furry comfort
The Healing Power of Pets

PI and Fatigue: Can It Be Treated?

How to Assess Your Home for Safer Living

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Editorial
Approaches to Feeling Better in the New Year
By Ronale Tucker Rhodes, MS

Abbie’s Corner
Appealing Insurance Denials
By Abbie Cornett

Features

Furry Comfort: Animal-Assisted Therapy for Chronic Illness
By Trudie Mitschang

PI Patients and Fatigue
By Terry O. Harville, MD, PhD

Improving Safety, Mobility and Activities of Daily Living at Home
By Matthew D. Hansen, DPT, MPT, BSPTS

Understanding Ehlers-Danlos Syndrome
By Meredith Whitmore

Sources

Product Guide
Understanding the ‘Pros’ of Probiotics
By Trudie Mitschang

Resource Center
Community foundations, associations, forums and other resources

Columns

Let’s Talk — Pete Atherton
By Trudie Mitschang

Patient Perspective — When the Impossible Happens, Is Anything Possible?
By Stacy Oliver

Life as a 20-Something — The Fine Line Between ‘Fear’ and ‘Vigilance’ at the Doctor’s Office
By Ilana Jacqueline

Parenting — Making Hospital Stays Easier
By Jessica Leigh Johnson

Departments

Faces of IG
From our Facebook page

Immuno101
DiGeorge Syndrome Development: The “Timing Is Off” Part 7 (Parathyroid Glands)
By Terry O. Harville, MD, PhD

Ask the Experts
Healthcare professionals’ responses to patient questions

Clinical Brief
Toward the Ultimate Cure: Gene Therapy for Severe Combined Immunodeficiency
By Keith Berman, MPH, MBA

In the News
Research, science, product and insurance updates

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

IG Living, (ISSN 1949-4548), published bimonthly, is a community service provided by FFF Enterprises, 41093 County Center Drive, Temecula, CA 92591, (800) 843-7477 x1362, fax (951) 699-9655.

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

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UP FRONT

Approaches to Feeling Better in the New Year

AT THE END of 2014, Lottie Ryan, who has suffered with chronic illness for 19 years, posted her New Year’s resolution on her blog, suggesting others with chronic illness make the same resolve: “Every day I will concentrate on what I can do (and never dwell on what I can’t do).” While the traditional time for New Year’s resolutions has come and gone, it’s not too late to resolve to take steps that could improve quality of life in 2017. That’s the focus of this issue’s articles.

I can’t think of a more pressing resolve for so many with primary immunodeficiency disease (PI), or any other chronic illness, than finding ways to fight fatigue. So, we turned to immunologist Terry Harville to explain why PI patients experience fatigue and how it can be treated. In his article “PI Patients and Fatigue,” Dr. Harville defines the medical causes of fatigue, specifically for those with PI, and explains that while there is no one treatment for alleviating fatigue, it is solvable with individualized intervention.

Other debilitating symptoms faced by PI patients can be a result of gastrointestinal issues, caused not just by bacteria but by frequently used antibiotics that destroy bad bacteria and also wipe out good bacteria. Fortunately, probiotics can help to minimize these issues. In “Understanding the ’Pros’ of Probiotics,” we provide a reference to the probiotic strains to look for on product labels, as well as a short shopping list to guide patients to products containing them.

Chronic illness often comes in the form of physical and/or mental conditions that can complicate daily activities. In those cases, a home assessment for health hazards is critical to ensuring patients’ safety. In his article “Improving Safety, Mobility and Activities of Daily Living at Home,” physical therapist Matt Hansen points out the benefits of a home safety assessment. While it’s possible to have one conducted by a health professional, Hansen provides a detailed indoor and outdoor checklist that anyone can easily use to conduct such an assessment.

Finally, there is perhaps nothing that can make most people feel better than furry companionship. Indeed, studies show that pets can do wonders for patients, including having positive psychosocial and psychophysiological effects, easing feelings of loneliness and isolation, taking their mind off their illness and even healing chronic pain. Our article “Furry Comfort: Animal-Assisted Therapy for Chronic Illness” discusses the benefits of animal-assisted therapy, providing real-life examples of how it has helped. It also outlines some precautions for immune-compromised patients.

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

Ronale Tucker Rhodes, MS

Appealing Insurance Denials

By Abbie Cornett

AS A PATIENT ADVOCATE, I help patients with a wide range of concerns, but one of the most common issues I am asked to address is how to appeal insurance denials. Typically, chronic illnesses often require patients try multiple different medications and treatments to narrow down the one that works best, and in many cases, a particular treatment or medication is denied or switched for nonmedical reasons. Being denied needed medical treatment is never easy, and is an all-too-common issue.

If denied insurance coverage, patients have the right to know the reason their claim is being denied and how to appeal the decision. The first step is called an internal appeal, during which patients ask for a review of the decision to deny coverage. If there is an urgent medical need, the insurance company must conduct the review in a timely manner.

A second appeal option is to request an independent third party to review the decision. The independent review organization must not be connected to either the patient or the insurer, and must provide a review from a physician that specializes in the area of medicine to which the claim is related. If the independent reviewer overturns the denial, the insurance company must approve the claim.

There are several dos and don’ts of the appeal process that patients need to observe for best results.

Don’t

1. Don’t start the appeal process over the phone. Insurance companies will frequently ask patients to call them regarding a denial. But, be aware that many insurers consider a phone call regarding an appeal the appeal. If no new information is submitted, this can be a reason to deny. Instead, all information should be submitted as part of a package either through the mail or electronically, not over the phone.

2. Don’t take it personally. Patients should resist the impulse to respond on an emotional level. The denial is a business decision for the insurance company. This means patients should not write a letter ranting at the insurance company, or one that merely says the medicine is needed. Any communication with the insurer should be professional, and should include medical documentation backing up the claim.

Do

1. Determine under what type of plan the claim falls. There is a difference between a fully insured plan and a self-funded plan, also known as an Employee Retirement Income Security Act (ERISA) plan, which is governed by state laws.

2. Read the policy thoroughly to determine coverage. Under the Affordable Care Act, patients have 180 days to file an internal appeal.

3. Collect all medical records, tests, lab results and doctors’ notes. Many times, denials occur when insurance companies think there may be a less-expensive treatment available. The patient’s doctor can write a letter explaining why the medical test or medication is necessary, and why the less-expensive treatment either didn’t work or isn’t equivalent.

4. If patients do have to call their insurer for information regarding the denial, they should make sure to state that they are not starting the appeal process with the call. Also, a record of the call should be kept, including the date, time and name of the person spoken to. If communicating by mail, send all letters by registered mail with a return receipt.

Denial Is Just the Beginning

What is important to remember is that being denied coverage is not the end of the fight, but only the beginning of what can be a long and stressful battle. Patients shouldn’t lose hope, though. More than 70 percent of appeals are successful.

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

Sources


**IG Living Reflects Reader’s Needs**

*IG Living* is my absolute favorite magazine! When it arrives, I stash it away, waiting for some uninterrupted time to savor the information and connection that it brings. The articles always help [me cope] with the many sides of living with a chronic illness. The October-November 2016 issue, however, really hit home for me in a way that no other issue ever has; this was the first issue that spoke to me personally.

Ronale Tucker Rhodes (Up Front, page 5) said that patients treated with IG run “the gamut; some live normal lives not hampered by chronic recurring infections, while others are not so fortunate and constantly struggle to function in the activities of daily living.” This is my story: I am chronically ill and severely disabled by my immune deficiency. Too often, I read that the chronically ill should “refuse to let their diagnosis define who they are or what they can accomplish.” Not all of us have that luxury! In fact, I often feel that despite my education, drive, self-discipline and type-A personality, my illness does dictate so much of what I cannot accomplish, where I cannot go, what I cannot do and whom I cannot see. Don’t get me wrong, there are still many things I can do, and [there is] an entire new life I have chosen for myself — but it is not the life I wanted or imagined. After 28 years on disability, I still grieve many losses. Grief does not just end; some grief is unattenuated and seems to compound with time, even as I grow wiser and gain more coping skills.

Trudie Mitschang’s article, “Goal-Setting for the Chronically Ill: Planning for an Uncertain Future” (pages 22-25), was excellent. She did not repeat the often-heard mantra to “think positive, and achieve anything.” Instead, she was realistic in saying that the chronically ill may face “daily roadblocks to accomplishing even basic tasks.” She acknowledged that even setting goals might feel impossible to the chronically ill, and went on to mention the very real limitations and ongoing grief over the interference with social, career and family goals: “No matter how well one lays out goals or commits to achieving them, the fact is a chronic illness does retain a measure of control over what can and cannot be achieved on any given day.”

For me, at age 64, with my personal/professional successes/losses, the uncertainty of my future and even my energy level on any given day, I take heart in her kind words of inclusion. Those of us who experience this level of illness are a small percentage of those living with immune deficiency. Still, it is nice to know that our needs can be reflected in this magazine. What I have learned over the years is that in addition to my determination to shower, put on a pair of earrings and fix my hair every day no matter how ill, I am also learning to surrender gracefully and accept that there is so much I will not do, so many places I will never go, and that I will not achieve what I set out to do. This takes great courage. Thanks for including all of us on the journey.

— Marilyn McVicker

**Critical Coverage Needed for Many Disease States**

I have stiff person syndrome (SPS) and paraneoplastic syndrome, which has turned into cancer. Unfortunately, I have never seen either of these diseases written about in your magazine. Would you please consider covering some of the more rare diseases for which immune globulin (IG) could be an effective treatment?

— Unnamed

**Reply from the editor:**

We have covered and/or made mention of SPS in several issues: April-May 2007 (page 30), October-November 2007 (page 17), April-May 2009 (page 40), February-March 2010 (page 36) and June-July 2013 (page 22), all of which are available on www.IGLiving.com. However, it has been several years since we devoted an article specifically to SPS, which is why we have one scheduled for the August-September 2017 issue. In addition, please look for coverage of the benefits of high-dose intravenous IG treatment for paraneoplastic syndrome in the April-May 2017 Clinical Brief column.
How did you feel when you were diagnosed?

Kind of relieved. However, I didn’t realize myasthenia gravis is a disease that many doctors don’t have a lot of information about. I realized quickly that I had to learn everything possible to keep myself functioning to whatever degree possible. Worse yet, going into the hospital is a nightmare because it’s up to me to keep everyone informed.

— J Sonkin

Relieved! I did not like the diagnosis or the treatment, but I was relieved I had a diagnosis about my health issues. It helped some people believe me, while others were still skeptical, which was really sad, but I found relief in knowing it wasn’t in my head like so many others, including my primary care physician (PCP), thought. After 25 years, I finally switched my PCP.

— D Sprayberry

Were you believed [about your illness]?

I do not have the energy to get into my own story, but I will say how sad it makes me that most of us carry the weight of disease on our shoulders with little to no support from our closest friends and family members. I feel blessed for the ones I do have.

— KH Russell

To this day, there are doubters. When a new problem or diagnosis comes up, I feel like “here we go again.” I’m also frustrated that people seem to think the keys to my good health are to eat better, take supplements or try anything else new out there that is the latest fixer. I try to always thank the person giving me “tips” about how to cure my disease, but sometimes I want to scream: “This is not curable, only treatable.”

— J Gardner

I especially like my friends who are selling some expensive juice or pill that will cure everything from AIDS to ZES and then try to make me feel bad when I don’t buy, saying: “I guess you just don’t care about your health that much. This will really fix you right up, and you don’t even want to give it a try.”

— EJ Hiten-Kizer

I was completely gobsmacked! I wanted to know everything about common variable immunodeficiency, and my primary concern was for my daughter.

— M Catanzaro

I was actually relieved. When I was going from doctor to doctor knowing something was wrong with me, I came home one night and told my husband I just wanted a ‘name’ for whatever was wrong with me. I had been told I was a hypochondriac, I needed counseling, I was looking for attention and this was not a good way to get it, and on and on it went. When I got my diagnosis, I was thrilled. That lasted for about a week until my follow-up appointment, and I realized my life had just changed drastically and maybe not positively. Moral of the story: Be careful what you ask for.

— J Gardner

With my cancer diagnosis, I was terrified. With my common variable immunodeficiency diagnosis, I actually was annoyed that I would be dealing with another chronic condition, but I was happy it could be treated.

