young children are highly perceptive and will begin to sense when something is wrong, while older children may overhear conversations or notice symptoms, and jump to the wrong conclusions.

While some parents worry that talking with their children may cause fear, honest communication actually breeds security because it builds trust. Sharing the challenge with children in age-appropriate ways has the potential to help them become more emotionally competent. But, careful planning is needed; the information must be communicated to them on their developmental level for them to properly process it.

According to Kathleen McCue, MA, LSW, child life specialist at the Cleveland Clinic Foundation, older children should be told the name and symptoms of the disease. They should be asked what they already know or what they have heard, and any wrong information they may have should be corrected. Younger children may need reassurance that their parent’s illness is not a punishment for their bad behavior, it is not a “monster that comes to get them” and it is not contagious. Most important, children of all ages need to know they will still be loved and given the care they need.1

Helping Kids Cope: A Guide for the Ages

The way children respond when a parent is chronically ill is influenced by many factors, including their own developmental stage. Identifying age-appropriate ways to communicate and relate as a family can ease stress for both parents and their children.

Infants. Infants whose parent has a chronic illness may experience significant changes in their routine, which in turn can cause them to become agitated or exhibit difficulty eating and sleeping. Stress may also make them more susceptible to colds or indigestion. What parents can do:

- Provide infants with extra physical contact and attention.
- Maintain a regular routine for physical needs, including feedings, sleep schedules, walks and playtime.

Toddlers. Very young children can sense feelings of frustration and fear when an illness is present, even though they may not understand what is happening. What parents can do:

- Talk to them using simple, honest words about the illness.
- Allow them to ask questions over and over.
- Let the children make choices about exposure to medical treatments such as being present during an infusion.

School-age children. The family is the basis of security for school-age children. Home is the main environment where they learn how to express their feelings. What parents can do:

- Continue to answer questions honestly and as many times as asked.
- Offer to include the children in seeing some of the medical aspects of the illness (always ask whether they want to be involved and how).
- Read books together that share stories about the illness.
• Consider peer support groups.

*Tweens.* Tweens alternate between their family and peer relationships for support. Their emotions are heightened by the onset of puberty. What parents can do:
• Expect children to experience emotional ups and downs. Assure them you are there for them.
• Understand their emotions may manifest in feeling physically ill (i.e., headaches, stomachaches, colds).
• Look for age-appropriate peer support groups, and encourage peer relationships and involvement.
• Answer questions honestly and thoroughly.

*Teens and young adults.* The primary support for teenagers is their peer group. While the family remains a significant resource, it is not uncommon for teenagers to act ambivalent about depending on their family. Having a parent diagnosed with a chronic illness can present a conflict for teens striving to assert independence. What parents can do:
• Make time for teens, even if they seem preoccupied with other priorities.
• Expect that teens may exaggerate the importance of certain aspects of this experience (strong feelings may seem out of proportion, but they must be allowed and accepted).
• Continue to encourage questions and provide honest answers.
• Encourage peer group support.

**Finding Community: Parents Are Not Alone**

It can be challenging for parents to help their children with their feelings when simultaneously managing their own emotions. Therefore, it is crucial for parents to find an outlet for their own evolving feelings about their illness. Just as their children look to them for help in acknowledging and processing their emotions, parents also need trusted peers to help them develop coping skills. Many of the national organizations for invisible and chronic illness offer peer support groups and hotlines. Social media groups are also popular sources of support and encouragement. "Research shows that understanding partners and peer support from similarly situated parents are particularly helpful in navigating the challenges of parenting while chronically ill," says Katie Willard Virant, MSW, JD, LCSW, a psychotherapist practicing in St. Louis, Mo. “Friends, relatives and therapists also can help us work through our own feelings and provide the emotional fortitude to parent well in difficult circumstances.”

Without question, every parent faces ups and downs while managing the daunting task of child-rearing, and for parents with chronic illness, it can be tempting to give in to hopelessness. But, by gleaning advice from others who have walked in their shoes and learning to remain optimistic when the odds seemed stacked against them, it is possible to find hope for better days ahead. “On days like today, where I was just released from the hospital after a long stay and I cannot go to track and field day, or the pre-k play tonight, or my son’s Boy Scout Derby, I try so hard to be thankful that I am still here, and I get to hug them when they come home and run into my room (and ask if I’m feeling better),” says J. Ann Marie. “They are too little to know this will always be this way — some good days, a lot bad, but that is fine. I want them to be too little as long as they can. I answer, ‘Yep! I am feeling better.’ Their faces light up, and they run off to play, and I take more medicine that makes me feel sick and try to rest some more and hope tomorrow I can get up and go with them.”

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

**References**

**IG Living**

**IG Living.com**
Organizing the Chronically Ill Chaos

This system of organizing medical data will help doctors and specialists understand a patient’s current state of care.

By Denna McGrew
ORGANIZED CHAOS is honestly the best I can hope for most days. By the numbers, my family is as complex as it gets since three of the four of us are chronically ill. Together, we have:

- 6 rare diseases
- 15 other major diagnoses
- 2 primary care physicians
- 14 specialists
- 8 to 12 monthly treatment days
- 4 to 6 doctor visits per month
- 10,000 miles travelled annually to see specialists

That’s a lot to manage for anyone! As such, I’ve learned a few things along the 17-year journey of our chronic life, including two main strategies for getting the best possible outcomes.

The first strategy, and most important, is to find the right specialist for your own disease. For our family, that means travel. I cannot overstate the importance of finding the doctor for whom your rare disease is of research interest. This “unicorn” of a specialist is one who has dedicated his or her career to learning about and treating your disease (even though this disease is one that most doctors need to Google).

Once you have that unicorn specialist on board, you must implement the second strategy: Arm him or her with your medical information in a way that makes it easy to understand and utilize. I’ve developed a system that works for my family and one that our doctors report finding useful. In fact, a couple of them have commented that my son wouldn’t be alive today without this system.

In the rare disease community, patients see many specialists who may be located all over the country. Busy doctors do not have the time to keep each other in the loop, so that arduous chore falls to the patient and/or caregiver (such as the patient’s parent). I think of it as being a little like football: The specialist is the coach and calls the plays, whereas the patient or caregiver is the quarterback, implementing and carrying out the plays determined by the coach. Rare diseases further complicate matters since there are multiple coaches, so it’s our job to make sure they have all the information so they can call the right plays.

My system involves keeping a single document in Microsoft Word that I update before every appointment and hand a hard copy to the doctor (Figure). Here are the basic components:

1) **Header:** In the header section of the document, include the patient’s name, date of birth and the date on which the document was created or most recently updated/revised, as well as the page number and number of pages. This header will automatically be present on every page, which helps if the pages get separated. If the person preparing the document is someone other than the patient, consider adding that information to the header as well. For example: Prepared by Denna McGrew, Jenna’s mother.

![Figure. Medical Information Sheet](image)

- **Patient Name**
  - Name:
  - Date of Birth:
  - Created/Revised:

- **Diagnoses**
  - Diagnosis
  - (Date)

- **Medications**
  - Prescription:
    - Drug:
    - Dosage:
    - Frequency:
  - PRN (as needed):
    - Drug:
    - Dosage:
    - Frequency:
  - OTC (over the counter):
    - Drug:
    - Dosage:
    - Frequency:
  - Recently tried medications:
    - Drug:
    - Dosage:
    - Frequency:
    - Reason stopped:

- **Allergies**
  - Drug and reaction:
  - Environmental allergies and reaction:
  - Food allergies and reaction:

- **Doctors**
  - Name, Specialty, Location, Phone/Fax/Email:

- **Major Symptoms**
  - Symptom:
  - Symptom:

- **Narrative** (reverse chronological order)
  - 0/0/0000
    - Summarize medical event in short form such as doctor visit, lab performed, medications prescribed and reason:
  - 0/0/0000
    - Summarize medical event in short form such as doctor visit, lab performed, medications prescribed and reason:

Developed by Denna McGrew; dennamcgrew.com
Important Safety Information

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

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

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

Hizentra is a prescription medicine used to treat:

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

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

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

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

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

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

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

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

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

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

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

Initial U.S. Approval: 2010

**BRIEF SUMMARY OF PRESCRIBING INFORMATION**

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

**WARNING: THROMBOSIS**

See full prescribing information for complete boxed warning.

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

**INDICATIONS AND USAGE**

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

**CONTRAINdications**

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

**WARNINGS AND PRECAUTIONS**

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

**ADVERSE REACTIONS**

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

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.
2) 

**Diagnoses:** In this section, all the patient’s diagnoses should be listed followed by the year in which the diagnosis was made. This section can be in reverse chronological order or in order of the severity of the diagnoses (my preference).

