Staying Active

How to Keep Moving with Chronic Illness

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Exercises for Better Balance

What Is Now Known About SAD?
Assistive Devices for Staying Mobile
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SUMMER IS fast approaching, which means warmer weather that entices us to go outdoors, become more active and, perhaps, take a vacation. Unfortunately, chronic illness can make activity more difficult depending on symptoms and their severity. That’s why, in this issue, we present several articles that can help you overcome obstacles that may prevent you from accomplishing the things you set out to do.

If taking a trip is something you are setting your sights on, don’t let illness deter you. Planning is the key to making your vacation safe and enjoyable. In our article “Travel Tips for Those with Chronic Illness” (page 16), we provide a host of suggestions for planning your trip before you go and while you are traveling — from making sure your destination is safe, packing medications, booking flights and hotels, and taking care of yourself by not overdoing.

If mobility is an issue, exercise and assistive devices can help you to move around more easily. For many with primary immunodeficiency and/or autoimmune diseases, balance is a problem. Matt Hansen, a physical therapist who specializes in motor development, outlines in his article “Exercises to Improve Balance” (page 20) the many exercises you can perform at home. He includes strengthening and functional exercises for ankles, hips and knees, as well as modifications to them and higher-level activities.

To help you move about more freely and gain more independence both at home and during travel, assistive devices may be the answer. We describe a number of different options in our article “Mobility Management” (page 42). In addition, we list a sample of products in our product guide for your perusal to help you keep doing the things you love.

Of course, in addition to these suggestions to help you stay active, many more educational and insightful articles are presented in this issue of IG Living.
IF YOU HAVE been diagnosed with chronic illness, chances are that diagnosis didn’t come overnight and it came with a big price tag. Chronic illness imposes a heavy toll of challenges that affect all parts of patients’ lives, from simply accessing appropriate care to paying for it. Unfortunately, one of the most common challenges chronically ill patients face is also frequently overlooked by family and the medical community: isolation!

Isolation is most often a consequence of no longer being able to work or interact with family and friends as you have in the past. Actually, the sense of isolation doesn’t happen overnight; it is a gradual process. Over time, you may receive fewer and fewer invitations from friends and family because your illness or physical limitations have caused you to frequently cancel plans in the past. Further, because of medical and other costs, you may not be able to afford to go out and do things you enjoyed before, even if you feel like it. Experts agree, it is extremely important for you and your loved ones to find ways for you to stay connected. If isolation is not dealt with, it can lead to loneliness and depression.¹

Here are some suggestions for preventing isolation:

1) Find a support group. Support groups, gatherings of people who share a common illness, offer many benefits. They provide a great way to learn more about your illness, connect with other people who are dealing with the same issues, and learn what resources are available for your particular disability. Oftentimes, these organizations hold in-person meetings and mixers, but there are also online support groups for times when you don’t feel like leaving the house.²

2) Use technology. Technology has created social networks that span the globe. The upside of technology is you don’t have to leave the house to stay connected with family, friends and the world through text, Facebook, Twitter and Instagram, to name a few.

3) Don’t give up on friends and family. Talk with them about your illness, and plan events that make allowances for your limitations. Be creative. Instead of going out to dinner and a movie, ask them to watch a movie with you at home and order takeout. Remember, it’s about the quality of the time you spend together versus the quantity.

4) Start writing a blog. Blogging is a great way to express yourself and share your experiences. Keep in mind you may be able to impart valuable information to another person suffering from the same condition.

5) If you are having trouble staying connected, find a therapist who specializes in patients with chronic illness. A therapist can help you find ways to cope with your illness and the changes it has caused in your life. If you are housebound, many therapists offer sessions via Skype or FaceTime.¹

Having a chronic illness doesn’t mean you are doomed to isolation. With a little work, preplanning and creativity, you can make lasting connections. The benefits are truly worth the effort. ■

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References
Have you thought about growing older?

I have so much wrong: kidney failure, multiple myeloma, myasthenia gravis, common variable immunodeficiency and so much more. My husband is 81, and it terrifies me that I might outlive him. I guess I am willing to keep my head stuck in the sand and ignore this possibility, live each day I have left to the fullest and enjoy the days that are halfway decent. Our son just drove us to Southern California for my nephew’s ordination into the ministry. We took the motor home so I could lie down. We enjoyed seeing my sister and all her family. Now, I will rest today. I guess I can only expect to live one day at a time, just like everyone else, and hope God sees fit to call me home before a nursing home decision is made. Death does not scare me; outliving my time is more worrying.