— D Konrad

By Terry O. Harville, MD, PhD

PREVIOUSLY, WE discussed the improper timing and sequence of the formation of anatomic structures that result in DiGeorge syndrome (DGS) and partial DGS (PDGS) features. In this issue, we continue the discussion of the improper timing of the parathyroid glands.

The parathyroid glands are critical for maintaining calcium and phosphorus levels in the body. They do this by secreting the parathyroid hormone (parathormone, or PTH) and calcitonin (a polypeptide hormone) that regulate absorption of calcium and phosphorus from the intestines, kidneys and metabolism into the bones (vitamin D also plays a critical role in these processes), which is required to maintain the levels in narrow ranges for appropriate cardiac muscle, skeletal muscle, peripheral nerve and brain function. Too much calcium (hypercalcemia) may result in muscle weakness, twitches/spasms, nerve irritability and abnormal cardiac rhythms, which can be fatal. Too little calcium (hypocalcemia) can result in numbness and tingling of the extremities, muscle spasms and extreme contraction of muscles (tetany). Seizures and cardiac rhythm abnormalities can also occur, which can be fatal. Thus, maintaining appropriate serum calcium levels is critical for well-being.

There are four parathyroid glands that begin development in the tissues of the third and fourth pharyngeal pouches (embryonic “gill” slits), as does the thymus. Eventually, the glands migrate to their normal final positions adjacent to the wings of the thyroid gland in the neck, hence their name. Because the parathyroid glands and thymus begin developing at the same time, disruption of the development of the thymus, as found in DGS/PDGS, may also disrupt the development of the parathyroid glands. This can be variable, though, depending on when the timing of the disruption occurs. For instance, there can be essentially absence of the thymus, resulting in complete DGS, but still with sufficient parathyroid tissue in the neck near their origins to maintain calcium metabolism. Alternatively, the thymus may be near normal in its formation, but have essential lack of parathyroid glandular tissues and severe hypocalcemia.

Hypocalcemia, due to decreased presence of parathyroid glands, is one of the original hallmarks described in DGS. Generally, an infant born with DGS/PDGS, who does not have an initially fatal cardiac lesion, will suffer seizures due to hypocalcemia about nine days after birth. In some cases, this may be the first indication that DGS/PDGS may be present. Regardless, the presence of neonatal hypocalcemia should be a signal to initiate a more comprehensive workup for DGS.

Once hypocalcemia is recognized, and can be attributed to decreased or absent parathyroid tissue, it can be treated with hormone replacement, vitamin D supplementation and calcium supplementation.

We will continue in the next issue with more discussion of the resulting issues when the timing is off during fetal development.

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.
Hizentra is the only subcutaneous Ig treatment with over 70,000 patient-years of experience

Important Safety Information

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

- Weekly: Start Hizentra 1 week after last IGIV infusion
  Initial weekly dose = Previous IGIV dose (in grams) x 1.37
  No. of weeks between IGIV doses
- Biweekly: Start Hizentra 1 or 2 weeks after the last IGIV infusion or 1 week after the last weekly Hizentra/IGIV infusion. Administer twice the calculated weekly dose.
- Frequent dosing (2 to 7 times per week): Start Hizentra 1 week after the last IGIV or Hizentra/IGSC infusion. Divide the calculated weekly dose by the desired number of times per week.
- Adjust the dose based on clinical response and serum IgG trough levels.

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

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As tolerated

DOSE FORMS AND STRENGTHS
0.2 g per mL (20%) protein solution for subcutaneous injection

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

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

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

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

USE IN SPECIFIC POPULATIONS
- Pediatric: No specific dose requirements are necessary to achieve the desired serum IgG levels.
**Abbie** I spoke with immunologist Terry Harville, MD, and he said hair loss is not commonly associated with PI or with IG replacement therapy; however, it can occur due to autoimmunity, changes in hormones, toxins/poisons and infections, among other things.

According to Dr. Harville, hair follicles live in different states of being, including the two major states: 1) active growing and 2) dormancy. The ratio of these essentially determines how much hair someone has. He suggests that something may have affected your numbers of growth versus dormant follicles. It may be worthwhile to see a rheumatologist or dermatologist to determine whether an autoimmune disorder associated with alopecia is occurring. He also recommends having a biopsy to evaluate the relative level of active versus inactive follicles.

**Michelle** It is always possible that IVIG won’t work to treat a disease. However, because it sometimes takes between three months and six months before improvement is seen, its efficacy can’t be determined until that time.

Many patients treated with IVIG or subcutaneous IG (SCIG) experience mild to moderate side effects. There are many ways to treat or eliminate side effects. I recommend speaking to your physician about premedicating with antihistamines and/or anti-inflammatory drugs and hydrating before and after infusions. You should also discuss slowing your infusion rate or switching to SCIG. More information on treating side effects can be found on our blog at [www.igliving.com/BlogEngine/post/Treating-Infusion-Side-Effects](http://www.igliving.com/BlogEngine/post/Treating-Infusion-Side-Effects).

**Question** Are gastrointestinal issues common after an immune globulin (IG) infusion?

I am a 60-year-old female with common variable immune deficiency, and I have been receiving Gamunex infusions for the past six years. I also have irritable bowel syndrome (IBS), and right after an infusion, I get nauseous and constipated. It usually takes a week for my body to get back on track. Is this common?

**Question** Can immune globulin (IG) therapy result in hair loss?

I have a primary immunodeficiency disease (PI) and have just finished my first full subcutaneous dose of Hyqvia. I have always had very thick hair, but after three weeks, I noticed extreme hair loss. My physician tested my hormone, iron and thyroid levels, and all were normal. I have been taking a B complex vitamin since my PI diagnosis, and I just started taking Biotin. Are there any explanations for the hair loss and how to counteract it?

**Question** Do some people fail to respond to intravenous immune globulin (IVIG) therapy, and are severe side effects from treatment normal?

I have chronic inflammatory demyelinating polyneuropathy for which I have had two months of IVIG. So far, I have seen no improvement at all, and I have been experiencing severe headaches, nausea, vomiting and pain in the back of my neck when I receive treatment. Does IVIG fail to work for some people? And, are the side effects normal?

**Michelle Greer**, RN, is senior vice president of sales at NuFACTOR Specialty Pharmacy.

**Abbie Cornett** is the patient advocate for IG Living magazine.
Toward the Ultimate Cure: Gene Therapy for Severe Combined Immunodeficiency

By Keith Berman, MPH, MBA

PRIOR TO THE successful use of hematopoietic stem cell transplantation (HSCT) to reconstitute the immune system, profound derangements of both cellular and humoral immunity in newborns born with severe combined immunodeficiency (SCID) were always fatal in infancy. Clinical experience accumulated over the last two decades has helped refine HSCT therapy and, thus, improve the odds of long-term survival for children born with this extremely rare genetic disorder. It is now apparent, for example, that for certain patients, the likelihood of long-term survival improves with a less intensive myeloablative conditioning regimen, or no conditioning at all. It’s now well-documented that survival odds are much better in SCID infants diagnosed and transplanted in the first three-and-a-half months after birth, or who are fortunate enough not to have experienced an infection prior to their procedure. This understanding of the importance of very early transplantation was a major impetus for universal newborn SCID screening with the T-cell excision circle (TREC) assay, which is now in place or is being implemented in 47 states.

But the predominant factor that influences long-term survival is outside anyone’s control: the availability (or nonavailability) of blood-forming hematopoietic stem cells (HSCs) from an HLA-identical sibling donor. For SCID infants with a matched sibling bone marrow or peripheral blood stem cell donor, the prospects for long-term survival are now edging toward 100 percent. Unfortunately, less than 25 percent of patients have a matched sibling donor.

SCID patients for whom only a matched unrelated or haploidentical HSC donor is available face far higher risks of life-threatening complications and death. Many continue to suffer severe recurrent infections due to only partial engraftment and incomplete restoration of immunity. HSCT will fail to adequately restore B-cell immunity in as many as one-half of these patients, necessitating chronic antibody replacement therapy with intravenous immune globulin (IVIG). Repeated breakthrough infections and complications of HSCT, including graft-versus-host disease, can
cause cumulative damage to the lungs and other vital organs. Many of these children experience failure to thrive and cognitive deficits. More than one in four will succumb within the first five years after HSCT.¹

The one remaining therapeutic option has been the dream of clinicians for decades: to treat the disorder by correcting it at its most fundamental genetic level, with gene therapy.

**The Infancy of Gene Therapy**

By the 1980s, genetic flaws causing the two most common forms of SCID had been identified and fully characterized:

- **ADA-SCID**: A defective gene encoding adenosine deaminase (ADA) results in a deficiency of this metabolic enzyme, which is critical for lymphocyte differentiation and growth.

- **SCID-X1**: A defective gene on the X chromosome that encodes the common gamma chain of the interleukin-2 (IL-2) receptor (IL2RG) results in profound disruption of the development of T lymphocytes and natural killer (NK) cells.

In a handful of cutting-edge laboratories here and in Europe, techniques were developed to exploit the ability of gamma-retroviruses to ferry and insert normal copies of the affected genes directly into the DNA of CD34+ lymphocytes collected and purified from the patient’s own bone marrow or blood circulation. The idea was to expand these gene-corrected cells ex vivo and reinfuse them into the patient, to find their way into the bone marrow space and differentiate into the T and B lymphocytes and NK cells that mediate immune function (Figure 1).

On Sept. 14, 1990, after much testing in animal models, U.S. investigators at the National Institutes of Health initiated the world’s first human gene therapy trial in a 4-year-old U.S. girl with ADA-SCID.⁴ Shortly thereafter, Italian investigators at the San Raffaele Telethon Institute for Gene Therapy initiated their own trial, employing different gamma-retrovirus vectors to insert the normal ADA gene into defective CD34+ lymphocytes harvested from patients with ADA-SCID.⁵

In simultaneous reports published five years later in the journal *Science*, the Italian and U.S. teams announced highly encouraging findings in a total of four ADA-SCID patients who had previously been supported with exogenous ADA replacement therapy.⁶⁷ The T cell and NK cell counts normalized in all patients, as did a number of cellular and humoral

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**Figure 1. Schematic Representation of a Gene Therapy Procedure for SCID**

1. Bone marrow or peripheral blood CD34+ stem cells are harvested from the patient
2. Stem cells are cultured with a modified viral vector carrying the normal, corrective gene sequence
3. The viral vector inserts the corrective gene into the stem cell cDNA
4. The gene-corrected stem cells are reinfused, migrate to the bone marrow and engraft
immune responses. ADA gene expression persisted in the two U.S. patients at four-year follow-up. At that point, it appeared that the dream might soon become reality.

**Protocol Change Transforms ADA-SCID Gene Therapy**

Unfortunately, it soon became evident that the initial engraftment of retrovirus- and ADA gene-transduced T lymphocytes did not sustain itself sufficiently over the long term; the small proportion of genetically corrected HSCs that did engraft failed to provide adequate protection against severe infections. Fortunately, later experimentation demonstrated that mild cytoreductive “conditioning” with low-dose busulfan or similar agents, essentially making room in the bone marrow for the reinfused gene-corrected CD34+ lymphocytes, was highly effective in facilitating immune reconstitution.

In 2002, the San Raffaele Telethon team was the first to describe sustained engraftment of genetically engineered HSCs using nonmyeloablative conditioning, with long-term increases in T-cell counts, normalization of T-cell function and restoration of a robust humoral response to vaccine challenges in two ADA-SCID patients. Seven years later, this same team reported the outcomes of gene therapy using autologous CD34+ bone marrow cells transduced with a gammaretroviral vector in 10 children with ADA-SCID and no available HLA-identical sibling donor: All 10 patients were alive after a median of 4.0 years (range 1.8 to 8.0) with stable engraftment of HSCs. Nine of the 10 had normalization of T-cell function, and five no longer required IVIG replacement therapy. “Effective protection against infections and improvement in physical development has made a normal lifestyle possible,” the investigators reported. Other gene therapy research teams in Europe and the U.S. have published similar results with different ADA-SCID gene therapy protocols using a conditioning regimen and conventional gamma-retroviral vectors to insert normal copies of the ADA gene into CD34+ HSCs.10,11

New findings published this year by an international consortium again led by San Raffaele Telethon confirm 100 percent long-term survival in 18 consecutive ADA-SCID patients receiving gene therapy, with normalization of T-cell populations, reduced need for IVIG replacement and a 10-fold mean reduction in severe infection rates. Further, in contrast to other severe primary immunodeficiency disorders treated with gene therapy, no cases of retrovirus-mediated insertional mutagenesis have been identified in any of the roughly 60 ADA-SCID patients treated to date.