3) 

**Medications:** This section potentially contains four separate components. The first is prescriptions and should include all medications taken on a routine or scheduled basis. Next, list medications taken on an as-needed basis. Then, list all over-the-counter medications, vitamins and supplements. Finally, list any medications that have been tried and stopped, and indicate the reasons they were stopped and the dates. Each of these sections should include the same information about the medication: drug name, dosage, frequency and route of administration. For example: Armour Thyroid, 120 mg, one per day in the morning; Gammagard IVIG, 20 grams every two weeks via IV.

4) 

**Allergies:** For those with known allergies, this section is quite important. List the allergen and the reaction. Include drugs/medications, environmental allergens, foods, medical supplies such as latex or adhesives. Note the specific reaction the patient has when exposed to each individual allergen.

5) 

**Doctors:** List each doctor or specialist. Include their specialty, location and contact information such as address, phone and fax number, as well as their email if available. This facilitates specialists being able to contact one another should they choose to do so.

6) 

**Major symptoms:** Indicate the major presenting symptoms in this section. Include any problems that cause the patient difficulties in daily life such as severe fatigue, recurrent infections, syncope episodes and tachycardia.

7) 

**Narrative:** This is the historical section that will be developed over time. I started my children’s documents when they were born since they were both sick from birth and their narratives are quite complete. For myself, I began getting very ill in 2011, and was diagnosed with common variable immunodeficiency in 2013. To compile my narrative when I started it in 2012, I had to do some digging. I pulled out my insurance explanation of benefits statements and used them to reconstruct the journey over the previous couple of years. Thankfully, today that information is more easily accessible via the insurance company’s website.

This narrative should be a short summary of any major event that occurs, including doctor visits, labs, imaging and other tests, and episodes of illness or flares. Include the date and then a relevant and concise summary. Keep it short and with the most recent events at the top (reverse chronological order).

This document will become your “go-to” for every doctor visit and can guide the appointment, thus saving time and confusion on the part of the provider. Our specialists literally sit knee to knee with us and go through the narrative items that have transpired since the last visit. And, the medical assistants use the medication section to update that information in their systems. It’s a simple way to make sure everyone is on the same page.

To ensure I have these documents with me at all times, I save the latest version as a PDF file and keep it in the free online document sharing tool “Drop Box,” which I can access from my computer, phone or iPad. That way, if I’m away from home and need to have a conversation with a provider, I have my cheat sheet. I can quickly email the PDF document to a specialist upon request.

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Once you have that unicorn specialist on board, you must implement the second strategy:

Arm him or her with your medical information in a way that makes it easy to understand and utilize.

This chronic life is not easy, but developing a system to wrangle the chaos can take away some of the stress. As caregivers and patients, the more we put on paper, the less we try to keep in our heads, and that equals reduced stress. Spend some time on the front end developing your systems, and enjoy the fruits of that labor for years to come.

DENNA MCGREW is a rare disease warrior for herself and her two children and often writes while sitting on one of Florida’s beautiful Treasure Coast beaches.
Managing Chronic Pain with Medical Marijuana

Can medical marijuana help patients manage chronic pain? The jury is out, according to research, but it is an option for those unable to manage pain with other medications.

By Ronale Tucker Rhodes, MS

MEDICAL MARIJUANA, also known as medical cannabis, dates back to 2737 B.C., when the mystical Emperor Shen Neng of China prescribed marijuana tea for the treatment of gout, rheumatism, malaria and poor memory. The drug’s popularity as a medicine then spread throughout Asia, the Middle East and down the eastern coast of Africa, with ancient physicians prescribing marijuana for everything from pain relief to earache to childbirth.¹

In the U.S., medical marijuana use began in the 1850s to treat afflictions such as neuralgia, tetanus, typhus, cholera, rabies, anthrax, leprosy, tonsillitis, dysentery, insanity and excessive menstrual and uterine bleedings. However, there was a prohibition on the use of medical marijuana in 1937 with the passage of the Marijuana Tax Act. While marijuana is still classified as a Schedule I drug (a category of drugs not considered legitimate for medical use), in 1996, patients and advocates turned to states for access to marijuana for medicinal purposes. Since then, voter initiatives in a host of states have passed, allowing access to medical marijuana, despite federal law prohibiting its use.²

According to surveys in recent years, public approval of medical marijuana has remained above 77 percent since 2011. Medical marijuana patients seem to also be satisfied with the treatment they experience, with a majority reporting they would be highly likely to recommend it to friends or family for treatment.³

More importantly, at a time when chronic pain is the most common cause of long-term disability in the U.S. — affecting more people than cancer, heart disease and diabetes combined⁴ — a number of studies are exploring the medical properties of marijuana to manage chronic pain. And, many researchers, including those funded by the National Institutes of Health, are continuing to explore the possible uses of cannabinoids (chemicals comprising the marijuana plant) for medical treatment, including for chronic pain management.⁵
What Is Medical Marijuana

The term “medical marijuana” refers to using the whole unprocessed plant or the chemicals contained within it to alleviate symptoms of certain conditions or diseases. The marijuana plant is comprised of more than 100 cannabinoids (chemicals), each of which have different effects on the body. The two main cannabinoids used in its medicinal application are tetrahydrocannabinol (THC), the psychoactive compound in marijuana (i.e., the element that produces the high), and cannabidiol (CBD), the substance that does not produce any psychoactive effects (medical marijuana has a higher CBD content, so it doesn’t produce the euphoria associated with its recreational counterpart). 6

THC can increase appetite and reduce nausea. It may also decrease pain, inflammation (swelling and redness) and muscle control problems. CBD may be useful in reducing pain and inflammation, controlling epileptic seizures, and possibly even treating mental illness and addictions. 4

Doctors prescribe medical marijuana to treat muscle spasms caused by multiple sclerosis, nausea from cancer chemotherapy, poor appetite and weight loss caused by chronic illness such as HIV or nerve pain, seizure disorders and Crohn’s disease, among others. 7 In addition, the California Medical Association states marijuana may also be used to help treat AIDS, anorexia, arthritis, cachexia, cancer, chronic pain, glaucoma, migraine, persistent muscle spasms, seizures, severe nausea and any other chronic or persistent medical symptom that limits a person’s ability to conduct major activities in life or can cause serious harm if not relieved. 6

How Does Medical Marijuana Work?

THC is what causes people to feel high and also what gives cannabis some of its medicinal properties such as increased appetite. The human body produces endocannabinoids, its own natural version of cannabinoids. Studies show the endocannabinoid system (ECS) helps to regulate the body’s responses to a variety of stimuli. The body will produce endocannabinoids when needed, but sometimes the effect is very brief. Endocannabinoid receptors are found throughout the body, but are especially prominent in the brain. The cannabinoids in marijuana, like THC, bind to these receptors, producing various effects, some medicinal such as reducing pain or anxiety, but also the feeling of being high. 9

CBD influences the body to use its own endocannabinoids more effectively. According to one study, this is because CBD does very little to the ECS. Instead, it activates or inhibits other compounds in the ECS. For example, CBD stops the body from absorbing anandamide, a compound associated with regulating pain. So, increased levels of anandamide in the bloodstream may reduce the amount of pain a person feels. CBD may also limit inflammation in the brain and nervous system, which may benefit people experiencing pain, insomnia and certain immune-system responses. 10

Types Available

Medical marijuana comes in many forms: pills, oils, vaporized liquids, nasal sprays, food, dried leaves and buds, and plants. It can be smoked, vaporized (heated until active ingredients are released, but no smoke is formed), eaten (usually in the form of cookies or candy), taken as a liquid extract and rubbed onto the skin. 7, 11

There is a distinction between medical marijuana, however, and cannabis extracts that are also made from CBD. CBD extracts are derived from hemp, while CBD products classified as medical marijuana are extracted from marijuana — even though the CBD used to make extracts is the same compound used to make medical marijuana. The laws governing CBD started with the Controlled Substances Act of 1970, which labeled all varieties of the cannabis plant, hemp included, a Schedule I drug. But a “hemp amendment” (Section 7606, Legitimacy of Industrial Hemp Research) in the Farm Bill of 2013 (signed into law in 2014) changed those rules. Previously, hemp could be imported but not grown in the U.S. The amendment allows states to create pilot programs to research and cultivate hemp, which the legislation defines as a cannabis plant containing 0.3 percent or less of THC by weight. (Marijuana plants grown today contain THC levels hovering around 20 percent.) The bill also allows hemp products to be marketed to the public. 12
While not for the treatment of chronic pain, the U.S. Food and Drug Administration (FDA) has approved three drugs made from synthetic forms of ingredients found in marijuana to treat other conditions. These include dronabinol (Marinol, Syndros) and nabilone (Cesamet), which can be legally prescribed for the treatment of nausea and vomiting caused by chemotherapy when other treatments have failed. Dronabinol may also be used to treat anorexia associated with weight loss in people with AIDS. The third FDA-approved medicine is a CBD-based liquid medication called Epidiolex to treat two forms of severe childhood epilepsy: Dravet syndrome and Lennox-Gastaut syndrome.