— Joy H-K

Yes, and I think we all have once we start hitting our late 40s. Unfortunately, I don’t think I will really make it to a late age, so [I am] loving and living life now while I can.

— Dave S

Yes, and I find it kind of scary. In my mind, going into a nursing home is no different than being in the hospital, because I would imagine both of these types of facilities would have the same challenges when it comes to a healthy environment. You definitely have given me something to think about.

— Jenny G

How do you respond to unkind remarks about your illness?

It’s hurtful, but what can we do? People say what they want to anyone these days, and they really don’t think about how it affects others. Most of the time, I don’t really care.

— Jennie B

Usually, I confront the person to clarify their meaning and intent. If it was intended [as unkind], then I would follow up 95 percent with a sharp response or, on a rare occasion, kill them with kindness.

— Dave S

It depends on who, what and why. Who said it? When was it said? Why was it said? That allows me to measure my response by which hat I have to wear. On some occasions, I have to wear the educator hat and then the caretaker hat. At its worst, out comes the witch hat.

— Janet S-D

Were you affected by the 21st Century Cures Act?

It would seem that if, as a patient, I need a certain drug or a certain way to administer that medicine, it should not even be up for discussion. I realize my thinking at times is fairy tale. I do not wish my disease on anyone, but I would really love to know how someone keeping a treatment from me would feel if this in some way impacted their life or the life of a loved one. I would love to pose that question to anyone coming between me and my treatments, but I know that is not realistic.

— Jenny G

I receive intravenous immune globulin infusions at home. We thought we were going to lose them in October 2017, but Congress came together, and the infusions are now paid through 2019. We now need to push the bill through to cover 2019 and 2020. It shouldn’t be so hard to cover the most vulnerable patients in this country.

— Lia F

» Join the conversation! Connect with other immune globulin patients through IG Living’s Facebook page at www.facebook.com/IGLivingMagazine. See our daily posts of interesting articles and facts, as well as thought-provoking questions that you can chime in on. Following are some snapshots of what’s being discussed.
Leslie » While you may have normal total IgG levels, if you are low in IgG2 and IgG3, you may have a functional (qualitative) deficiency. You need to see an immunologist to have a complete immune system workup. An immunologist can complete the appropriate vaccine testing, which would include measuring titers to determine if your immune system is working correctly. The usual vaccine testing process involves measuring prevaccine titers, giving the vaccine and then waiting four to eight weeks to measure postvaccine titers. However, depending upon how long ago you received the last pneumonia vaccine, the immunologist may be able to draw your titers without revaccinating.

Question » How Is an Immune System Problem Diagnosed in Someone with Low IgG Levels and Numerous Bouts of Pneumonia?

For years, my IgG2, IgG3 and IgM levels have been below the normal range. I have many respiratory problems, including pneumonia, for which I was in the hospital again two weeks ago. I was taken off of methotrexate that I take for rheumatoid arthritis. Yet, I had a physician’s assistant laugh at me and say I do not have an immune system problem. How low do my IgG levels have to be to have an immune system problem? Also, I have had pneumonia shots but I don’t know if they are working. My second-to-last case of pneumonia was a month after the shot.

Leslie and Dr. Harville » Radiofrequency ablation is akin to providing “microwaves” (like a microwave oven) to a very small, localized area. Use in the stomach to cauterize areas of bleeding should not be a problem for CIDP patients, although undergoing anesthesia and other aspects of an operation could be. These should be carefully discussed with the physicians involved.

Question » Could Radiofrequency Ablation Cause Problems in CIDP Patients?

I have chronic inflammatory demyelinating polyneuropathy (CIDP) and watermelon stomach with frequent bleeding. I have had several endoscopies and cauterizations to no avail. A new procedure known as radiofrequency ablation has been proposed, but I’m concerned it might cause nerve problems with my CIDP.

Question » Are There Cancer Treatment Guidelines for CVID Patients?

What are the medical guidelines for patients with common variable immunodeficiency who get cancer and need both chemotherapy and radiation? I know patients would discuss this with their immunologist and oncologist, but I would appreciate knowing what the guidelines are.

Have a question? Email us at editor@IGLiving.com. Your information will remain confidential unless permission is given.

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LESLIE J. VAUGHAN, RPh, is senior vice president of clinical programs at NuFACTOR Specialty Pharmacy.
DiGeorge Syndrome: Thymus Development and the Initiation of T Lymphocyte Development

By Terry O. Harville, MD, PhD

IN PREVIOUS issues, we discussed features of DiGeorge syndrome (DGS) and partial DGS (PDGS) resulting from the consequences of improper timing of the sequence of events during early phases of embryonic development. In this issue, we will discuss how maldevelopment of the thymus results in the immunologic problems associated with DGS/PDGS.