A landmark event in May 2016 marked the culmination of a 25-year journey by these investigators, their clinical collaborators on three continents and the courageous families that agreed to participate. Acting on a recommendation by the European Medicines Agency — Europe’s equivalent of the U.S. Food and Drug Administration (FDA) — the European Commission approved Strimvelis (autologous CD34+ cells transduced to express ADA) for the treatment of patients with ADA-SCID for whom no suitable HLA-matched related stem cell donor is available. It is the world’s first licensed corrective childhood gene therapy.

Strimvelis will be marketed in European Union countries by GlaxoSmithKline, which also collaborated in its final stages of development. This individually customized treatment for ADA-SCID fulfills the promise of gene therapy: to essentially cure the more than three-quarters of ADA-SCID children who do not have a suitable donor for HSCT.

**Gene Therapy for SCID-X1: Full Stop to Full Speed Ahead**

Concurrent with preliminary ADA-SCID trials in the early 1990s, other investigators were actively testing similar gene therapy protocols to treat male children with X-linked severe combined immunodeficiency (SCID-X1) and a poor HSCT prognosis. SCID-X1 is the predominant disease variant, accounting for 50 percent to 60 percent of all SCID cases. Preclinical studies confirmed that gamma-retroviral vectors effectively inserted the healthy gene for IL2RG into harvested CD34+ cells. A decade later, after incorporating the mild nonmyeloablative conditioning that produced durable engraftment in ADA-SCID gene therapy trials, small published SCID-X1 patient series documented persisting normalized T, B and NK cell counts and restored immune functions.13,14

But a shocking and unexpected setback put a halt to progress in SCID-X1 gene therapy: Five of the first 20 patients treated in these trials developed T-cell leukemia between two and five years after their procedure. In all cases, evidence pointed to aberrant activation of nearby oncogenes triggered by a powerful oncogene “enhancer” element within the gammaretroviral vector.

Investigators immediately set about developing safer gene transfer vectors. Two types are now being evaluated in clinical trials with encouraging efficacy results and, thus far, without any evidence of a leukemogenic effect. One is modified gamma-retrovirus vectors entirely devoid of enhancer sequences,15 and the second is novel “self-inactivating” lentiviral vectors designed to have a reduced risk of activating oncogenes.16 Extended patient accrual and follow-up will be necessary to establish whether these alternative vectors prove to be safe in current SCID-X1 gene therapy trials, as well as in ongoing trials evaluating
gene therapy for two other primary immunodeficiency disorders — Wiskott-Aldrich syndrome (WAS) and chronic granulomatous disease (CGD) — in which earlier use of gamma-retroviral vectors was also associated with fatal acute leukemia (Table 1).

**On the Horizon: More Approvals for More Uses**

Past challenges of long-term engraftment of gene-corrected progenitor cells and vector-associated leukemias risk appear to have been resolved. All evidence now suggests that gene therapy is largely curative for most patients with ADA-SCID and SCID-X1, without the risks of curative for most patients with ADA-Deficient Immunodeficiency (SCID).

Additionally, findings from six currently ongoing gene therapy clinical trials will be of value to further optimize patient outcomes. All evidence suggests that it is now only a matter of time before gene therapy protocols for these two rare genetic disorders — and potentially CGD, WAS and other life-threatening primary immunodeficiencies — secure FDA approval for commercialization.

Meanwhile, what has been learned about how to optimize the safety and efficacy of gene therapy from experience with SCID patients has helped researchers to design better gene therapy vectors and clinical protocols for far more common genetic disorders, including, for example, sickle cell disease, beta-thalassemia and hemophilia A and B. Thanks to this pioneering SCID research, thousands of patients with these debilitating disorders may not need to wait so long for potentially curative gene therapy: All three are currently the subjects of active clinical trials.

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**References**


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**Table 1. Active U.S. Gene Therapy Trials for Primary Immunodeficiency Disorders**

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<tr>
<th>Disorder</th>
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<td>Adenosine Deaminase-Dependent SCID (ADA-SCID)</td>
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<td>Autologous Transplant of EFS-ADA Modified Bone Marrow Cells for ADA-Deficient Immunodeficiency (SCID)</td>
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<td>Gene Transfer for SCID-X1 Using a Self-Inactivating Gammaretroviral Vector</td>
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<td>Wiskott-Aldrich Syndrome (WAS)</td>
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<td>Pilot and Feasibility Study of Hematopoietic Stem Cell Gene Transfer for Wiskott-Aldrich Syndrome</td>
<td>NCT01410825</td>
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Research

Study Shows PI Patients Could Be at Risk from MMR Vaccine

A new study shows that the measles-mumps-rubella vaccine, part of the routine recommended vaccines schedule for children, could be dangerous for primary immunodeficiency (PI) patients. The study, conducted at the Children’s Hospital of Philadelphia (CHOP), looked at data from 14 patients in the U.S. Immune Deficiency Network and the Clinical Immunology Society immune deficiency Listserv (12 from the U.S. and two Europeans) who had cutaneous granulomas. PI patients typically suffer from infections, one of which can be inflammatory skin lesions known as cutaneous granulomas, which are masses of immune cells that can become ulcerated and, ultimately, fatal. Using immunofluorescence staining, the researchers determined that seven of those patients tested positive for rubella virus, acquired during vaccination at an earlier point. At least one tested positive on a bone biopsy. Three, all children, died from serious infections.

While the researchers are unsure whether the rubella virus was a cause of the children’s deaths, Kathleen Sullivan, MD, PhD, chief of the division of allergy and immunology at CHOP and an author of the study, believes the vaccine caused the granulomas to form and that PI patients are unable to rid their bodies of the small, weakened dose of rubella they receive in the vaccine. “Our study clearly demonstrates that people with moderate T cell defects should not receive it,” said Dr. Sullivan. Moreover, she says, “people with antibody defects [who are] on immunoglobulin replacement are already protected by the immunoglobulin product they receive. [And,] more severe T cell deficits usually are treated with transplantation, and [patients] can be safely vaccinated after transplant.”

Legislation

21st Century Cures Act Impacts SCIG Therapy Reimbursement

In December, former president Obama signed the 21st Century Cures Act, which took effect Jan. 1. The act’s major provisions will reform the current standards and appropriations for biomedical research, provide $1.75 billion annually for the National Institutes of Health (NIH) and $110 million for the U.S. Food and Drug Administration (FDA) — funding that will end after five years. Along with an increase in NIH and FDA funding, the bill will reduce regulations on access to medical research and expedite the testing processes of new drugs. However, two minor provisions of the act directly impact how Medicare reimburses for subcutaneous immune globulin (SCIG) therapy.

Section 5004 of the act modifies reimbursement for drugs covered under the durable medical equipment benefit effective Jan. 1, 2017. The reimbursement will change from 95 percent of the first published average wholesale price to the average sales price (ASP) plus a 6 percent add-on, less a 2 percent reduction of payment due to the sequester. Applying the sequester changes, the actual reimbursement is ASP plus 4.3 percent. At this reimbursement rate, the cost to purchase an SCIG product exceeds the reimbursement for many specialty pharmacies. This means that access to an SCIG product is now limited by reimbursement, and many primary immunodeficiency patients may be required to switch back to intravenous IG, change specialty pharmacies or return to a hospital outpatient infusion center to continue therapy.

The second provision, Section 5012, allows the Centers for Medicare and Medicaid to reimburse “qualified home infusion therapy suppliers” for providing infusion therapy services in a beneficiary’s home for Medicare Part B and Part D beneficiaries receiving services at home. However, this does not take effect until Jan. 1, 2021. This provision will come with a broad list of new requirements and standards for suppliers of home infusion, and it will require Medicare to reimburse home infusion therapy suppliers based on a single, all-inclusive payment. In addition, as of this writing, it excludes payment for these services for Hizentra (CSL Behring) because the product is on the self-administered drug list.
Co-Pay Assistance

CSL Behring Raises Out-of-Pocket Co-Pay Assistance for Hizentra

CSL Behring has updated its Hizentra Co-Pay Relief Program for eligible patients to assist with out-of-pocket expenses. Launched in May 2014, the original program provided up to $4,000 in out-of-pocket assistance per patient per year. As of Sept. 12, the program now provides $5,000 per patient per year for existing and newly enrolled patients with no monthly cap for co-payments, deductibles and coinsurance. Eligibility guidelines are as follows:

• Prescription must be for primary immunodeficiency diagnosis, and patients must express financial need.
• Patients must be at least 2 years of age and must receive Hizentra through a specialty pharmacy or physician office.
• Patients must have coverage under a private U.S. insurance plan. The program is not valid for prescriptions eligible for reimbursement by any federal or state healthcare program such as Medicare, Medicare Advantage plans, Medicaid, PCIP, Champus, TriCare, Veterans Administration or any other state or federal program. In addition, patients whose insurance policies prohibit co-pay assistance are not eligible.

Those already enrolled in the program do not need to do anything to receive the increased level of assistance. Patients interested in enrolling must contact their specialty pharmacy or physician, express their need for financial assistance and ask the pharmacy or physician to enroll them via Medmonk (administrator of the program). Additional information can be obtained at CSL Behring’s IgIQ Resource Center at (877) 355-4447 or www.Hizentra.com/copay.
Plasma Safety

PPTA Says There Is No Risk of Zika from Plasma Donations

Based on robust virus clearance capacity during manufacturing of plasma-derived products and current regulatory guidance in Europe and the U.S., the Plasma Protein Therapeutics Association (PPTA) states it is assured that existing manufacturing methods are fully effective against Zika virus (ZIKV) and, consequently, the safety of plasma protein therapies is not affected by ZIKV. In addition, PPTA does not consider deferral or donation testing is necessary for plasma used for manufacturing into plasma-derived therapies.

According to PPTA, “ZIKV is of intermediate size (approximately 40-60 nm in diameter), has a lipid envelope and is therefore similar to other flaviviruses such as West Nile (WNV), dengue, yellow fever and Japanese encephalitis viruses. This group of viruses is highly susceptible to manufacturing steps with virus inactivation and removal capacity as typically used in the production of plasma-derived medicinal products such as caprylate- or solvent-detergent treatments, low pH incubation, pasteurization, dry-heat treatments, nanofiltration or plasma fractionation processes. The effectiveness of these processes has been clearly demonstrated using closely related lipid-enveloped model viruses belonging as ZIKV to the flavivirus family, e.g., bovine viral diarrhea virus, or tick-borne encephalitis virus or WNV. In addition, donor screening procedures make it highly unlikely that any person showing disease symptoms typical of ZIKV would be accepted for donation.”


Research

Researchers Call for Studies of IVIG for Treatment of ILD

Researchers at Harvard Medical School in Boston recently explored the use of intravenous immune globulin (IVIG) to treat interstitial lung disease (ILD) and found that while there is lack of evidence for its use, IVIG is increasingly being prescribed. Further, they recommend that clinical trials be conducted to determine IVIG’s effectiveness in treating ILD.

According to the researchers, IVIG use has expanded in recent decades to include treatment of autoimmune and inflammatory disorders due to its immunomodulatory effect, acting via neutralization of pathogenic autoantibodies, alteration of immune cell effector function, suppression of cytokine and chemokine activity and interference with activation of complement. Because ILD is frequently a complication of autoimmune disorders and connective tissue disease, IVIG is increasingly being used in its treatment. While no large studies are currently available to support the use of IVIG to treat ILD, it is being used off-label for refractory cases that have failed to respond to standard immunosuppression.


Duke Children’s Hospital in Durham, N.C., has established a Jeffrey Modell Diagnostic and Research Center, made possible by a partnership between the Jeffrey Modell Foundation (JMF) and CSL Behring. The new center will offer advanced diagnostic evaluation to patients with suspected primary immunodeficiency disease. John Sleasman, MD, chief of the division of allergy and immunology at Duke University School of Medicine’s Department of Pediatrics, will serve as the center’s inaugural director. For more information about JMF, visit www.info4pi.org.
“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
furry comfort

Animal-Assisted Therapy for Chronic Illness

From mood elevation to proven pain relief, research shows pet ownership could be a prescription for better health.