In addition, FDA has approved two clinical research projects for new forms of marijuana ingredients. One clinical trial is testing the drug Sativex for breast cancer pain. Sativex is a combination of chemicals from the marijuana herb and is sprayed into the mouth. It is approved in more than 20 countries to treat muscle spasms from multiple sclerosis and cancer pain.

It’s important to note there are side effects of marijuana that usually don’t last long, including dizziness, drowsiness, short-term memory loss and euphoria. More serious side effects include severe anxiety and psychosis. In addition, only people 18 years and older can be prescribed medical marijuana. Further, its use is advised against in people with heart disease, pregnant women and people with a history of psychosis.

Where Can Medical Marijuana Be Purchased?
The FDA does not approve or recognize marijuana as a medicine. However, medical marijuana is legal in many states today. California became the first state to legalize it in 1996, and since then, 33 states have followed suit (Figure).
Medical marijuana laws, usage and prices vary from state to state. Not only do prices differ dramatically by state, there are also significant differences in prices between cities within the same state. In addition, the number of medical marijuana dispensaries in each state varies, as do the number of medical marijuana patients and caregivers. For example, while New Mexico had a legal medical marijuana patient rate of 25 per 1,000 residents in May 2018, Illinois had a rate of under two per 1,000 residents.9

To get medical marijuana, you need a written recommendation from a licensed doctor in states where it is legal. And, a person must have a condition that qualifies for medical marijuana use. Each state has its own list of qualifying conditions. Some states may also require an individual to get a medical marijuana ID card. With that card, medical marijuana can be purchased at a store called a dispensary.15

CBD extracts can be purchased over the counter at drug stores and through online retailers with no state or federal oversight.12

Studies Pertaining to Chronic Pain

Many studies show medical marijuana is effective in managing chronic pain; however, there are other studies showing no beneficial effect. Indeed, more than 45 studies have looked at marijuana and pain related to chronic diseases such as cancer, diabetes, fibromyalgia, multiple sclerosis, HIV, rheumatoid arthritis and spinal injuries. The studies have included smoked marijuana, along with herbal and man-made forms, the majority of which showed improvement in pain relief in comparison to a placebo or to other traditional pain medications. However, about a quarter of the studies showed no improvement.13

A study conducted in 2015 that reviewed the pharmacology, indications and laws related to medical marijuana use found marijuana for chronic pain, neuropathic pain and spasticity due to multiple sclerosis is supported by high-quality evidence. The study looked at six trials that included 325 patients examined for chronic pain, six trials that included 396 patients with neuropathic pain and 12 trials that included 1,600 patients with multiple sclerosis. While several of these trials had positive results, suggesting marijuana or cannabinoids may be effective for these indications, the authors noted there are also many that do not.16

In 2018, The New England Journal of Medicine published a case vignette of a 31-year-old woman with a long history of complex regional pain syndrome in her right leg and foot. After being treated with several opioids, regional and sympathetic nerve blocks, transcutaneous nerve stimulation, lidocaine and compounded salves, behavior modification, acupuncture and alendronate infusions, she continues to suffer what she calls excruciating pain. Two physicians were asked to comment, based on published literature, experience, recent guidelines and other sources of information, as appropriate, about which option they would recommend: 1) Prescribe medical marijuana, or 2) Discourage the use of medical marijuana.

Benjamin Caplan, MD, chose option one: “Cannabis added to her regimen could alleviate emotional distress and provide a more direct route to pain relief…. Cannabis has been shown to contribute, through cannabinoi-d-receptor and non–cannabinoid-receptor mechanisms, to anti-inflammatory and neuroprotective effects that may alleviate chronic pain. These effects appear to be dose-dependent with respect to synaptic transmission within the dorsal horn of the spinal cord, and inhibition of this communication may play a role in the development of chronic pain associated with local inflammation or nerve injury. For example, in a murine model of neuropathic pain, administration of cannabis significantly reduced allodynia [a rare type of pain, generally on the skin] in a dose-dependent manner. Furthermore, in a recent study of refractory pain, cannabis showed efficacy in patients for whom traditional treatment options had failed.”

Edgar Ross, MD, who chose option two, explained: “Literature reviews on the efficacy of medical marijuana are cautionary about effectiveness and side effects. Although the cannabinoid compounds are almost certainly safer than long-term opioid therapy, studies in humans that suggest efficacy are limited in quality and scope…. Since the mechanism underlying cannabis-associated psychosis is unknown, establishing a safe dose for routine use is difficult. Controversy about the addictive potential of cannabis and the risk of cannabis withdrawal syndrome is also ongoing.”

But, there have been studies that show medical marijuana is safe for chronic pain, at least among people with some experience using the drug. In one study, the first and largest of the long-term safety of medical marijuana use by patients with chronic pain in seven pain treatment centers across Canada, researchers followed 215 adult patients with chronic pain who used medical marijuana for one year and compared them with a control group of 216 chronic patients who didn’t use medical marijuana. Participants who used medical marijuana were provided leaf marijuana containing 12.5
cannabis-based medications “might be effective for chronic pain treatment, based on limited evidence, primarily for neuropathic pain patients.” And, a systematic review of the efficacy of cannabis in patients with neuropathic pain or multiple sclerosis or receiving chemotherapy concluded “there is incomplete evidence of the efficacy and safety of medical use of cannabis” and that “confidence in the estimate of the effect was again low or very low.”

Caution Is Advised

Clearly, some studies show there are may beneficial properties of medical marijuana, while others do not. And, currently, it is not an approved FDA medicine because FDA requires clinical trials in hundreds to thousands of human subjects to determine the benefits and risks of a possible medication. According to FDA, researchers have not conducted enough large-scale clinical trials that show the benefits of the marijuana plant (as opposed to its cannabinoid ingredients) outweigh its risks in patients.5

Still, many patients and studies can attest to medical marijuana’s management of chronic pain. However, it’s important to note that medical marijuana is not monitored like FDA-approved medicines. In fact, there are many unknowns, including its potential to cause cancer, its purity, potency or side effects. Considering these risk factors, only people who have a prescription from a doctor should use medical marijuana.7

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Ronale Tucker Rhodes, MS, is the editor of IG Living magazine.

Many patients and studies can attest to medical marijuana’s management of chronic pain.
Cannabidiol has been touted for its beneficial effects for managing chronic pain. But, with no large-scale clinical trials, it is still not an approved medicine by the U.S. Food and Drug Administration. Nevertheless, CBD products cultivated from hemp are allowed to be marketed to the public. But, because research results are mixed on whether CBD is beneficial for improving pain, it can be difficult for consumers to decide whether using it is the correct choice. For a professional perspective, we discussed the use of CBD with Lisa Allen-Khalil, MD, an internal medicine specialist in Brooksville, Fla., who is a proponent of CBD.

**IG Living:** What led you to recommending CBD to manage pain?

**Dr. Allen-Khalil:** Cannabis has been used medicinally for more than 3,000 years, beginning in the 19th century when Western medicine recognized its benefits for treating pain and spasms. Just recently, medical cannabis has become available in my state of Florida. In my internal medicine practice, I have seen patients with chronic pain issues who have not responded to conventional treatment modalities. Either the side effects of these modalities were unacceptable or the efficacy was poor.

**IG Living:** Is there any particular research you are familiar with that has led you to believe in CBD’s benefits for pain management?

**Dr. Allen-Khalil:** There isn’t a lot of research regarding the efficacy of using CBD for chronic pain. But, what is available is promising, and more research is being conducted. So, we will learn more as results become available. Studies published in the *Journal of American Medicine* and *Annals of Internal Medicine* give moderate evidence and low-quality evidence, respectively, supporting the use of cannabinoids for chronic pain, particularly neuropathic pain, spasticity, sleep problems, HIV-associated wasting, chemotherapy-related nausea and vomiting, and Tourette syndrome.

**IG Living:** Tell us about your personal experience with CBD?

**Dr. Allen-Khalil:** I have used CBD myself to treat pain as a result of sports-related injuries with fair results.

**IG Living:** What factors have you found determine which CBD products are the most effective for managing pain?

**Dr. Allen-Khalil:** Since CBD is currently not regulated by the U.S. Food and Drug Administration (FDA), finding a standardized formulation is very difficult. As such, the efficacy of the many preparations is hard to gauge. I have recommended to my patients who want to try CBD to talk with their pharmacist to find a reputable company that makes a quality CBD product. Having stated this, until CBD is regulated by FDA, people may not be entirely sure they are getting what they pay for.