As previously discussed, the thymus develops from the normal layers of the skin. The skin infolds in the region of what will become the neck, and essentially “pinches off” so that what was the outer skin layer of the neck has become the inner core of the thymus, and the previous inner layer of skin of the neck is now the outer boundary of the thymus. Thus, overall, the thymus is essentially histologically indistinguishable from normal skin, but inverted. These skin-derived cells become known as thymic epithelial cells (TECs). Additionally, specific immune-related genes become activated in TECs, which allow them to help with the development of T lymphocytes.

The development of a T lymphocyte is a long journey, beginning in the bone marrow. The bone marrow contains unique cells, known as pluripotent stem cells (PSCs), that have the potential to develop into any cell the body needs. It is estimated adults have only about 10,000 PSCs, a very tiny percentage of the trillions of cells that make up the human body. After early embryonic development, PSCs primarily replenish the hematopoietic system, generating the red blood cells, platelets and white blood cells needed to function, grow old and die. PSCs are slow-growing and slow-dividing. When one does undergo a growth cycle and divides, one of the daughter cells remains as a PSC (to keep that population always present), and the other continues to divide, becoming more specialized along the way. The continuous divisions along the differentiation pathways expand the number of cells so that sufficient numbers can be achieved as they reach their mature developmental stage. The PSC daughter cell grows and divides a few times, and becomes what is known as a multipotent stem cell (MSC). The MSC commits to develop along specific pathways (it is thought it cannot go back to becoming a PSC, under normal circumstances). Most typically, the MSC will develop into cells that replenish the needed hematopoietic cells.

Following the MSC stage, after further growth and divisions, greater cell lineage commitment is achieved. A myeloid-erythroid progenitor cell type (MEPC) and a monocyte-lymphoid progenitor cell type (MLPC) arise. The MEPC further grows and divides, with further lineage commitment, into myeloid progenitor cells (MPCs) and erythroid-platelet progenitor cells (EPCs). MPCs grow and divide, giving final lineage development into primarily neutrophils, basophils and eosinophils. EPCs grow and divide, developing into red blood cells and megakaryocytes. The megakaryocytes become the “factories” for generating platelets.

At some point, as the MLPC is further developing, cells that will become monocytes/macrophages diverge from the lymphoid lineage, and the lymphoid-lineage cell (LLC) emerges.

The LLC lineage further grows and divides, committing to the development of B-lineage and T-lineage lymphocytes. The B-lineage lymphocytes remain in the bone marrow to complete their development into mature B lymphocytes to provide antibody production. The T-lineage lymphocytes exit the bone marrow and traverse to the thymus to undergo their development. Some of the T-lineage lymphocytes do not enter the thymus and become natural killer cells.

We will continue the development of T lymphocytes in the thymus with our next discussion.

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.
What Are Immunoglobulins?

By Michelle Greer, RN

THE IMMUNE SYSTEM is a complex network of cells, tissues and organs that protect the body from bacteria, virus, fungi and other foreign organisms. The primary functions of the immune system are to recognize self (the body’s own healthy cells) from nonself (anything foreign), keep self healthy and destroy and eliminate nonself. Immunoglobulins take the lead in this process.

A Review of Terminology

Understanding a few related terms and their function can provide a better appreciation of immunoglobulins and how they contribute in the immune system.

An antigen is anything foreign entering the body such as bacteria or a virus, or in cases of transplant, another person’s organ, tissue or cells. Antigens are identified by the immune system by a marker molecule, which enables the immune system to differentiate self from nonself. Lymphocytes (natural killer cells, T cells and B cells) are one of the subtypes of white blood cells in the immune system. B cells secrete antibodies that attach to antigens to mark them for destruction. Antibodies are antigen-specific, meaning one antibody works against a specific type of bacteria, virus or other foreign substance. Just as every lock has a single key, an antibody has a single antigen key.

When a B cell encounters its triggering antigen, it gives rise to many large cells known as plasma cells. Every plasma cell is essentially a factory for producing an antibody. Antibodies are also known as immunoglobulins. Antibodies, or immunoglobulins, are glycoproteins made up of light chains and heavy chains shaped like a Y (Figure 1). The different areas on these chains have different functions and roles in an immune response.