By Trudie Mitschang
ANYONE WHO HAS owned a pet will agree that animals are a great source of unconditional love and support. But, for a patient with chronic illness, a furry friend can provide much more than faithful companionship and a good cuddle. Studies show that animals can help with everything from lowering stress and blood pressure to battling depression and healing chronic pain.

“The evidence favoring the health value of pets is so compelling that if pet therapy were a pill, we would not be able to manufacture it fast enough,” says Larry Dossey, MD, author of *Reinventing Medicine*. Dr. James Rising, MD, DVM, a physician and veterinarian in Wallace, Idaho, agrees. In fact, he goes so far as to "prescribe a dog" for some of his patients. “Then I’ll tell them to go to the animal shelter and have it filled,” he says.1

While much of the medical literature focuses on the role of pet ownership in disease prevention, a number of studies have explored the benefit of animal companionship for people who live with a wide range of chronic illnesses and disabilities. In one study, researchers explored the relationship between pet ownership, AIDS and depression and found that people with AIDS who had pets reported less depression than those with AIDS who did not have pets. Other studies have uncovered equally positive effects of pet ownership on patients with Alzheimer’s disease, autism and mobility or hearing disabilities.2

The History of Animal-Assisted Therapy

Animal-assisted therapy (AAT) is the term most often used when animals are utilized as a therapeutic treatment option for disease or illness. Although AAT has gained increased acceptance in recent years, the concept is hardly new. Florence Nightingale is believed to have used animals in healthcare for the first time in 1860. She recorded that small pets were good companions, especially for the chronically ill, and described the benefits of an animal-companion as a source of therapy to her patients. She also used pets with wounded soldiers in the early 19th century and determined that pets were influential in the healing process.3

Sigmund Freud has also been credited with recognizing the role of animals in therapy. Freud believed the calming presence of his own canine was especially useful with the children he worked with, and was known for bringing his beloved chow chow into his therapy sessions.3

Another name linked to pioneering research surrounding AAT is Dr. Boris Levinson. A practicing child psychologist in the 1960s, Dr. Levinson theorized that his patients were less anxious and were less resistant to therapy when his dog, Jingles, was involved in the sessions. He noticed that one of his patients, who had previously refused to speak during sessions, would interact and speak to Jingles. Dr. Levinson proceeded to observe similar results in other children who had difficulty communicating. Based on his collective experience, he went on to author *Pet-Oriented Child Psychotherapy*, which later earned him the moniker “the father of AAT.”4

Pet ownership lowers blood pressure response to mental stress.

While much of the early research into AAT focused primarily on dogs, over time, cats, horses, birds and a variety of other animals have been used therapeutically, with equally promising results. According to Pet Partners, a national leader in demonstrating and promoting AAT, “Therapy animals aren’t just dogs. Cats, horses, rabbits, pigs, birds, llamas and alpacas, guinea pigs and even rats are eligible for evaluation through the Pet Partners program.”5

Citing numerous studies, the Pet Partners website touts the following patient-specific benefits offered by therapeutic companion animals:6

• Fibromyalgia patients spending time with a therapy dog instead of in an outpatient waiting area at a pain management facility showed significant improvements in pain, mood and other measures of distress.

• A walking program that matched sedentary adults with therapy animals resulted in an increase in walking over a 52-week graduated intervention, with participants stating their motivation for adherence was “the dogs need us to walk them.”

• The presence of an animal was shown to significantly increase positive social behaviors among children with autism spectrum disorder.

• Pet ownership lowers blood pressure response to mental stress.

• Pet owners have higher one-year survival rates following heart attacks.

• Pet ownership was associated with a reduced risk for non-Hodgkin’s lymphoma and diffuse large cell lymphoma.

• Human health savings of $3.86 billion over 10 years related to a decrease in doctor visits in studies in Austria and Germany have been linked to pet ownership.
Pets and Chronic Pain Relief

Nancy Gordon, founder of Paws for Comfort, has personal experience with the healing power of pets. A fibromyalgia sufferer who endured crippling chronic pain for years, Gordon found relief when she was introduced to a rare dog breed called the Xoloitzcuentli (Xolo), also known as a Mexican hairless. Gordon got her Xolo when she discovered the breed’s size and unique body warmth make it ideal for use as a living “heating pad” for muscle pain.

According to Gordon, the symptoms of her chronic fibromyalgia have been significantly lessened thanks to “Toaster,” her pet Xolo. She has since become an enthusiastic proponent of the breed, which has a centuries-old reputation for healing. “Before I discovered Toaster, my pain management required a microwaveable neck wrap around my neck almost 24/7. When I discovered Xolos, I was amazed at the healing power of their bodies when pressed against the skin for 15 to 30 minutes,” Gordon explains. “I believe that animals are angels for us, healing us in more ways than we can imagine,” Gordon says. “They provide us with compassion and unconditional love. They help us remember universal love and the pure pleasure of play, which can be forgotten by those tired and beaten down from a pain-filled day.”

Two articles on the National Fibromyalgia Association’s website note that Xolos’ usefulness in pain relief is due to their hairless nature, which allows their body heat to be easily felt — and to provide surprising warmth when placed against a person’s skin. Because Xolos are hairless, they are also hypoallergenic and odorless, making them excellent pets for anyone with asthma or allergies.

Using hairless dogs as heating pads is obviously an unconventional pain management solution, but additional research supports the benefits of animals in chronic pain relief (wearing the pet not required). In a 2012 study, it was concluded that therapy dog visits in an outpatient setting provided significant reduction in pain and emotional distress for chronic pain patients. The study also found therapy dog visits significantly improved emotional distress and feelings of well-being in family and friends accompanying patients to medical appointments.

More research is needed before AAT becomes a mainstream pain management solution, but with many studies supporting the use of pets in alleviating everything from chronic aches to post-surgical discomfort, AAT seems to provide a relatively easy and cost-effective intervention with no negative side effects.

The Feel Good Factor

Chronic illness and depression tend to go hand in hand. Whether a patient is having a bad day due to a physical setback or is battling a more serious bout of the blues, AAT may offer healing and hope. AAT is recognized by the National Institute of Mental Health as a type of psychotherapy for treating depression and other mood disorders. Being around pets appears to feed the soul, promoting a sense of emotional connectedness and overall well-being. “Some of the problems associated with depression are social withdrawal and feelings of loneliness,” says Steve G. Kopp, a licensed mental health counselor and marriage and family therapist with Genesis Health Systems. “Animal-assisted therapy gives a person a feeling of companionship and acceptance.” Animal therapy can have profound effects, Kopp adds, explaining that when patients soothe animals they are interacting with, they often end up soothing themselves.

There’s no question that snuggling a puppy simply feels good, but research also points to physical changes that occur within the body during patient/pet interactions. According to a 2012 paper published in *Frontiers in Psychology*, the increased oxytocin observed during human-animal interaction is proposed as the key reason for its positive psychosocial and psychophysiological effects. Oxytocin, a hormone secreted by the posterior lobe of the pituitary gland, is sometimes known as the “cuddle hormone” or the “love hormone” because it is released when people snuggle or bond socially, resulting in feelings of warmth and affection.

Science aside, there are several practical reasons why pets provide mood-boosting benefits. If chronic illness has caused mild to moderate depression, a pet can offer:

- **Uncomplicated love.** Chronic illness can significantly strain relationships with family and friends. A pet can offer judgment-free affection and will never say, “You don’t look sick.”
- **Responsibility.** Taking care of someone other than oneself
might feel daunting, but experts say that adding a little responsibility to one’s life can actually be helpful. Caring for an animal adds a new and positive focus to life while giving a sense of accomplishment.

• **Activity.** For those barely getting off the couch, a dog can give them a reason to take a brisk walk around the block.
• **Routine.** Having a daily schedule helps people with depression. An animal’s natural routine can offer much-needed structure.
• **Companionship.** Chronic illness is isolating. Having a pet is proven to ease feelings of loneliness and isolation.

### Safety Concerns for the Immune-Compromised

Patients who are immune-compromised can tremendously benefit from AAT, but safety concerns do exist. While each patient will need to assess, with their physician, the benefits versus the risks of pet interaction, it is a fact that infectious disease transmission from pets to people is a potentially dangerous possibility. For most patients, however, proper safety precautions can ensure a loving owner/pet relationship that offers more health benefits than hazards.11

One of the most common ways infections can pass from pet to owner is from contact with feces or urine. When cleaning up after a pet, patients should be sure to wear gloves, use a pooper scoop and make sure to wash hands thoroughly afterwards.

Patients will also want to prioritize their pets’ health. Regular worming is vital since many of the intestinal parasites pets get can infect humans, too. In a household with an immune-compromised member, monthly deworming is optimal. Animals should be treated against fleas and vaccinated regularly. It is also important to take animals to the vet at the first sign of illness, particularly vomiting, diarrhea, coughing and sneezing. Many of the bugs that cause these problems in pets can also pass to humans.

Patients with cats should try to discourage hunting, which could expose them — and patients — to any number of infectious diseases. For this reason, many doctors will advise keeping cats indoors.

Snuggling with a pet is one of the primary ways we show and receive affection. However, immune-compromised patients should discourage pets from “kissing” their face to avoid unwanted germ transmission. Likewise, they should be extra careful when it comes to pet bites or scratches. If an injury breaks the skin, the area should be washed and medical advice sought. Chances of injury can be reduced by avoiding rough play and regularly trimming pets’ nails.

Finally, pets should be chosen wisely. Some patients might gravitate toward more exotic pets thinking they may require less maintenance and interaction than a dog or cat, but pets such as reptiles are often infected with salmonella, while birds can be a carrier of Cryptococcosis and psittacosis, two highly infectious airborne respiratory illnesses.11

### Welcome Companionship

Life with chronic illness is difficult. In addition to the physical symptoms caused by the illness itself, there is the mental and emotional toll that comes from a dire medical diagnosis. Exploring the mood and health-enhancing effects of animal ownership can offer a welcome antidote to loneliness, mental distress and physical pain, while providing much-needed loving companionship. Whether choosing a shelter animal, a pick-of-the-litter puppy or a trained service animal skilled in AAT, adding a furry friend to a patient’s life might just make getting through the rough times a little bit easier.

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

### References

Fatigue is a common symptom of primary immunodeficiency, and while it is better understood today, its cause and treatment vary from patient to patient.

By Terry O. Harville, MD, PhD
IN THESE MODERN times, it’s common for people to complain about “fatigue.” Bright lights at night, television, computers, social media, telephones and other handheld devices, etc., all contribute to keeping us awake and have disrupted the normal diurnal rhythms of our bodies, particularly melatonin secretion in our brains. Yet, people with acute or chronic illnesses such as primary immunodeficiency disease (PI) complain about fatigue that is beyond the disruption of normal sleep — even when appropriate treatment has begun. What is the basis of this fatigue in patients with PI, and are there possible therapeutic interventions?

What Is Fatigue

The older definition of fatigue is “extreme tiredness, typically resulting from physical exertion.” This is distinguished from “malaise,” which is a medical term defined as a “feeling of weakness, overall discomfort, illness or simply not feeling well.” Thus, when most patients with chronic disease lament about fatigue, they are not exhausted due to physical exertion, but are suffering from the medical definition of malaise. However, since the newer definition of fatigue includes “mental exhaustion and tiredness due to illness,” fatigue and malaise may be used somewhat interchangeably.

Feeling fatigue while ill has always been an issue. When someone becomes ill, it is often recommended to “let the patient rest.” But, it is important to determine what is actually causing fatigue. For instance, many years ago, I helped to manage an athlete who competes in Ironman triathlons and runs 10 miles every day at noon in the Florida heat. She called me to complain about feeling fatigued, and I suggested she may be overtraining, causing her to become physically fatigued. But, she felt it was different than physical exhaustion, so I examined her and found signs of sinusitis. Two days after starting antibiotics, she called to tell me the fatigue she had experienced was gone. This and many other examples demonstrate that fatigue usually occurs when an infection is present. As such, those with arthritis conditions, autoimmune disorders or any state of immune system activation can experience fatigue.

THOSE WITH ARTHRITIS CONDITIONS, AUTOIMMUNE DISORDERS OR ANY STATE OF IMMUNE SYSTEM ACTIVATION CAN EXPERIENCE FATIGUE.

What Causes Fatigue?