**IG Living:** Have you prescribed CBD to your patients, and if so, what has been their experience?

**Dr. Allen-Khalil:** The few patients for whom I’ve recommended CBD have had mixed results. This is due to the lack of availability of standardized preparation and the uniqueness of each patient’s physiology.

**IG Living:** What advice would you give to chronic pain sufferers who are unsure if they should try CBD?

**Dr. Allen-Khalil:** I would advise them to obtain a consultation with a pain management specialist familiar with CBD who can make sure they are prescribed the proper formulation and dose of CBD to treat their specific conditions.

**References**


MANY PATIENTS WITH chronic illness also suffer from chronic pain. And, considering the epidemic of opioid addiction, there are, thankfully, nonnarcotic options available that are effective in helping to relieve pain and reduce inflammation. The two types of over-the-counter pain relievers are nonsteroidal anti-inflammatory drugs (NSAIDs) and acetaminophen (Tylenol).¹

NSAIDs are a class of medication used to treat acute or chronic pain. They can provide relief for mild to moderate pain and are often used to treat joint and muscle pain, and in some cases nerve pain. NSAIDs are nonaddictive and, in lower doses, can be obtained without a prescription. Currently, there are nearly two dozen categories of NSAIDs available that are marketed under 76 product names,² nine of which are the most common (see Common Over-the-Counter NSAIDs).³

While acetaminophen can be an effective pain reliever, it is commonly mistaken for an NSAID. Acetaminophen is classified as a miscellaneous analgesic that doesn’t fit into a particular class because it works in different ways to relieve pain compared to either an opioid or an NSAID.⁴ It is used to treat many of the same conditions as NSAIDs, but it doesn’t have an anti-inflammatory component, and it typically has a lower effect on pain.³ However, it is the pain reliever of choice for those who cannot tolerate NSAIDs or who must take a blood thinner.

How Do NSAIDs Work?

NSAIDs work much like corticosteroids (steroids) for reducing pain and inflammation without the associated side effects. They are commonly used to treat temporary conditions such as menstrual pain, muscle strains and sprains. However, they can also be used to treat a wide variety of chronic conditions such as osteoarthritis, rheumatoid arthritis, lupus, fibromyalgia, backaches and muscle pain, to name a few. Further, they are effective for treating headaches and fever.

NSAIDs work by blocking the production of prostaglandins that have several effects on the body, including causing pain and inflammation.⁶ Blocking these enzymes alters the sensation...
of pain.

Additionally, they work to reduce swelling often associated with certain types of pain. While all NSAIDs reduce pain and inflammation, their effects differ from person to person, which means one type may prove more effective in managing pain than another for an individual.

Generally, NSAIDs are taken orally in either pill or liquid form. If a patient is unable to take the oral form due to upset stomach, nausea or bleeding, topical NSAIDs may be considered. Topical NSAIDs, which come in gels, creams and patches that are applied directly to the skin, can be purchased either over the counter or by prescription. These are convenient because they can be applied directly to the area of inflammation, and they can be very effective for short-term relief. Further, they pose less risk for typical NSAID side effects.

Like any medication, it is important NSAIDs are used as directed. Oral and topical NSAIDs should not be combined without consulting a healthcare professional, and they shouldn’t be used in conjunction with bandages or heating pads. Application of a bandage or heat increases blood flow to the area, which can increase the absorption of the NSAID into the body and possibly lead to an overdose.

**Proper Use of NSAIDs**

Many people assume if they can purchase a medication without a prescription, it is safe. But, this can be a dangerous assumption. The term “over-the-counter” doesn’t necessarily mean the medicine is safe for everyone all the time, that it can be taken indefinitely or that it doesn’t have side effects.

Chronic illness patients using NSAIDs for pain are probably going to be taking them on a regular basis over a long period of time and, thus, the potential for serious side effects increases. Studies have shown NSAID use in older adults can exacerbate several chronic illnesses, including heart failure and hypertension, and they can interact with a number of other drugs. To avoid these complications in older patients, other pain-relieving options can be considered. For instance, analgesics such as acetaminophen, a short half-life NSAID (ibuprofen) or low-dose opioids have a lower risk of adverse reactions.

NSAIDs are intended for short-term use, and the length of time they should be taken depends upon the reason for their use. No over-the-counter NSAID should be taken continuously for more than three days for fever relief and 10 days for pain relief without consulting a healthcare professional. If NSAIDs must be taken for a longer period of time, patients can experience side effects. As such, patients should be carefully monitored by their physician so they can develop an appropriate treatment plan and change treatment if problems occur.

The most common side effect of long-term NSAID use is gastrointestinal (GI) problems. In fact, some 10 percent to 15 percent of patients are unable to tolerate NSAIDs because of side effects including but not limited to gas, bloating, heartburn, stomach pain, nausea and diarrhea. This is because NSAIDs reduce the amount of prostaglandins that help protect the lining of the stomach, which can cause the stomach acid to make small erosions in the stomach wall and duodenum, and cause discomfort.

While GI issues are the most common complaint, NSAIDs can also result in other common side effects, including headaches, dizziness, skin rashes, drowsiness, dry mouth and extreme weakness or fatigue. In addition, approximately 15 percent of those who use NSAIDs long-term develop peptic ulcers (ulcerations of the stomach or duodenum). Unfortunately, many times, patients have no early symptoms so they are unaware of the ulcers, which puts them at risk of developing dangerous ulcer complications such as bleeding or perforation.

NSAIDs work much like corticosteroids (steroids) for reducing pain and inflammation without the associated side effects.

Other serious side effects that may occur due to long-term NSAID use include:

- Allergic reactions such as hives, wheezing, difficulty breathing and swelling of the throat, tongue or lips
- Muscle cramps, numbness or tingling
- Rapid weight gain
- Black, bloody or tarry stools
- Bloody urine or vomit
- Decreased hearing or ringing in the ears (tinnitus)
- Jaundice (yellowing of the skin and the whites of the eyes)
A black box warning is the U.S. Food and Drug Administration’s (FDA) most serious warning for drugs and medical devices. Intended to inform users about serious or life-threatening risks, they are added when FDA has received research that confirms such a risk. In July 2015, FDA strengthened its box warning for NSAIDs after evidence showed people who used them had an increased risk of heart attack and stroke. The new warnings included the following information:

- The risk of heart attack or stroke can occur as early as the first weeks of using an NSAID. The risk may increase with longer use of the NSAID.
- The risk appears greater at higher doses.
- It was previously thought all NSAIDs may have a similar risk. Newer information makes it less clear the risk for heart attack or stroke is similar for all NSAIDs; however, this newer information is not sufficient for us to determine that the risk of any particular NSAID is definitely higher or lower than that of any other particular NSAID.
- NSAIDs can increase the risk of heart attack or stroke in patients with or without heart disease or risk factors for heart disease. Numerous studies support this finding, with varying estimates of how much the risk is increased, depending on the drugs and the doses studied.
- In general, patients with heart disease or risk factors for it have a greater likelihood of heart attack or stroke following NSAID use than patients without these risk factors because they have a higher risk at baseline.

Consult a Healthcare Professional

NSAIDs are a popular form of pain relief for millions of people. While they are easy to obtain, they are intended only for short-term use. Those who have underlying health conditions who need to take them for an extended period should consult their healthcare professional. It is important patients understand all the facts about the NSAIDs they are considering taking, since all carry risk of side effects.

ABBIE CORNETT is the patient advocate for IG Living magazine.

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

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

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

— Jenny Gardner

*Chronic Inspiration* can be purchased on iTunes, Amazon and Barnes and Noble.com.
THE NUMBER OF people developing or living with an autoimmune disease has increased noticeably over the past 30 years. The National Institutes of Health estimates approximately 24 million Americans have an autoimmune disorder, although organizations such as the American Autoimmune Related Diseases Association estimate the number to be much higher. The Benaroya Research Institute at Virginia Mason, a research institute in Seattle, Wash., dedicated to discovering causes and cures to eliminate autoimmune and immune system diseases, notes on its website that the incidence of many autoimmune diseases is increasing possibly due to changes in the environment that impact the immune system. This increases the likelihood that someone who is genetically predisposed to a particular autoimmune disorder will in fact develop it.

Can diet change the course of autoimmune disease? If we are what we eat, then linking diet with the development of autoimmune disorders makes a lot of sense. If only the relationship were that simple. What is known is diet can improve or worsen autoimmune symptoms, and autoimmune diseases often require diet changes to manage them. This article presents an overview of several research-based dietary approaches to autoimmune disease, along with practical tips from dietitians working with clients who have immune deficiency disorders.

**The Elimination Diet**

People with high levels of autoimmune antibodies are thought to have more intolerances to foods. In one study, cow’s milk, casein protein, wheat and egg white were

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**By Mindy Hermann, MBA, RDN**

Research shows many different dietary approaches are beneficial in preventing and treating autoimmune disorders, but some approaches still require additional study. 

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**Dietary Approaches to Autoimmune and Immune Deficiency Disorders**

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linked to the greatest antibody reaction, while no antibody reaction resulted from eating vegetables, fish or meat.²

The elimination diet is a challenging but important first step in identifying foods and ingredients that may be causing immune disease symptoms to worsen. In the first phase of this diet, a person cuts out all common food allergens from the diet: milk, eggs, nuts, peanuts, wheat, soy, fish, shellfish and beef. Careful label reading is essential because many ingredients contain common allergens! It may take several weeks for symptoms of allergy or intolerance to subside. And, because an elimination diet is highly restricted and nutritionally unbalanced, it should be closely supervised by a doctor and dietitian.

Once symptoms stabilize or disappear, allergenic foods are added back to the diet one by one, and tolerance is monitored for several days before trying a different allergenic food. People who are reintroducing foods with the help of a dietitian and physician must keep a log of foods and symptoms. Any food that appears to make symptoms worse stays on the list of foods to avoid.

The elimination diet can be an effective tool for directing disease management. A 2017 study showed improvement of symptoms and inflammation following a six-week elimination diet and five-week maintenance phase to help manage Crohn’s disease and ulcerative colitis in young adults.³

Fasting

Fasting long has been part of certain religious observances. In Judaism, fasts last a full 24 hours, while Islamic fasts for Ramadan require avoiding food and beverages during daylight hours. More recently, variations on fasting have become a popular strategy for weight loss. These modified fasts restrict eating and drinking to a set number of hours, say eight hours, during the day.

Early evidence suggests fasting may benefit the immune system. In a group of mice with a multiple sclerosis-like illness who were placed on a modified fast, the disease became less severe, and symptoms completely reversed in some mice.⁴ The research team explained that diet and nutrients affect the formation, function and lifespan of lymphocytes, and diets that imitate fasting seem to improve health.³

Modified fasting should be undertaken only with supervision from a doctor or dietitian to make sure food choices are nutritionally adequate.

Fiber

The typical Western diet is high in fried foods, butter and processed meat, and low in fruits, vegetables, whole grain and fiber. A European research team points out this type of diet can contribute to inflammation in the intestine and throughout the body and increase the prevalence of autoimmune diseases.⁵ The team suggests increasing dietary fiber could benefit T cells to help lessen inflammation and restore the health of the intestine.

The topic of fiber has become much more exciting as researchers continue to discover new roles for fiber in nourishing the bacteria in the intestinal tract, called the microbiome. The microbiome in turn helps control immune function. Different types of fiber improve the health of the microbiome and are readily available in a balanced diet that includes fruits, vegetables, whole grains and legumes (chickpeas, kidney beans, peas, lentils and others). In one study, the type of fiber found in common fruits and vegetables improved immune response in mice with an experimental form of multiple sclerosis.⁷

Gluten-Free

Gluten is a protein found in wheat, rye and barley. People with celiac disease have an immunological reaction to gluten that damages the surface of the intestine, causing diarrhea, poor absorption of certain nutrients and an autoimmune response throughout the body. Some immune disorders, including autoimmune thyroid disease, also are linked to celiac disease. A gluten-free diet may improve autoimmune disease symptoms throughout the body, for example, in women with Hashimoto’s thyroiditis.⁸

A gluten-free diet is well-established as part of the management of celiac disease. People following a gluten-free diet must
eliminate all sources and derivatives of wheat, rye and barley in their diet. Gluten-free counterparts for traditional sources of gluten such as bread, muffins and snack bars can be found in supermarkets and online. Eliminating ingredients with gluten, as well as foods made in a facility that also processes ingredients that contain gluten, requires careful label reading and shopping.

Sodium Management
The connection between salt and autoimmune diseases might seem surprising. However, it appears a high-sodium diet (sodium is one of two elements in salt, so eating a lot of salty or processed foods increases sodium intake) may prevent certain types of T cells from controlling inflammation, while also stimulating other cells that cause inflammation. This effect has not yet been studied in humans, but various studies confirm the relationship between a high-sodium diet and multiple sclerosis, lupus nephritis, rheumatoid arthritis, colitis and Crohn’s disease in animals. Avoiding eating too much salt is a good strategy for general health.

Supplements
Any decisions regarding supplements should be made with a doctor or dietitian and should focus on shortfalls in nutrients that play a role in immune health:
• Vitamin A is involved in components of innate and adaptive immunity, including natural killer cells, macrophages, neutrophils and lymphocytes. It is stored in the body, and high doses can be toxic.
• Vitamin B6 takes part in several immune functions.
• Vitamins C and E work as antioxidants that protect cells from damage by reactive compounds in the body.
• Vitamin D helps regulate various types of immune cells: monocytes, macrophages, dendritic cells and activated T cells. “Vitamin D deficiency appears to be the most consistent micronutrient deficiency associated with autoimmunity,” explains Anthony Thomas, PhD, director of scientific affairs for Jarrow Formulas. “Essentially, supplementation is warranted to improve and maintain vitamin D status, particularly in people without adequate skin sun (UVB) exposure.”
• The mineral selenium is part of enzyme reactions related to immune function. A recent paper, however, points out that study results regarding supplements and immunity are not consistent, so further research is needed before making recommendations regarding dietary supplements for autoimmune disease.
• Omega-3 fatty acids, found in fish oil supplements, may be considered for people who don’t get omega-3s in their diet from canola oil, walnuts, at least two weekly servings of salmon or other higher fat fish, or other foods.

Overall Diet
Dietary approaches to autoimmune disease are thought to benefit the body by reducing inflammation, repairing existing damage and preventing further damage. Many of the dietary measures that appear to help individuals with diseases such as multiple sclerosis also are part of a well-balanced healthful diet: fish that provide omega-3 fats, dairy products that supply vitamin D, plenty of fruits and vegetables, and fiber-rich foods, along with less meat and processed foods that are low in fiber and high in saturated fat.

One of the challenges in dietary management is people respond differently to restrictions and nutrients. A group of researchers from a hospital in Romania gathered research on various vitamins, minerals, fatty acids, phytochemicals and other food components that have been associated with management of lupus. They concluded a personalized diet
rather than a single type of eating plan could help keep the body healthy, improve remission from lupus symptoms, prevent negative reactions to medication, and improve physical and mental well-being.

“Diet and nutrition answers are not simple as every person has different needs,” says Joanne Gardner, MS, RD, LDN, integrative dietitian nutritionist and certified LEAP therapist for food hypersensitivities at Duke Integrative Medicine in Durham, N.C. “We personalize recommendations. Some people find an approach through their self-directed explorations and experimentations that serves them well. Yet, as with any health improvement protocol, dietary approaches tend to be generalized and will need to be modified for each individual.”

Angela B. Coate-Hermes, RDN, LD, CLT, who works with clients with autoimmune diseases through her company Nourishing Transitions Nutrition Services in Beaverton, Ore., shares a few general tips for people with autoimmune diseases. “I cannot emphasize enough the importance of drinking plenty of water. Water helps to carry oxygen to your cells, protects your kidneys and is essential for removing waste from the system. Proper hydration is also important for digesting nutrients, including water-soluble vitamins that the body cannot properly utilize if you are dehydrated. Chronic dehydration also can contribute to symptoms that are associated with autoimmune conditions, including headaches, fatigue and muscle and joint pain. A lack of proper hydration can also play a role in gastrointestinal issues such as constipation. The current recommendation is to drink at least half of your body weight in ounces every day. For example, if you weigh 150 pounds, you should drink at least 75 ounces of water each day.”

“Next, be sure to have your vitamin D levels checked. Vitamin D deficiency can contribute to a compromised immune system and is linked with many different autoimmune conditions. Having low vitamin D levels can also prevent your immune system from fighting off bacterial and viral infections. And also get tested for food sensitivities if you have not yet been tested. Most people with autoimmune conditions have food sensitivities that lead to inflammation and symptoms associated with inflammation. These can include headaches, joint and muscle pain, fatigue, depression, anxiety and gastrointestinal problems. Identifying your trigger foods and eliminating them from your diet can help to greatly reduce your symptoms related to your autoimmune condition. Work with a registered dietitian nutritionist who is trained in food sensitivities and can help you plan a well-balanced diet that also eliminates your trigger foods.”

MINDY HERMANN, MBA, RDN, is a food and nutrition writer and communications consultant in metropolitan New York.