The basis of fatigue in patients with illness was in large part discerned when HIV became an issue. Before effective therapy was available, most patients infected with HIV, and particularly those who developed AIDS, exhibited fatigue and frequently lost interest in eating. Sometimes, this was severe enough that
patients would waste away, and a condition known as “chronic wasting syndrome” was named. When animal studies were conducted, a protein found in the serum could result in “normal” animals becoming lethargic, seeking a warm, dark and quiet place, and not wanting to eat. If the serum was continually injected, the animals would not move about, would lose weight and eventually die from starvation. The serum factor was termed “cachexin,” since the general clinical features were those of cachexia (lack of activity with extreme weight loss). Eventually, cachexin was found to be tumor necrosis factor-alpha (TNF). When an infection occurs and the immune system becomes activated, TNF is one of the factors released early in the process. Thus, TNF can make a person feel tired and cold, have photophobia (light bothering the eyes), be bothered by noises and cause anorexia. This is highly relevant from an evolutionary perspective: If one can rest and heal from an illness, then one is more likely to survive, rather than wander about looking for food, thereby becoming a target for a larger predator.

### Fatigue in PI Patients

Today, more than 300 genes with mutations have been identified as having roles in causing immunodeficiencies. What is unexpected is most of the same gene mutations can also result in autoimmune disorders! Previously, it was thought that “overactivation” of immunity was the driving force behind autoimmune disorders, whereas “underactivation” of immunity led to immunodeficiencies. This implied that either different genes were involved or, if the same gene was involved, there were different mutations with opposite effects. Demonstrating that the same mutation in a gene can result in autoimmunity in some patients and immunodeficiency in others has obviously changed the paradigm. This helps to explain issues such as fatigue that are shared by those with autoimmune and immunodeficiency; in both cases, the immune system is likely inappropriately overactivated.

Another aspect of fatigue is “mental fatigue” or “mental fog,” even when a patient may not be experiencing physical fatigue. Patients describe mental fatigue as being unable to think clearly, having difficulty with decision-making and having faulty memory. It is now known that immune system-activating factors and cytokines such as TNF, as well as inflammatory byproducts produced in the brain, can contribute to this phenomenon. Thus, delirium during a fever is incited by the inflammatory cytokines. Reduction in inflammation can go a long way toward resolving delirium and body fatigue. Again, from an evolutionary standpoint, this cause and effect was likely to keep the mind from being active to promote rest until the illness resolved.

While things such as muscle strength can be measured in patients with weakness, fatigue is difficult to quantify because it is considered subjective. Fortunately, there are quality-of-life (QoL) questionnaires that can quantify the extent of fatigue in patients, as well as monitor changes over time. Unfortunately, these are not commonly used in the routine clinical setting; they are more typically used when conducting studies (for example, testing a new immune globulin [IG] replacement product). Therefore, while these questionnaires are helpful for indicating in those with medical conditions in which the immune system is not performing correctly.

Normally, once an infection has been cleared and the immune system dampens down the activated state, the factors and cytokines such as TNF that are promoting fatigue wane, and the fatigue resolves. Unfortunately, for those with states of chronic activation of the immune system such as in autoimmune disorders and immunodeficiencies, fatigue does not wane.

### Fatigue in PI Patients

Normal, once an infection has been cleared and the immune system dampens down the activated state, the factors and cytokines such as TNF that are promoting fatigue wane, and the fatigue resolves. Unfortunately, for those with states of chronic activation of the immune system such as in autoimmune disorders and immunodeficiencies, fatigue does not wane.
that fatigue is a major limiting factor for PI patients, practical use in the day-to-day management of PI patients is lacking for several reasons: 1) patients may not feel well enough to complete the questionnaire appropriately at each visit; 2) there may be staffing limitations in the clinic to administer and interpret the results; and 3) slight variations in successive visits may not be very meaningful, especially if visits are far apart. However, simplified versions of QoL questionnaires with fewer questions and use of a visual analog scale of perception of the disease state can be useful in successive visits, and many clinics do use these to help monitor patients’ levels of fatigue. These measurements can be especially useful when altering patients’ therapies to try to improve the level of fatigue.

Treating Fatigue

In patients with arthritis, anti-TNF medications have not only greatly alleviated symptoms and features of the disease, but have also greatly reduced fatigue in most. And, while this supports the role of TNF in fatigue in such conditions, there is a downside. Some of the medications may allow reactivation of latent tuberculosis or fungal infections. And, there is concern they may allow some cancers to develop. Therefore, the routine use of these types of medications solely for helping to reduce fatigue in PI patients who do not have a specific arthritis condition has not occurred because the risks may outweigh potential benefit.

At this point, there is no single successful therapy for alleviating fatigue in PI patients. For many patients, especially younger ones, merely introducing the correct treatment can improve fatigue levels. There could be a couple of reasons for this. One is that young patients have not acquired chronic indolent infections such as chronic sinusitis. Another is that they have not accumulated as many autoimmune-provoking components in their immune system as older patients have over time. Thus, the great burden of fatigue tends to be in adult PI patients, especially in those with mixed autoimmunity and immunodeficiency and/or chronic infections such as chronic sinusitis.

Treatment of the underlying components of disease is the first step to help ameliorate fatigue in PI patients. In some cases, this means giving higher doses of intravenous or subcutaneous IG than the minimal starting doses. Or, it may mean prolonged usage of antibiotics until chronic lung, sinus or gastrointestinal infections are cleared (or even continuing antibiotics indefinitely). A patient’s nutritional status should also be assessed. Improvements in dietary intake, especially supplementing nutrients that may be deficient, can be very helpful. Undertaking mindfulness exercises, participating in stress reduction, performing yoga, etc., can also be very helpful, even though fatigue may not be fully relieved. In some cases, antidepressant medications may be required. Interestingly, especially for patients with mental fog, medications such as Ritalin and Adderall that are commonly used to treat attention deficit hyperactivity disorder in children may be useful. Lastly, vitamin supplementation, including vitamin B12 injections and especially vitamin D, has received more attention as being beneficial in helping relieve symptoms of fatigue. This remains controversial, however, since there are no good widespread studies that indicate overall benefit.

When fatigue is not improved despite treatment, investigating additional issues may help identify treatable conditions that are causing fatigue. For example, has the patient developed diabetes mellitus or hypothyroidism? Does the patient have allergic disease or asthma that is not being fully addressed? Is the patient anemic? Are the liver and kidneys functioning normally? Has the patient’s uric acid level gone up (mild gout)?

Common, But Not Unsolvable

Unfortunately, fatigue is a common problem in PI patients and in patients in whom activation of immunity may be present. Many things can contribute to fatigue, including emotional distress from having a chronic disease. Therapeutic intervention must be individualized for each patient, with his or her physician willing to continue to investigate for potential causes that can be treated or otherwise ruled out as contributing to fatigue. Regrettably, despite the best efforts, patients may remain fatigued. This should not be taken as a sign to give up, but as an indication to delve further into the potential causes until optimal therapeutic intervention has been achieved.

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Improving Safety, Mobility and Activities of

A home safety assessment could help prevent health hazards for patients with physical and/or mental health conditions.

By Matthew D. Hansen, DPT, MPT, BSPTS
HOME SHOULD BE a haven, but too often, it unnecessarily becomes a hazard. This can be especially true for people with chronic illnesses and those who are returning home from a hospital stay.

One tool that can help make the home a safer place is the home safety assessment. Indeed, patients who initially think they don’t need one are often surprised and appreciative at the potential dangers discovered and the recommended modifications to resolve them.

A home safety assessment can be performed by discharging facilities or home health or hospice agencies that will be continuing care for patients. In addition, physical therapists, occupational therapists and nurses are uniquely qualified to perform the assessment should patients be fortunate enough to have access to one. If access is not available, following is a systematic review of items patients, family members and caregivers can consider as part of a home safety assessment, followed by several adaptations to make the environment safer if concerns are found.

**Outside the Home**

Safety at home begins outside the home. In fact, a 2006 study reported that “falls occurred outdoors more often than indoors among most age groups…. Most outdoor falls (73 percent) were precipitated by environmental factors, such as uneven surfaces and tripping or slipping on objects, and usually occurred on sidewalks, curbs and streets. Walking (47.3 percent) was the most common fall-related activity.”

Efforts to make the home and yard safer may require simple free adjustments, or they may entail modifications that cost a bit of time and/or money. Either way, they more than pay for themselves. The average national cost per day for an inpatient hospital stay is approximately $2,000 and can be much higher based on the reason for hospitalization. The average cost per day in a skilled nursing facility is about half the cost of a hospital. And, the average hospital stay after a hip fracture is more than 10 days. A lot of home modifications could be made with the money saved from not having to pay one large co-pay or health insurance deductible!

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**Outdoor Checklist**

- **Walkways and driveways**
  - Are they in good repair (e.g., no cracks or unevenness)?
  - Are they free of obstacles (e.g., garden hoses or fallen branches)?
  - Are they free of wet leaves, ice and snow?
  - Is there adequate lighting?
  - Is the slope of the driveway low enough to not be problematic?
  - Consider repair of walkways, use of flood and landscape lighting and behavioral changes (loading/unloading car only at the top of the driveway; driving to the bottom of the driveway or asking for help to pick up mail from the mailbox, etc.)

- **Steps/stairs**
  - Are walkways and porch steps clearly visible?
  - Are handrails present, sturdy and in good repair?
  - Are stair treads level and sturdy?
  - Are stair risers of equal height?
  - Consider placement of nonskid stair tread and/or reflective decals to mark each riser.

- **Ramps**
  - Are ramps sturdy, in good repair and free of obstacles?
  - Is there a minimum run-to-rise slope ratio of 12:1 (i.e., 12 inches of ramp length for every 1 inch of height)?
Is there a smooth transition between the ground and the ramp surface?
Do railings extend beyond the ramp to help with transitions?
Do ramps have nonskid surfaces?
Is there at least 36 inches between ramp handrails to accommodate walkers and wheelchairs?
Does the landing provide enough space to safely open the door without causing the patient to back up to the edge of the landing or onto the ramp?
Patients dependent on a wheelchair or two-handed device (e.g., adaptive walker) should have a ramp for at least one of the entrances to their home. Some local civic groups and other non-profit organizations may help to build ramps for people in need. There are also a number of metal commercial ramps that can be purchased to navigate one or two steps without breaking the bank.

**FOR THOSE WHO HAVE DIFFICULTY TURNING A DOORKNOB, CONSIDER REPLACING KNOBS WITH LEVER HANDLES.**

- **✓ Doors**
  - Are doorways wide enough to accommodate a wheelchair?
  - Are locks, latches and door handles in working order and easy to use?
  - Do doors open and close easily without sticking?
  - If doors are on springs, do they close slowly enough so they will not close on someone crossing the threshold?
  - Do doormats have nonskid backing without large cracks or upturned corners?
  - Are thresholds low enough not to cause a significant tripping hazard?
  - For those who have difficulty turning a doorknob, consider replacing knobs with lever handles. Decals may be placed at eye level on glass doors for patients with low vision or dementia to help prevent them from walking into the door (it may help keep birds from flying into the glass, too).

- **✓ Garages/carports**
  - Are cars easily accessible and walkways free of obstacles?
  - Is there sufficient lighting with an accessible light switch near the doorway?
  - Is the patient able to get into and out of the primary vehicle safely?
  - Auto transfer aids (e.g., Stander HandyBar) can help patients with decreased strength to get into and out of a car more easily.

- **✓ Other**
  - Are trees, shrubs and bushes pruned so they don’t encroach on walkways or cause low-hanging hazards?
  - Do porches/decks have railings to prevent the patient from stepping or falling off?
  - Are porch/deck floorboards secure with no protruding nails or large splinters?

**Inside the Home**
Areas inside the home can present a number of safety hazards besides fall risks. Some situations may not be dangerous to well-bodied people, or may not have been unsafe at another time during patients’ lives, but they may now present a threat because of physical and/or mental changes that have taken place related to their health conditions. Keep this in mind when reviewing this checklist.

**Indoor Checklist**

- **✓ Passageways and floor coverings**
  - Are floors and transitions between living spaces level and in good repair?
  - Is the patient able to safely navigate between doorways with or without assistive devices or a wheelchair?
  - Are hallways and pathways unobstructed of clutter and wide enough to allow passage?
  - Are indoor stairs and their floor coverings in good repair?
  - Do indoor stairs have sturdy handrails?
  - Is there sufficient natural and/or working artificial light in all areas?
  - Are there any fall hazards (e.g., rugs/mats, pets, cords, oxygen tubing)?