References

Resources
- National Institute of Environmental Health Sciences — Autoimmune Disease: www.niehs.nih.gov/health/materials/autoimmune_diseases_508.pdf
- Benaroya Research Institute — Autoimmune Diseases: www.benaroyaresearch.org/what-is-bri/disease-information/autoimmune-diseases
Brandon Dillon has come a long way since his CVID diagnosis in 2010. His IVIG infusions make it possible for him to remain healthy and participate in running, cycling, bowling and other physical activities.

Profile:
Brandon Dillon
By Trudie Mitschang

Brandon was in his mid-20s when his first symptoms of common variable immunodeficiency (CVID) began, but it was almost a decade later when he was finally diagnosed. Since then, this active and inspirational athlete has continued to beat the odds by training and participating in numerous athletic events (he completed his first full marathon just six months postdiagnosis). Brandon first shared his story in IG Living in 2014. We recently caught up with him to see how he’s continuing to inspire and motivate others.

Trudie: For readers not familiar with your story, tell us when you suspected something was wrong with your health.

Brandon: I started experiencing recurring sinus infections and bouts of pneumonia at age 25. From then on, what normally started as cold symptoms would usually turn into sinus infections.

Trudie: Describe what happened when you were 37.

Brandon: In October 2010, a week after my 37th birthday, I was experiencing another cold-turned-sinus infection, and I just couldn’t seem to shake it. I ended up going to urgent care because it hurt like crazy just to breathe. The doctor ordered a chest X-ray and blood work, and when he came back into the exam room, he told me I had good news and bad news. The good news was I had pneumonia, and he was sure it could be treated; the bad news was he was sending me to the hospital because my white blood count was through the roof. I ended up being hospitalized for four days. Luckily, the doctor treating me wanted to know why a relatively healthy 37-year-old was in such bad shape (not only was I fighting pneumonia, I had bacteria in my bloodstream). They ran some tests and consulted with an immunologist at the University of Utah Medical Center who diagnosed me with CVID. Had I waited just one more day to get looked at, I probably wouldn’t be here now.

Trudie: What was your life like prior to CVID?

Brandon: Prior to diagnosis, I lived a very normal life. I had a full-time job working for an engineering company, hung out with friends, bowled in leagues a couple of nights a week and was fairly active with running and cycling.

Trudie: What was it like when you got the news?

Brandon: When the doctor came into my hospital room and told me the reason I was so sick was because I had a primary immune disorder, I was terrified. I had no idea what that meant or what my life would be like going forward.

Trudie: How has intravenous immune globulin (IVIG) helped your quality of life?

Brandon: My immunologist told me
with the IVIG infusions, I should be able to maintain a very normal quality of life and do the things I’ve done in the past. I just have to be a little more conscious about my health and avoid people who are sick, which can be challenging.

Trudie: What advice did your immunologist give you that was especially helpful?

Brandon: One thing he told me on my first visit with him that has stuck with me to this day was the goal from here on was to die with CVID, not of CVID. I know it sounds kind of morbid, but it really is true.

Trudie: What motivated you to run that first marathon after diagnosis?

Brandon: When my immunologist told me I would be able to have the quality of life I had before my diagnosis, I took that to heart and signed up to run a marathon the following spring. I started training in January, and six months to the day of lying in a hospital bed and receiving my diagnosis, I ran my first full marathon. Up to that point, the longest race I had participated in was a half marathon.

I feel fortunate we found a treatment regimen that works for me and allows me to keep doing what I love. Saying that, I know there are people out there who have health issues that are a lot worse than mine, so there is no way in my mind I can sit back and take pity on myself.

Trudie: What has changed since you were last featured in our magazine?

Brandon: Since I was last featured in IG Living, I feel like my fitness level has improved. I have taken on and finished two Half Ironman-distance triathlons, my race times seem to be improving, and I just feel good. For example, I participated in an Olympic-distance triathlon last August, finishing fourth overall and first in my age group. That was an awesome feeling.

Trudie: What advice do you have for others who struggle to stay active following a CVID diagnosis?

Brandon: Set aside time each week to get out and do something active — whether that’s going for a walk, bike ride, run or anything else you enjoy. You don’t have to run marathons or compete in Ironmans, just as long as you feel accomplished at what you’re doing. I really feel that getting out and doing something active helps clear the mind and makes you feel good. I might add that it helps to seek out people with similar goals to keep you on track with your fitness goals. Also, signing up for an event such as a 5K, a walk for a cause, etc., is a good way to stay motivated.

Trudie: How do you avoid self-pity?

Brandon: I feel fortunate we found a treatment regimen that works for me and allows me to keep doing what I love. Saying that, I know there are people out there who have health issues that are a lot worse than mine, so there is no way in my mind I can sit back and take pity on myself.

Trudie: What are your future goals?

Brandon: I’m getting the itch to run another full marathon, maybe later this fall. I also plan to do another half-distance triathlon this summer because I have some things I need to work on before I take on a full Ironman, which is my ultimate goal.

Trudie: What has living with chronic illness taught you about yourself?

Brandon: It has taught me I definitely cannot take being healthy for granted because sickness still happens and it will happen. There are times when I’m not feeling well while I’m in the middle of training for an event, and I have to take some time off to feel better and not beat myself up about it. At the end of the day, the running, cycling, bowling and being active are just hobbies I enjoy and are not my full-time job, and if I end up not performing up to my hopes or expectations, then so be it, there is always next time.

Trudie: What are your future goals?

Brandon: I’m getting the itch to run another full marathon, maybe later this fall. I also plan to do another half-distance triathlon this summer because I have some things I need to work on before I take on a full Ironman, which is my ultimate goal.

Trudie Mitschang is a contributing writer for IG Living magazine.
Look Again

By Stacy Oliver

AT FIRST GLANCE, if you were to see me sitting at an outdoor café on a warm summer’s day, you’d think I am the picture of health. I gesture and laugh, cross my legs, take a sip of iced tea. You would assume I’m “normal” (whatever that signifies in society). You might even be intrigued by me, my gregariousness and red hair in the sunlight. But, I’m anything but run-of-the-mill ordinary. I favor my left hand because the neuropathy in my right hand is so bad I can’t use it for certain functions like holding an iced tea. I’ve added Stevia to my tea so I can taste it. My Sjögren’s has changed my taste buds; food and drink smell and taste a bit odd now. The hostess gives me my walker when I leave because of the neuropathy in my legs. I have multifocal motor neuropathy (MMN), the disease no one has heard of, so I have to explain it every time. The sun has shifted, so I must put my hat back on. I also have lupus, so the sun is not my friend. Except for my right hand looking a bit atrophied, you’d never know I have all these diseases. Oh, and there’s the port in my chest. I am treated with immune globulin (IG) infusions three times a month. This is my “normal.”

People are too quick to judge. For instance, they make remarks about people’s weight. Maybe the person is on steroids; maybe he or she has already lost 50 pounds. Or, someone acts upset with another for a trivial reason. Maybe someone they know just died; maybe they are a stressed-out caregiver; maybe they just got a diagnosis that isn’t that great. We can all remember to try and be more generous with others. What we see isn’t always what it looks like.

So many people with chronic illness look like everybody else. When I encounter other people, they sometimes make assumptions based only on what they know. “You use a walker, so your back is out or healing.” “Is MMN like multiple sclerosis?” Or, they have no idea. “What is immuno-what?!” I have what is termed a “fringe disease.” It’s not a typical one most people would understand; it’s rare, and not many have it. (I wish I got a pretty fringe scarf with this fringe disease, but alas, I got stuff way less nice.)

I recently took a sewing workshop. I knew I would be much slower than the other ladies. I told some of them a little about myself so they wouldn’t be surprised by my actions. I’ve had to learn to adapt to my hobbies, and it can look strange. They were very kind and helpful. I actually needed some cloth ironed, and several volunteered. For time’s sake, some of the staff helped me along, knowing I could do it, but just to keep me up to pace. Everyone was very thoughtful, and I had fun doing what I could, being creative and making something.

See, it’s as simple as this: People don’t know unless you tell them.

Don’t make others guess why you’re struggling to do something, or why you’re slower to get things done. Why make people guess? That only results in them feeling bad they didn’t know and you feeling bad because you can’t function the way you want. Or, it leads to you being the odd person out, not able to do what everyone else is doing. Yes, you might have to educate them a bit because most folks don’t know about IG, let alone the rare diseases it treats.

By being honest with others, you’re actually being honest with yourself. Especially if you are like me and became ill later in life. I physically move differently now with MMN, which I got in my 40s.

So, let’s try to be kind when interacting with our fellow neighbors on this planet Earth. I think there is enough discord and misinformation around us; we don’t have to add to it. That means you need to be vulnerable. Brené Brown, who studies human connection, says “vulnerability is the birthplace of love, belonging, joy, courage, empathy and creativity.” I have found when people are given the chance to understand, they rise to the occasion. I feel at ease knowing I can be myself, handicaps and all. The next time you’re about to criticize someone else, take another look at them.

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

By Ilana Jacqueline

WHEN I WAS younger, I had this terrible feeling in my stomach every time I needed to see the doctor. It wasn’t fear, though, it was shame. I felt shame because my doctor would constantly tell me I was faking my symptoms, I wasn’t taking my antibiotics correctly or I was somehow causing or worsening my symptoms. I didn’t know any better at the time, so I walked around with this cloud over my head, knowing I would be in for 10 minutes of disappointed eye-rolling and long-suffering sighs.

It wasn’t until I was well into adulthood that I realized so many of my health problems weren’t my fault. I became better at advocating for myself, but I still — to this day — have doctor appointments that send me to my car crying.

Learning how to cope with shame from medical professionals can seem insurmountable in the moment. My brain might say: “This is just one bad doctor with no bedside manner.” But my heart asks: “Is this all my fault?”

Here are some ways I’ve learned to cope with shame at the doctor’s office:

1) Speak out. Shame is best diluted by sharing it. When you bottle up that feeling, it festers and grows bigger and uglier in your head. And it isolates you. The reality is everyone feels shame, and medical shame is no different. So, whether you post about your experience on a Facebook support group or with your mom on the way home from that terrible appointment, you need to get it out there.

2) Fight back. During some of my more traumatic doctor appointments, I didn’t feel like I could do or say anything in the moment to defend myself. It all happened so fast, and I was so overwhelmed. By the time the doctor walked out of the exam room, I hadn’t said a word in my defense. It wasn’t until I got home, digested what had happened and logged onto my computer to write a review online. It’s important not to get too enraged in your response. Don’t curse. Don’t go full caps lock. Just write about how the appointment made you feel. I’ve used the public review to ask the doctor: “How would you feel if someone had dismissed your daughter the way you dismissed me?”

3) Work it out. Sometimes a phone call with a friend, a post on social media or even leaving a negative review in your defense isn’t enough to clear those feelings of shame. It’s important for patients with chronic illnesses to go to therapy, and shame should be something you discuss at length. Whether you need to act out the scenario again, cry it out or just take a moment to put all of your feelings on the table, therapy helps.

4) Mitigate triggers. If you know you have to follow up with a doctor who has brought you shame in the past, don’t go alone. Bring a friend or family member to speak up on your behalf and to be there to comfort you. Doctors tend to alter bad behavior when a patient has an advocate in the room.

5) Seek support. You are not alone in having felt guilted or emotionally violated by a medical professional. It happens to patients of all kinds, every day. One way of coping with it is to connect with other patients and support them. You can find plenty of first-person stories about the topic on TheMighty.com. You can find your tribe on Inspire.com. Or, you can join local support groups in your area.

Shame can either be a fleeting emotion or a deeply buried one. The choice is yours for how you want to deal with it. But choosing not to deal with it is the only bad choice you can make.

ILANA JACQUELINE is a 29-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.
Ready-for-School Preparations for Preschoolers

By Jessica Leigh Johnson

SENDING A CHILD to preschool for the first time can be bittersweet for parents. While it’s exciting to help her pick out that first-day-of-school outfit or take pictures outside with that way-too-big backpack weighing him down, this transition also marks the end of toddlerhood and the beginning of school years. As parents with older kids know, once a child starts school, time seems to fly by. Pretty soon, that preschooler will be a high schooler prepping for the ACT and going on college visits. As the parent of a high school junior, I often find myself thinking back to my daughter’s first day of school, wondering: “Where has the time gone?” That’s why it’s important to cherish these special firsts in a child’s life, and thoroughly prepare her for what’s to come. Also, keep in mind that a child might be as apprehensive about this transition as her parents. Talking about what to expect and what that first day will look like can go a long way in easing her first-day jitters.

Several preparations need to be initiated months or years before a child starts school, and taking care of these things ahead of time will prevent panic when late August rolls around.

Make sure the child is up to date on immunizations. For a child with chronic conditions such as a primary immunodeficiency disease (PI), allergies and neurological conditions, immunizations are not an option or are not effective. That’s why it’s important for parents of those who can be immunized to make sure their children are up to date before they start school. Although this topic has become increasingly controversial, vaccines are still the best-known way to prevent severe complications from illnesses such as meningitis, measles and whooping cough, to name a few. The Centers for Disease Control and Prevention also recommends children 6 months and older receive an influenza (flu) shot every year.1 Most schools require children to be up-to-date on their immunizations before they are enrolled. For a child with PI or other contraindicative conditions, states offer medical (and religious) exemptions. Exemption forms need to be filled out and signed by a physician before the child can start school.

If necessary, request an individualized education program (IEP). If a child attending preschool has any of the 13 conditions covered under the special education law’s Individuals with Disabilities Education Act (IDEA), he will qualify for an IEP, which requires the school to provide special education services and accommodations to help him maximize his learning potential. Some of these conditions include visual, hearing or “other” health impairments, learning disabilities, autism spectrum disorders, traumatic brain injury, speech or language impairment, along with several others.2

The first step is to request an evaluation by the IEP team, which might include professionals such as a speech-language therapist or occupational therapist. If the team finds the child eligible, parents will work with these professionals to create the IEP. There’s not much else needed to get the IEP started; it should happen within 30 days.2 Therefore, if parents think their child might require special education services or certain accommodations, it would be wise to request an evaluation early enough to have the IEP in place by the time the child begins preschool.

For a child with chronic health concerns, specific medical accommodations can be added to the IEP. These might include unlimited excused absences for medical appointments, steps for addressing/cleaning open wounds (in the case of an immune-compromised child), parent notification when certain transmissible diseases are present in the classroom (flu, strep, chickenpox, etc.), and any other allowances that would help the child succeed in school without being penalized for things that cannot be
helped. It’s really up to the parents and their IEP team to create an individualized, child-focused education plan.

*Purchase school supplies.* A child’s preschool will likely send home a list of recommended school supplies several weeks to months before the first day. On the list will be the usual supplies such as crayons, pencil boxes, a backpack and lunch bag. If a child has a chronic illness or PI, a few alterations might need to be made to the list, and these changes should be discussed with the teacher ahead of time. For example, if the student is required to bring in a box of crayons, ask the teacher if the crayons will become “community supplies” that are shared by all the students. Come flu season, parents of a preschooler with a chronic illness might not want their child sharing supplies — especially if they knew where all those little hands were before using those crayons! It’s not unreasonable for parents of an immune-deficient child to request their child has her own personal school supplies.

Along the same lines, parents can ask if their child can bring her own water bottle to school. This could cut down on germ exposure at the drinking fountain.

Other supplies usually needed in preschool may include an extra change of clothes in case of accidents or run-ins with playground puddles, pull-ups for those not fully potty-trained and a blanket or sleeping pad for naptime, if applicable. The latter should be taken home regularly for cleaning.

Over-the-counter and prescription medications the child uses regularly can be provided to the school nurse, who will administer them to the child at the appropriate times.

*Teach personal hygiene.* One of the first things parents can do to prepare their child for preschool, as well as the school years that follow, is to require good personal hygiene. This training can be done at as young an age as the parents are willing to start. Something as simple as handwashing before meals and after using the restroom goes a long way in preventing illness. Taking on this challenge is worthwhile as most parents are aware preschools and daycares are breeding grounds for germs.

Preschool children ages 3 years to 5 years don’t think twice before putting their fingers into their mouths and noses, and then picking up toys or touching doorknobs, books and anything else in the classroom. All a schoolmate has to do is touch these surfaces and proceed to touch his eyes, nose or mouth to have potentially come in contact with a contagion. So, the earlier a child adopts good hygiene habits, the better. Parents should remind their child to wash her hands with soap under warm running water for at least 20 seconds and to scrub between her fingers. Also, all children should be reminded to always cover their mouths when they cough or sneeze, and use tissues to wipe their noses to help stop the spread of sickness to classmates.

Visit the school ahead of time. To ease first-day jitters and remove the fear of the unknown, parents should take their child to visit their new preschool several weeks before the first day. Ask about scheduled building tours or if one can be arranged. After exploring the school building, let the child play on the playground a few different times. This will give the child a sense of familiarity and increase his comfort with the new setting, and give him something to look forward to when school starts.

This time in a child’s life can be filled

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**One of the first things parents can do to prepare their child for preschool, as well as the school years that follow, is to require good personal hygiene.**

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### References


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

By Heather Bremner Claverie

THE TEARS. The jitters. The nausea. For individuals with an aversion to needles, these reactions can interfere with necessary medical procedures. This can be especially true for adults and children in the immune globulin (IG) community whose medical conditions require frequent infusions.