  - Floor rugs are common fall hazard culprits. If patients are not willing to have rugs taken up and put away, a nonskid backing can be placed between rugs and floors to help reduce slipping and sliding. Patients who use supplemental oxygen can request tinted oxygen cannula tubing versus standard translucent tubing that is notorious for being almost impossible to see until it’s too late. Those who truly are not safe negotiating stairs may have to consider moving their living quarters to the home’s main level, find a way to cover the costs of a stair lift or consider moving to a ground-level apartment or ranch-style home.
Electrical and fire
Are any fireplaces, lit candles or space heaters in use? Fireplace flues should be clean and unobstructed, and screens should be used when lit. Candles should be discontinued, and space heaters should be in good working condition.
Can the patient safely access and open/close locked windows?
Are there any electrical concerns in the home (e.g., overloaded outlets, electric cords with a short, hazardous placement of cords)?
If supplemental oxygen is in use, are “oxygen in use” signs placed somewhere visible to visitors before entering the home?
Does the patient or visitors smoke in the home when oxygen is in use?
Are oxygen tanks secure and kept in a safe location?
Is the stove in safe and working condition?
Is there a working fire extinguisher accessible?
Are smoke and carbon monoxide detectors present and in working order?

Bathrooms
Are toilets, sinks and showers/tubs working and accessible?
Does the patient have a walk-in shower, or do they have to step into a bathtub basin?
Do showers/tubs have a nonslip surface?
Are there floor mats that could cause a tripping hazard?
Is the water heater set to a nonscalding maximum water temperature?
Is there a night light present in the bedroom or hallway for bathroom accessibility?
Grab bars can be a lifesaver — literally — for those who experience impaired balance and/or strength. The Internet has measurement and installation guidelines that can direct someone in the proper orientation and placement. Bathtub transfer benches, shower chairs, handheld shower heads, long-handled sponges and a number of other adaptive tools can also help to make the bathroom a safer place.

General
Is the patient able to safely transfer to/from all seating surfaces in the home without using an unstable surface (e.g., a side table) for assistance?
Is there a phone that is readily accessible from a standing or crawling position (in case of a fall)? Can the patient use a phone functionally?
Are prescriptions kept in a safe place away from visitors (especially minors), and is the patient able to administer his or her own medications accurately?
If obtainable, does the patient have an extra supply of essential prescription medications in case of an emergency?
Are counters, sinks and cabinets safely accessible without having to reach high overhead or climb on top of something?
Is food available in the refrigerator and cupboards/pantry? Patients may also want to think about keeping a 72-hour emergency supply of food in different parts of the home in case of a natural disaster.
Are living conditions in the home sanitary?
If there are any firearms in the home, are they safely secured and stored?
A number of transfer assist devices can help patients get up from furniture, including sit-to-stand recliner chairs, hydraulic patient lifts, transfer poles and “bed canes.”

Grab bars can be a lifesaver — literally — for those who experience impaired balance and/or strength.

Safety First
Patients living with an immune deficiency, autoimmune disease, potential secondary complications and/or neuropathies already have a lot on their minds. However, thinking about safety first, whether it pertains to infection prevention or an unnecessary accident that could occur at home, will help to preserve health and well-being so that life can be enjoyed and other special considerations are dealt with in the most effective way possible.

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Reference

Grant First
Often misdiagnosed due to a lack of awareness, EDS is more common than autism, can coexist with any illness and is believed to affect the immune system.
SOME PEOPLE HAVE a list of health-related “justs” that they have grown up with. “My skin just bruises easily,” or “I’m just clumsy,” they might say. Maybe they’re “just tired a lot,” or they “just have migraines” or “just get dizzy” when they stand. Sometimes their heart “just races,” too. People who have struggled since childhood with these symptoms, and many more, often believe everyone else has them, too.

A healthy person would certainly say no, they do not have such a list. But for individuals with Ehlers-Danlos syndrome, a condition that few doctors understand, let alone are able to identify, this list is par for the course and rarely questioned — until symptoms worsen to the point of pain or even debilitation.

What Is Ehlers-Danlos Syndrome?

Commonly called EDS, this syndrome is a group of six genetic connective tissue disorders that often seem to masquerade as a variety of other illnesses. As a result, EDS patients spend an average of 20 years looking for a proper diagnosis.1 During that time, they are often misdiagnosed with conditions such as irritable bowel syndrome, fibromyalgia, multiple sclerosis, scleroderma, depression, anxiety, seronegative rheumatoid arthritis, osteoarthritis, chronic fatigue syndrome, asthma, hypochondria, lupus, Sjögren’s syndrome and other autoimmune disorders. Like its cousin osteogenesis imperfecta, EDS is sometimes even misdiagnosed as child abuse since joint dislocations and bruising during normal activities are not uncommon.2

EDS can coexist with virtually any illness. Although it is not an immune disorder, EDS is thought to affect the immune system. The theory is — and at this stage, it is only a theory — EDS can predispose a patient to acquiring an immune-mediated disorder. And, immune-mediated disorders can definitely be exacerbated by preexisting, and usually undiagnosed, EDS. Symptoms such as infections, diarrhea, malabsorption, fatigue and headaches can all be worsened by EDS since it can share many immune system disorders’ problems and further weaken the body.2

EDS is caused by heritable defects in how the body produces collagen. Despite its relative obscurity, it is not a new condition. Around 400 B.C., Hippocrates was the first to describe the symptoms. It wasn’t until centuries later, however, that Edvard Ehlers, in 1901, and Henri-Alexandre Danlos, in 1908, conducted further research and discovered that hallmark features of the disorder are skin extensibility and fragility.3

Since collagen is found in bones, skin, tendons, ligaments, muscles, teeth, organs, corneas and blood vessels, all of which can be affected by the disorder, symptoms are diverse. An undiagnosed or diagnosed EDS patient’s doctors are typically inexperienced in diagnosing or treating a connective tissue disorder that manifests in virtually every bodily system. A significant problem, too, is that only 5 percent of those who have EDS know about the condition. Patients suspect that something is physically wrong with them at times — or perhaps much of the time — but the symptoms they suffer from often seem to be entirely unrelated to one another.2

Heidi Collins, MD, who specializes in physical medicine and rehabilitation at Beacon Healthcare Medical Group, South Bend, Ind., is an EDS patient. She often quips: “If you can’t connect the issues, think connective tissues.” She explains that EDS is misunderstood and underdiagnosed because doctors are not unlike the parable of the four blind men touching an elephant. Each man feels something unique and real about the area they touch, but none is able to put all of the parts together as a whole. Likewise, doctors see individual symptoms and, unless they speak to each other about a common patient, are usually unable to diagnosis a unifying disorder.2
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— Marcia Boyle
President and Founder, Immune Deficiency Foundation

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Unexpected But Not Common

Some statistics indicate that between one in 2,500 and one in 5,000 people have some form of EDS.4 Clinical observation, however, shows that 3 percent of the population has hypermobility EDS, by far the most common type.7 This is more people than those diagnosed on the autism spectrum. While virtually everyone knows about autism, EDS is only now beginning to be better known in the medical community. Beyond the difficulty of recognizing the disorder by the untrained eye, it was thought for decades to be rare enough to require little to no study in medical schools. In fact, EDS is one of the few disorders to have a mascot, one it shares with the primary immunodeficiency disease community. The zebra represents the disease because of a somewhat folksy saying among doctors: “When you hear hoofbeats, think horses, not zebras.” In other words, doctors are trained to look for the most common conditions, not the exotic. Even so, more and more physicians and patients are learning that EDS is not that exotic at all.2

Other EDS Types

Beyond hypermobility, other varieties of EDS include the vascular and classic types, as well as the extremely rare kyphoscoliosis, arthrochalasia and dermatosparaxis types. Each has in common joint laxity, velvety skin, easy bruising and various systemic manifestations. Still, each type is a distinct condition that runs in families, affecting both men and women of all racial and ethnic backgrounds.6 Vascular type, seen in 5 percent of the EDS population, often results in a shortened lifespan; aortic dissections, ruptured organs and aneurysms are among its symptoms.7 Classic and hypermobility types, though disabling, do not shorten a patient’s life but often result in joint pain, poor healing, dysautonomia (an impairment of the autonomic nervous system) and POTS (postural orthostatic tachycardia syndrome).2

Overall, symptoms vary widely depending on the EDS type because the illness is a spectrum disorder, with some patients mildly affected and others completely incapacitated. The most common signs, however, include hypermobility of joints, fatigue, chronic joint and musculoskeletal pain, migraines, dizziness when standing, velvety or hyperextensible skin, unexplained stretch marks, various gastrointestinal problems, easy bruising, seemingly spontaneous joint dislocations (known as subluxations), scarring, tachycardia, hernias and, more seriously, aortic dissection and aneurysms.8

Only a few of the genes responsible for causing EDS have been discovered. Biopsies and blood tests are used to identify some rarer types. There is no genetic test available for hypermobility type, so physicians rely on family history and a clinical diagnosis.6

The Road to Diagnosis — And Why It’s Important

Shani Weber, 48, of Mount Airy, Md., has experienced doctors’ mishandling of her symptoms. At age 16, after a doctor told her she did not have Marfan syndrome (a related condition), he deemed her “fine.” She attributed her widespread pain to gymnastics training and went on with life, although her “just” list was significant. She says that over the years, she had “lots of diagnoses for each symptom separately. Each knee had its own diagnosis of instability and tendonitis. My blood pressure drops were diagnosed as orthostatic hypotension. My shoulder was diagnosed with bursitis. And on and on.”

Doctors could not connect her problems until after an accident, when her shoulder would not heal. “I spent countless hours Googling to try to learn why I was not improving. I stumbled upon EDS, and it was a perfect match for my lifetime of joint issues, extreme hypermobility and more.” After educating herself and her physician about EDS and waiting a year to see a geneticist (doctors who can diagnose are in short supply), she was finally properly diagnosed and treated.10

Since EDS mimics other illnesses, it’s crucial in some cases to determine whether a diagnosis such as rheumatoid arthritis, lupus, multiple sclerosis or another condition could instead be EDS — or perhaps even be compounded by EDS. Dr. Collins recalls a colleague who had informally diagnosed herself with ALS, or Lou Gehrig’s disease. The woman was in tears of relief after Dr. Collins properly diagnosed her with EDS. “Everybody needs to keep this disorder on their radar,” says Dr. Collins. “She went through a box of tissues because I was telling her Ehlers-Danlos syndrome was causing her weakness rather than Lou Gehrig’s disease. She, a physician herself, was completely unaware that her symptoms were a poster child manifestation of Ehlers-Danlos syndrome.”2

Diagnosing EDS is much simpler than many doctors realize with a physical examination and two diagnostic scales, the Beighton and
Brighton. “My then 11-year-old son accurately applied the diagnostic criteria to some of his elementary school fellow students,” Dr. Collins explains, “And, funny enough, one of the students separately appeared in my clinic. So, my 11-year-old can diagnose it. If somebody is obviously hypermobile and very obviously hypermobile on the Beighton or Brighton, then a physician or patient can look at those lists of questions and apply them pretty simply.”

The Beighton scale, most commonly used, is a series of flexibility tests. Patients are asked to bend their little fingers back beyond 90 degrees, push each thumb to their forearms, put their palms on the floor without bending their knees and hyperextend their knees and elbows. For each extension an individual is able to do, one point is given, for a total of nine points. A Beighton score of 4 or more, along with other symptoms such as chronic joint pain, is required for a diagnosis.

Despite the relative ease of identifying EDS, Dr. Collins understands the frustration many patients experience when trying to find answers for their various symptoms. Undiagnosed “EDSers” are often accused of doctor shopping because they consult so many specialists — usually with no definitive answers. Tests such as MRIs, CT scans, blood work and X-rays usually come back negative or normal, so EDSers are frequently labeled hypochondriacs, even though nothing could be further from the truth.

Treating EDS

Since EDS is a genetic disorder, it has no cure. Treatments for each type are palliative, designed to help patients manage the most debilitating symptoms. Physical therapy can help to strengthen muscles and joints, adding to their stability, but patients must be sure to see a physical therapist who is familiar with the condition or willing to learn about it. The wrong exercise or articular manipulation can damage a patient’s already hypermobile joints and muscles. Ring splints for fingers and other braces for major joints are commonly used to prevent hyperextension and, thus, further strain or damage them.

In extreme cases, surgery may be necessary to repair damaged ligaments and tendons. The risk for some patients is poor healing, especially with the classic type, because the skin is prone to scarring and is slow to recover.

Chronic pain is another struggle, but a doctor can help patients determine which pain medications and diets work best to manage it. Opioids are used only for acute pain, as long-term use can cause dependence. Migraines can be managed or even controlled with a variety of medications. Blood pressure, which is often low, can be improved with proper hydration, exercise, diet and various medications.

Though treatment is somewhat limited, many patients still find hope as they learn how to modify their lifestyles to remain healthy.

A Person’s List of “Justs” Could Be Significant

The “just” list that most EDSers have, whether or not they realize they suffer from EDS, usually means something despite a person’s occasional ability to rationalize it. “I haven’t met an EDS patient yet who doesn’t say, ‘This is how I’ve been all my life, so I didn’t think it was a problem,’” says Dr. Collins.

Symptoms could be a path to something significant, however. If an individual suspects that they or someone they know has a form of EDS, the next step is to learn as much as possible about the disorder. They should educate themselves, and then educate their physicians, or find physicians who are experienced in dealing with EDS. Comprehensive information on EDS types and symptoms can be found at ehlers-danlos.com and other EDS organizations. Being proactive in the search for answers just might spare people years of pain and many, many “justs” that could be treated rather than accepted or deemed mysterious.

Meredit Whitmore is an English professor and freelance journalist in the Northwest.

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LET’S TALK

Diagnosed with CVID in his 40s, Pete Atherton has not let his illness thwart his career goals, earning a doctorate degree and working in executive roles.

PROFILE: Pete Atherton

By Trudie Mitschang

Pete Atherton is a successful business executive, devoted husband, father and recent grandfather who refuses to let common variable immune deficiency (CVID) stop him from achieving his life and career goals. With a career path that includes a doctor of philosophy in computer graphics, and executive leadership roles at successful venture-backed technology companies, GE’s Global Research Center and two mid-size manufacturing corporations, this busy 65-year-old is currently working in retirement with a tech startup focused on commercializing a breakthrough solar energy technology. Known for his positive attitude and tenacity, Pete offers encouragement and inspiration that a chronic illness need not derail hopes and dreams for a promising future.

Trudie: You were diagnosed with CVID in your 40s. Were there any clues growing up that you were chronically ill?

Pete: I learned later in life that my parents saw me as a sickly child, but I was fortunate because I never heard that as a child. I was very active growing up and never thought of myself as different. I remember a couple of knee scrapes that got quite infected, and I also got a lot of mouth sores as a child. Looking back on my teens and 20s, I was able to go hard in both sports and studies, although there were a few hints along the way that something was wrong.

Trudie: When did your symptoms get worse?

Pete: In my later 30s and 40s, I started getting more colds that turned into bad sinus infections. I recall the congestion being bad, but the soreness of my sinus tissue was awful and sometimes bled. Then, the sinus infections turned into pneumonia, sometimes two or three times a year. On top of that, it was tough to get to sleep. I was finally diagnosed with CVID at age 44, and I will always be very thankful to the doctor who diagnosed it and changed my life for the better.

Trudie: What was your original treatment plan, and how has it evolved?

Pete: I remember getting the first immune globulin (IG) infusion in a doctor’s office, and I had such a bad reaction while driving home that I had to pull off the road. Early infusions caused spikes in fevers and bad shakes. This went on for several months until we got advice to start infusions at a low rate and slowly ramp up to a moderate rate. The infusions went from one-and-a-half hours to six hours, and it worked! I am also very fortunate to be married to a wonderful woman for 40 years who has degrees in microbiology and nursing. She has contributed a lot of medical knowledge over the years, and has been my infusion nurse for the past 10 years.

Trudie: Have you had any adjustments in medication?
Pete: Over the years, I have been through a few changes in medicine for various reasons. I have been on Privigen infusions for four years at the recommendation of my current specialist, who is great, and it has worked very well for me. Heading into Medicare, I tried HyQvia (subcutaneous IG), but it did not work as well for me as it has for others, and I have returned to Privigen.

Trudie: Were you recently approved for Medicare?

Pete: Yes. I was recently accepted to the Medicare IVIG Demonstration project that allows me to continue infusions at home. Medicare Part B covers specific diagnoses, and I was very fortunate that my CVID diagnosis is one of them. The warning I have heard is that it is covered today, but that could change in the future.

Trudie: In what ways has living with chronic illness been challenging personally and professionally?

Pete: I was on football, wrestling and tennis teams in high school, wrestling and rugby teams at Penn State, rugby in graduate school at Cornell and a member of GE’s national corporate relays team in my 40s. I also coached kids’ soccer and wrestling for several years. I think because of the tough training and competition from sports, I just tried to get tough and power through. What choice did I have? I still had to support a family, and work was challenging. Among other things, I learned to use DayQuil and NyQuil frequently, along with coffee, and I got antibiotics when needed.

Trudie: You’ve had a high-profile, demanding career. Have you ever felt the need to hide your illness from colleagues or business associates?

Pete: Yes! After my CVID diagnosis, I kept the treatments to myself, always doing them at home and away from work hours when possible. Since CVID is not contagious, I wanted to keep it as private as possible. I also knew the cost was high, and did not want to risk my job over it. Very few people ever knew about it, although some did see some of the effects it had on me.

Trudie: What was one of your biggest hurdles?

Pete: Changing jobs was very tricky because insurance continuation was critical to avoiding the preexisting condition issues. Getting new treatment required a local doctor’s approval, getting a prescription and lining up an infusion service. All of that had to happen in a few weeks to get a smooth transition from a different city hundreds of miles away. The issue is that if continuity is broken, then the insurance companies can re-evaluate. Apparently, this is also true with Medicare supplemental plans and, maybe, Part D since those are provided by insurance companies. There is a period of time when you first sign up that they can not refuse you or raise the rates, but if you sign up after that time frame or change insurance companies, then they have the option of refusing you based on preexisting conditions.

Trudie: Do you have a favorite quote or life philosophy?

Pete: I never thought of myself as limited, and I do not think about CVID or other health issues very much other than trying to maintain a healthy lifestyle. I would advise that it is one of those many challenges that life throws at you. Solve it as best you can like other challenges, and move on. I would also advise not feeling sorry for yourself. Strive to accomplish what is important to you, and enjoy life’s journey.

Trudie: For someone wanting to make career advances despite chronic illness, what advice do you offer?

Pete: While it is good to share things, especially with close friends and family, dwelling on negativity can drag relationships down, and after a while, people don’t want to hear it. If you put yourself in a manager’s position at work, do you think they are going to favor someone who focuses on doing a great job or dwelling on health issues?

Trudie: Many people feel limited by CVID. Did you ever feel tempted to not push so hard?

Pete: I never thought of myself as limited, and I do not think about CVID or other health issues very much other than trying to maintain a healthy lifestyle. I would advise that it is one of those many challenges that life throws at you. Solve it as best you can like other challenges, and move on. I would also advise not feeling sorry for yourself. Strive to accomplish what is important to you, and enjoy life’s journey.

Trudie: For someone wanting to make career advances despite chronic illness, what advice do you offer?

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Trudie: Do you have a favorite quote or life philosophy?

Pete: Work hard, accept challenges, but don’t forget to smell the flowers along the way. Time goes by too fast. Don’t let your medical condition hold you back.

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.
When the Impossible Happens, Is Anything Possible?

By Stacy Oliver

AS YOU READ this, there’s only a month or two until the start of a new baseball season. But as I write this, it’s still October, and the impossible has unfolded before my eyes. I am a native Chicagoan watching the Cubs win the World Series. This is crazy! The lovable losers of baseball battled it out against the Cleveland Indians for a title they haven’t had since Mark Twain (Samuel Clemens) was alive. Oh, and a little author named Tolstoy (you know, the guy who wrote War and Peace) was alive and answering fan mail. That was 100 years ago. It’s been 75 years since the Cubs have even played in the biggest game of baseball. For me, whether they won or lost the series didn’t matter; the fact that they played in the series had far more reaching consequences.

As a lifelong Cubs fan to actually say “I watched the Cubbies play on TV in the World Series” is surreal. Every year, at the end of baseball season, this is the team about which you’d say: “Next year is the year.” We had a curse hanging over us from a goat. It’s a long story, but believe me, it was taken very seriously. So, what happens when the impossible happens? Is anything possible?

If the Cubs could actually get to the World Series, what else can happen? Maybe it’s not so crazy to think that one day there could be better immune globulin (IG) treatments. Instead of accessing ports and hours of getting infusions with a pump, we could have IG lip balm! Nourishment for our lips on the outside, while helping us fight our diseases on the inside. I mean, while we are batting a thousand, let’s go for a home run and a couple of cures!

Why is this just coming to me now as I watch the Cubs win? The world has always been full of impossible possibilities. Take the city I was born in, Chicago; it burned down to the ground and was rebuilt to be one of the greatest cities in the world. If I could get my disease from out of nowhere, who is to say there isn’t a chance it could get better or maybe even go away? It might take another 100 years, but: “Next year is the year.”

It was beyond exciting to actually win the World Series, but for me, the thrill was making it there. That seemed to be the eternal joked-about goal, as if it ever really could happen. And, now that it has, there has been a shift in the universe for me. There’s no going back on my personal projects or life goals. No excuses in making my life happen, no matter how poorly I may be feeling. I have to play through the pain. Next year is now; it is happening. The Cubbies have shown me that, indeed, anything is possible if you believe long enough and persevere. I will be a fierce fighter of all my diseases and slug away at them with IG until I hopefully knock them out of the ballpark.

STACY OLIVER was diagnosed in 2008 with multifocal motor neuropathy (MMN). She is the assistant director of the Center for the Writing Arts at Northwestern University, and she is working on her supersecret identity as Neuropathy Girl, who will one day save the world after her infusion and a nap.
I STOPPED BEING scared of going to the doctor at a young age. If I learned one thing growing up with a primary immune deficiency disease, it was that infections don’t just “go away” on their own. They stick around. They get worse. They replicate until your throat is white or you’re too weak to get out of bed. I learned it’s important to go to the doctor to get medicine. Even though I might have dreaded the strep test or the exasperated way my pediatrician looked at me, I knew that if I wanted to feel better, I had to go.

I grew up. I had surgeries under general anesthesia. I had scopes for my gastrointestinal and sinus problems. When I developed an eye infection in my early 20s, I allowed a doctor to pull stitches out of my numbed eyeball while I was wide awake. I was not someone who was afraid of the doctor.

When dysautonomia joined my bandwagon of unwanted conditions, I got over any fear of needles I once had. I had to have fluids multiple times a week through an IV, so there was no room for phobia. So, it surprised me the other day as I sat in my car in my infectious disease doctor’s parking lot, that I was flooded with an unfamiliar sensation of anxiety. I had just come out of an 11-day hospital stay during which I’d had two surgeries: one to have my port pulled out due to infection, and another to correct the damage done by the surgeon who pulled out the port.

The first surgery was mind-blowingly bad. I was wide awake for the entire procedure. I was given topical anesthesia, but I felt every slice, dice, yank and knit. I screamed for the doctor to listen to me; I pleaded with her to knock me out. By the time the surgery was over, I was in complete shock, shaking, unable to comprehend the situation I’d just been put through. She sewed up the infected area, and two days later, I was back in the operating room getting the infected tissue removed and leaving a giant hole in my chest.

Just looking down at the bandages made my knees weak. But it was one bad surgeon. One bad surgery out of the many I’d already had in my life.

There is an element of trust we put in our doctors. We hand it over every time we sign a consent form, accept a new prescription or close our eyes as we’re wheeled into an operating room. We hand our bodies’ well-being over, believing we are being taken care of by people who know what they’re doing. It’s the same consent we give to pharmaceutical companies each time we try a new drug, or bare our arms for a phlebotomist to find the perfect vein. It doesn’t matter how seasoned we are as patients. We are all vulnerable to having our trust broken. This last incident turned me upside down and inside out, and, to say the least, I’m still learning how to cope with it.

What I do know is not talking to someone about it made my anxiety worse. And not speaking with my doctor about it after it failed to resolve on its own continued to make it worse. And while I do think the anxiety of the botched surgery will fade over time, it gave me an invaluable reminder: Just because we can take the punches — the tests, the tubes, the intense medications — it doesn’t mean we’re invincible. We have to treat ourselves with the same kind of compassion and care we would recommend for others.