A Common Phobia

Fear of needles, or trypanophobia, isn’t an odd paranoia. It starts from the time babies get their first shots and scream at the pain of the poke. Fast forward to older children who are now aware the implement the nurse is holding won’t feel good. Then, those patients become adults who may prefer to look away rather than watch the nurse access a vein.

In 1994, needle phobia became a medically defined condition when the American Psychiatric Association’s Diagnostic and Statistical Manual of Mental Disorders added blood-injection-injury phobia into its 4th edition.

An Unavoidable Act

People with needle phobia aren’t aware of it until their first shot. And, since the etiology of needle phobia is an inherited vasovagal reflex (the sight of blood or certain stresses such as those triggered by a needle puncture), it can cause patients to learn to fear needles with each successive shot.

Yet, unlike some phobias that can be alleviated by avoiding the act or object, needles are a medical necessity. And, for individuals who rely on infusions every three or four weeks, it’s not the solution to the problem. In addition, some fear or anxiety around infusions may not be related to needles. For individuals who don’t have easily accessible veins and must deal with repeated pokes, infusions may be particularly painful and uncomfortable.

Out of Sight; Not Out of Mind

Not thinking about an upcoming infusion is one possible coping method, but it doesn’t serve patients well. Mental issues are only a part of the picture. When patients become nervous, their blood vessels dilate and their hearts race. This temporary hypertension causes blood to rush to the heart and chest, in turn making the veins difficult to access. That’s why it’s critical that individuals who suffer from infusion anxiety but rely on it for their health take steps to cope with the issue.

Developing trust with the nurse is one essential component. Letting healthcare professionals know they’re dealing with infusion anxiety gives the nurse a chance to address the issue and prepare their patient. Warming up the infusion sites before the procedure with a bath or heating pads is a simple way to alleviate pain. Also, applying a topical anesthetic that contains lidocaine and prilocaine beforehand will numb the infusion site. Wearing loose, comfortable clothing and drinking plenty of water throughout the day will help, too.

For younger patients, infusions can be a huge source of anxiety. The first step is to find an experienced pediatric nurse who specializes in infusions. Swaddling patients between the ages of 1 year to 2 years has been shown to help since it gives them a sense of security and safety. Older children need to be told about each step of the process. Make sure the nurse gives them details about the procedure before and during, and doesn’t make the mistake of saying it won’t hurt.

Some parents have discovered that the distractions provided by virtual reality glasses are a huge asset during infusions. Anything that lets the patient relax such as calming music or meditation is recommended.

After applying all these remedies, an adage comes into play: It’s difficult to be anxious in the face of humor, so adding humor into pre-infusion rituals may serve as the best medicine.

HEATHER BREMNER CLAVERIE is a contributing writer for IG Living magazine.
Google Daydream View gives patients the power to fly off to other lands and immerse themselves in another world. Simply slip on the comfortable headset and embark on adventures during an infusion session. Patients can stream thousands of movies and apps and share their experiences with friends and family. $59.95; amazon.com

Noise-canceling, wireless headphones are a nice accessory for infusion visits. These comfortable headphones will not only block out surrounding noise, but will give the patient the ability to listen to a steady stream of music, podcast or audiobook. They can even listen to guided meditations during procedures to calm them into a meditative state. $349.95; bose.com

Heating up infusion sites in advance can alleviate some of the pain associated with the procedure since it helps increase blood flow to those areas. The generous size of the Sunbeam Heating Pad for Pain Relief and its 9-foot-long chord gives patients the ability to relax while effectively warming up a 12-by-24-inch area. Three different settings let patients decide the intensity of the temperature, and a removable cover makes it easy to clean. $19.99; sunbeam.com

Herbs have been used for thousands of years to treat a wide variety of ailments, from colds to angst. Vetiver, a plant known as the “oil of tranquility,” can be dropped on the wrist, chest or neck or into bath water. Lavender is another plant well-known for its ability to alleviate anxiety, sleep disturbance and stress. Ingest it via capsules, inhale it or apply it topically. For kids, the Aura Cacia Kids Foam Bath smells good and calms high-strung youngsters. $3.29-$22.99; Sprouts Farmers Market and Target, or auracacia.com

A topical anesthesia such as EMLA cream can help ease some of the anxiety about pain. This U.S. Food and Drug Administration-approved product uses a combination of 2.5 percent lidocaine and 2.5 percent prilocaine, two substances that enter through the skin and block pain receptors in nerve endings. Adults and children can use this cream, but dosage guidelines should be discussed with their physician. Available by prescription only

Numerous studies have shown simple meditation is an effective tool to combat anxiety symptoms. But calming an active mind isn’t as simple as it appears. Guided meditations are a great way to start and can help calm the nerves before or during infusion procedures. Mindful is a great resource for a variety of guided meditations that can help individuals navigate difficult emotions and improve sleep and destress. free; www.mindful.org/mindfuls-top-10-guided-meditations-of-2018
Ataxia Telangiectasia (A-T)

**WEBSITES**
- A-T Children’s Project: www.atcp.org

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

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

Evans Syndrome

**ONLINE PEER SUPPORT**
- Evans Syndrome Research and Support Group: www.evanssyndrome.org

Guillain-Barré Syndrome (GBS)

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

Idiopathic Thrombocytopenic Purpura (ITP)

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

Kawasaki Disease

**WEBSITES**
- American Heart Association: www.heart.org/HEARTORG/Conditions/More/CardiovascularConditionsOfChildhood/Kawasaki-Disease_UCM_308877_Article.jsp#T1T2b0ePWE0
- Kawasaki Disease Foundation: www.kdfoundation.org
- KidsHealth: kidshealth.org/parent/medical/heart/kawasaki.html

Mitochondrial Disease

**WEBSITES**
- United Mitochondrial Disease Foundation: www.umdf.org
- MitAction: www.mitaction.org

Multiple Sclerosis (MS)

**WEBSITES**
- All About Multiple Sclerosis: www.mult-sclerosis.org/index.html
- Multiple Sclerosis Association of America: mymsaa.org
- Multiple Sclerosis Foundation: www.msfocus.org
- National Multiple Sclerosis Society: www.nationalmssociety.org

**ONLINE PEER SUPPORT**
- Friends with MS: www.FriendsWithMS.com
- MSWorld’s Chat and Message Board: www.msworld.org

Myasthenia Gravis (MG)

**WEBSITES AND CHAT ROOMS**
- Myasthenia Gravis Foundation of America (MGFA): www.myasthenia.org
- Genetic Alliance: www.geneticalliance.org

Myositis

**WEBSITES**
- The Myositis Association: www.myositis.org
- International Myositis Assessment and Clinical Studies Group: www.rheum.nhs.uk/research/resources/imascs

**ONLINE PEER SUPPORT**
- Myositis Support Group – UK: www.myositis.org.uk

Peripheral Neuropathy (PN)

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

Primary Immune Deficiency Disease (PI)

**WEBSITES**
- Immune Deficiency Foundation: www.primaryimmune.org
- Jeffrey Modell Foundation: www.info4pi.org
- The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/Pages/index.aspx
- American Academy of Allergy, Asthma & Immunology: www.aaaai.org
- International Patient Organisation for Primary Immunodeficiencies (IPOD) — UK: www.ipodi.org
- New England Primary Immunodeficiency Network: www.nepin.org
- Rainbow Allergy-Immunology: www.uhospitals.org/rainbow/services/allergy-immunology

**ONLINE PEER SUPPORT**
- IDF Friends: www.idffriends.com
- Jeffrey Modell Foundation Facebook Page: www.facebook.com/JMFworld
- IDF Peer Support Program: www.primaryimmune.org/idf-peer-support-program
- Michigan Immunodeficiency Foundation: www.idfworld.org/en/nonprofits/2432e2b2a15942206f23d8a8703c9e-michigan-immunodeficiency-foundation-monroe

Scleroderma

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

**ONLINE PEER SUPPORT**
- International Scleroderma Network: www.sclero.org/support/forums/a-to-z.html

Stiff Person Syndrome (SPS)

**WEBSITES**
- American Autoimmune Related Diseases Association Inc.: www.aarda.org
- Genetic Alliance: www.geneticalliance.org
- Living with Stiff Person Syndrome (personal account): www.livingwithspss.com
- Stiff Person Syndrome: www.stiffpersoonsnet.org

Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)

**WEBSITES**
- PANDAS/PANS Advocacy and Support: www.pas.care
- PANDAS Network: www.pandaznetwork.org
- Midwest PANS/PANDAS Support Group: www.midwestpandas.com

Pemphigus and Pemphigoid

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