ILANA JACQUELINE is a 27-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.

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IG Living | February-March 2017 | IGLiving.com 41
CHRONICALLY ILL CHILDREN are no strangers to hospitals. Whether for a visit to the specialist’s office or to the clinic for a regular treatment such as an intravenous immune globulin infusion, they’ve passed through those infuriating revolving doors so many times, it’s become old hat. So when it comes time for an extended hospital stay, chronically ill kids should be model patients, right? Not always.

It’s one thing for children to tolerate a hospital visit when they know they’ll be there only for an hour or two. But for something more involved, like surgery or a serious illness, the prospect of staying at the hospital for a lengthier amount of time can be scary, no matter how many times they’ve been there. Here are a few ways to make it a little easier for both children and their families.

Be honest. When children are scheduled for surgery or hospitalized due to an illness, it’s normal for them and their families to have questions. Parents can help their children cope with hospitalization by talking to them honestly about what is going to happen and why. For example, telling them that nothing will hurt or that there won’t be any blood tests isn’t going to help parents earn their trust. Instead, let children know that they aren’t alone, and that while a blood draw may hurt, it will help give the doctor important information to make them better.

Without overloading young kids with too much technical information, parents should try to answer any questions they have. If the answer is unknown, be sure to write it down and ask the doctor as soon as possible. When children are known to be easily affected by tension and anxiousness, stressful medical conversations should take place outside of their room.

It may also be helpful for parents to familiarize themselves with the illness or surgery by discussing it with the doctor ahead of time or researching online. The more parents understand about the tests and treatments their children will undergo, the more support they can offer during the hospital stay.

Visitors. When hospital stays are long-term, both parents and children can become bored. Visitors help to break the monotony and bring a little bit of home to an unfamiliar environment. Seeing a favorite relative or special friend can cheer up children who have been confined to their room for long periods of time. If kids are in an intensive care unit or under special precautions, like those for chemotherapy or bone marrow transplant patients, a nurse will need to be consulted about visitors. While they may not be allowed in the room, they can certainly visit with parents in the waiting area and pass along a card or gift.

Playing doctor. Younger children often benefit from having a doll or stuffed animal to practice medical procedures on. When our son Andy was 3 years old, he was admitted to the hospital for a long stay. Because he needed so many fluids and had multiple blood draws each day, a Hickman catheter was surgically implanted in his chest. The nurses at the hospital gave him a doll who had the same tubes (he called them “red and blue”) in its chest. When the lab technicians came to draw blood or give Andy medicine through his special tubes, he would do the same for his doll. This took his mind off what was happening to him while also making the entire process less scary.

Some hospitals provide toy doctor kits for their pediatric patients, which include pretend stethoscopes, syringes, blood pressure cuffs and many other items they can use to perform medical tests and procedures on their dolls or stuffed animals. This is a great tool to help children process what is happening to them and calm any fears they may have about their doctors. If a hospital stay is planned, these role-playing activities can be started several days ahead of time to help children feel more comfortable with what they are going to experience.

Bring a touch of home. Parents can’t always know when an extended hospital stay is in their children’s immediate future;
therefore, it may be difficult to pack everything that will make the stay more comfortable. When my son went to the emergency room in 2006, I had no idea we’d be loaded into an ambulance and taken to a hospital three hours from home, or that we’d be there for over a month. Needless to say, I brought nothing with me but the clothes on my back. But if parents do have time to prepare, there are some things they can pack that will help their children feel more “at home” while in the hospital.

Comfort items like a child’s own pillow, blanket, books and favorite stuffed animals help children feel secure and lend a sense of familiarity to a new and often sterile environment. Parents may also want to bring a pillow and blanket; it makes sleeping in a chair in a frigid hospital room or waiting room a little more pleasant.

Nonessential items such as noise-cancelling headphones, DVDs and downloaded movies on an iPad come in handy when the hospital TV only has three channels, or when children must share a room with another patient.2

It may be helpful for children to journal about their hospital experiences. Or, if children are too young to write, they could draw and color instead.1 Be sure to bring a notebook, journal or a pad of drawing paper, along with pens, crayons or markers.

Accept help from a child life specialist. In many hospital pediatric wards, there are child life specialists whose job it is to provide services to young patients and their families. These professional friends can be present with children prior to significant tests, surgeries and major medical procedures, and can offer counseling and comfort to families. For example, child life specialists can often be found wielding an iPad in the lab to distract an anxious child during a painful blood draw. They can also assist in the emergency room by teaching relaxation techniques to an anxious child. For children who are in the hospital for extended periods of time, a child life specialist may be involved in planning parties and activities for a child or his or her siblings.3

Parents can help their children cope with hospitalization by talking to them honestly about what is going to happen and why.

Even though it was 10 years ago that my two boys were hospitalized with complications from X-linked agammaglobulinemia, I still remember the family life specialist who worked with us. She brought games and toys to distract my older son while he underwent unpleasant procedures. She also entertained my daughter, who was simply along for the ride, with puzzles and crafts.

Long-term housing. For routine surgeries or short-term infections, children are often treated at their local hospital and the stay is limited to a few days. But for more complicated situations, patients and families may find themselves in a large hospital hours away from home for weeks or months. After a few sleepless nights spent on the recliner in the hospital waiting room with a major crick in the neck, they may be compelled to seek out alternative lodging. Hotels can be expensive, although many hospitals offer discounted prices for hospital guests. Many families have found comfort from special housing located near large hospitals such as the Ronald McDonald House that offer a place for families to rest and become refreshed for a very nominal daily fee.4 At least one meal per day is often provided, usually by volunteer groups.

This type of housing also offers a place for patients to stay when they are discharged from the hospital, but cannot go home because of frequent outpatient treatments. Entertainment, and in some cases education, is provided for siblings of patients, as well as weekly large-group events like scavenger hunts and movie nights for the whole family.

During our month-long stay at a large university hospital, we made the Ronald McDonald House our home-away-from home. Having a home base can make a lengthy hospital stay more survivable.

It’s never ideal to have a child in the hospital for any length of time, but as parents of chronically ill children know, sometimes it’s a necessary evil to keep them healthy. By following some of these steps, parents can help make the hospital stay, no matter how long or short, as positive an experience as possible. "

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

References
PRODUCT GUIDE

Understanding the ‘Pros’ of Probiotics

By Trudie Mitschang

BACTERIA. JUST THE thought of it brings to mind disease-causing microbes lurking in public restrooms or hovering over office cubicle desktops. But not all bacteria are bad — or bad for you. Probiotics, also known as beneficial or “good” bacteria, naturally live in the digestive tract and perform many essential functions to keep a body healthy. The term “probiotic” comes from the root words pro and biota, meaning “for life,” and is commonly used to refer to dietary supplements or foods that contain beneficial bacteria similar to those normally found in the body.

When used as a dietary supplement, probiotics are known to restore the balance of the intestinal microflora that can become unbalanced due to illness, stress, age, traveling or the use of certain medications. In recent years, there has been a growing public and scientific interest in probiotics and their potential health benefits. Researchers are studying whether probiotics taken as foods or supplements can help treat or prevent certain types of illness, including irritable bowel syndrome (IBS) and various digestive problems. While study results vary, there is encouraging evidence, especially for those with autoimmune and immune deficiency diseases, that probiotics may help minimize digestive track issues.

Probiotics Versus Antibiotics

Many patients with primary immune deficiency disease (PI) are frequent users of antibiotics. They are powerful aids in the fight against disease, but most antibiotics not only destroy harmful bacteria, they also wipe out the good bacteria the body needs to stay healthy. Many people who are regularly prescribed antibiotics know all too well that the positive effects of antibiotic treatment are often tempered by unpleasant side effects like diarrhea and IBS that may signal the antibiotics have upset the balance of good bacteria in the gut. Taking probiotics in food or as an oral supplement can help repopulate the beneficial bacteria and reverse the harmful after-effects of antibiotic treatment. Keep in mind the U.S. Food and Drug Administration does not regulate probiotics, so a doctor should be consulted prior to starting any supplement regimen.

Shopping Cheat Sheet

The names of the different strains of probiotics may initially seem foreign and very confusing. Although far from an exhaustive guide, here is a quick reference of the top probiotic strains to look for on product labels.

• *Lactobacillus.* This common probiotic is typically found in foods. It might also appear on a product label as acidophilus or L. acidophilus, or as Lactobacillus with another bacteria name such as bulgaricus. Lactobacillus is touted as a treatment for numerous health issues, including diarrhea, skin problems, lactose intolerance, high cholesterol, yeast infections and urinary tract infections. It is considered especially effective in treating diarrhea caused by viruses and medications.

• *Bifidobacterium.* This probiotic strain occurs naturally in the digestive system. Other label ingredient names may include B. bifidum, bifidus and B. breve. Bifidobacterium is recommended in supplement form for those who have been on antibiotics, and is therefore a strain that could be helpful to PI patients. Some studies suggest that Bifidobacterium can prevent intestinal infections and diarrhea caused by antibiotics.

• *Streptococcus thermophilus.* This friendly bacterium strain is found in fermented foods such as yogurt and mozzarella cheese. In the body, it helps treat gastrointestinal (GI) disorders and lactose intolerance.

• *Food-based options.* Many people associate yogurt with probiotics, but to get the full health benefits, make sure to look for yogurts with “live and active cultures.” Many commercial yogurts are heat-treated or pasteurized, resulting in the loss of these valuable cultures. There are also nondairy yogurt options containing live cultures that are made from rice, soy and coconut milk. Other dairy-free options for food-based good bacteria include brewer’s yeast, miso and sauerkraut.

Intestinal Wonder Workers

For PI patients who struggle with gut issues or who have been prescribed multiple courses of antibiotics, could a daily dose of good bacteria be the answer to improved health and wellness? Although still controversial in some medical circles, there seems to be increasing evidence that these intestinal wonder workers could hold the key to a more balanced GI track.

TRUDIE MITSCHANG is a contributing writer for *IG Living* magazine.
Saccharomyces boulardii is a probiotic yeast strain that survives passage through stomach acid and delivers its benefits to the intestinal tract. This strain helps protect and maintain a normal intestinal microflora, which in turn helps support intestinal health and immune response. It also works to restore the normal intestinal microflora when taking certain medications and during travel. $17.99; Vitacost.com

**Jarrow Formulas**
**Saccharomyces Boulardii Probiotic Supplement**

With a full line of age-specific products, Culturelle is one of the most popular probiotic brands in the U.S. Culturelle Digestive Health Daily Probiotic Formula contains naturally sourced ingredients that work with the body to support digestive health. Ingredients include Lactobacillus GG. 50 capsules $34.99; Walgreens.com

**Culturelle**

**Dr. Ohhira Probiotics**

Dr. Ohhira Probiotics contain 12 synergistic strains of probiotics that have been developed by a growth/production process that requires three to five years of fermentation. This extended fermentation process enables the strains of bacteria to grow and proliferate, and also results in the production of substantial metabolic byproducts and other accessory nutrients. Original formula (30 capsules) $28.35; drohhiraprobiotics.com

**Natren Healthy Trinity**

Each nondairy probiotic capsule contains a minimum of 30 billion colony forming units of three potent, super strains of beneficial bacteria. Healthy Trinity’s bile-resistant super strains are then released directly into the small intestine in the next stage of digestion, when bile breaks down the oil carrier and allows the beneficial bacteria to disperse through the gastrointestinal tract. $69.95; Natren.com

**Udo’s Choice Adult’s Blend Probiotic Capsules**

Udo’s Choice Adult’s Probiotic contains eight strains of adult-specific good bacteria. Each vegetarian capsule contains 12 billion viable cells at date of manufacture. 60 capsules $15.54; Amazon.com

**Kevita Cleansing Probiotic Tonic**

Described as a “deep hit of lemony sweetness combined with a tangy note of apple cider vinegar and a punch of citrus,” this probiotic tonic offers a drinkable method of ingesting good bacteria. Fermented with both apple cider vinegar and a proprietary water kefir culture, every bottle contains four strains of live probiotics. At health food stores nationwide and Kevita.com

These products are representative examples of what is on the market and are not endorsed as safe, effective or beneficial by IG Living. Patients are advised to check with their physician prior to introducing any products into their regimen.
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