Types of Immunoglobulins

There are several types of immunoglobulins and each has a different role in an immune response (Figure 2). Immunoglobulin G (IgG) is the most predominant antibody found in the blood, intestine and lymph. It comprises approximately 75 percent of all immunoglobulins. It also is the major antibody involved in an immune response, including neutralization of toxins, viruses and bacteria. It is the only antibody that can cross the placenta. IgG also has four subclasses: IgG1, IgG2, IgG3 and IgG4.

Immunoglobulin A (IgA) is the second most common antibody and is found in bodily secretions such as mucus, saliva and tears. Due to this, IgA can provide local immune protection.

Immunoglobulin M is the third most common antibody and is expressed first in an immune response and, therefore, assists in neutralizing antigens in the early stages of infection. It is found mainly in the blood and lymph.

Immunoglobulin E is found in very low levels in the body and plays a major role in allergic responses.

Immunoglobulin D was the last immunoglobulin to be discovered, and little is known about its function.

A deficiency in any component of the
immune system, including immunoglobulins, may result in an immune deficiency. There are over 300 types of primary immunodeficiencies, which are inherent genetic problems with the immune response. Other immunodeficiencies are secondary, meaning they are a result of specific external factors such as medication or malnutrition. Treatment for immune deficiencies varies greatly, and can include treating the resultant infections, immune globulin therapy and stem cell transplant. At present, most primary immunodeficiencies can be medically managed but not cured. Fortunately, in some cases, a secondary immune deficiency is reversible if the causative factor is identified and addressed.

A malfunction in any component of the immune system can result in an autoimmune disease. Malfunction occurs when the immune system sees self as nonself and creates an autoantibody to destroy it. This attack can occur in any system in the body (Table 1). There are many treatments for autoimmune diseases, but rarely a cure. Treatments, which include steroids, immunosuppressive medications, immune globulin, rituximab and other monoclonal antibody medications, suppress or modify the immune response to relieve symptoms.

A Complex System

The immune system is a multi-faceted defense system that protects the human body from foreign invaders, with immunoglobulins playing a major part in immune function. Even so, this system can malfunction, turn on itself and result in disorders in different parts of the body. Although much has been learned about the immune system’s components and their roles in disease and treatment, the need for more study is ongoing. The latest findings about immune deficiencies can be obtained from the Immune Deficiency Foundation at www.primaryimmune.org. More information about autoimmune diseases can be obtained from the American Autoimmune Related Diseases Organization at www.aarda.org.

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

References
Researchers in the Bone Marrow Transplant Department at St. Jude Children’s Research Hospital in Memphis, Tenn., believe they have found a cure for severe combined immunodeficiency (SCID) using a new gene-based therapy. The treatment uses an inactivated form of HIV to introduce genetic changes into the patient’s bone marrow cells so they start making B, T and natural killer (NK) cells. The researchers chose HIV because the virus naturally evolved to effortlessly infect human immune cells. Previous versions of this gene-based cure used a different mouse-derived virus, which tended to activate cancer-causing cells and produce leukemia in patients. With the HIV virus, it appears the immune system is being successfully infiltrated.

In addition to the HIV virus, babies undergoing this new gene therapy also undergo chemotherapy with the drug busulfan to prepare their bone marrow to accept the genetic changes. While doctors have previously been reluctant to use chemotherapy in SCID patients since they don’t produce any immune cells, babies with X-linked SCID only achieved a partial cure when they received gene therapy treatment without chemotherapy; their T cells returned, but not their B or NK cells. The combination of the new gene therapy with the chemotherapy appears to have restored all three types of immune cells in babies.

Six out of seven infants treated using this new gene-based therapy already are out of the hospital and leading normal childhoods, according to lead researcher Ewelina Mamcarz, MD. “They left the hospital after four to six weeks, and we’re following these babies on an outpatient basis,” she said. As of this writing, the last infant was barely six weeks past treatment, and his immune system is still in the process of constructing itself.

The oldest patient was about 15 months.

In response to reports by individuals with primary immune deficiencies (PIs) of lower-than-average body temperature, the Immune Deficiency Foundation (IDF) is conducting a study to determine if this is the case. Currently, no known literature exists on average body temperature in persons with PI. However, when PI patients have an infection, their temperature frequently does not rise to 100.4 degrees Fahrenheit, the threshold upon which doctors begin to prescribe antibiotics, which means they may not receive critical antibiotics due to missed signs of a fever.

The IDF-funded study is looking for households in which one adult with PI and one adult without PI, both of whom are 21 years of age or older, who are willing to participate. In the study, both the PI and non-PI adults will be asked to take their oral temperature three times per day on five consecutive days and record it in a paper logbook provided by IDF. A digital oral thermometer that participants may keep will also be provided. Participants will also be asked to disclose what medications, supplements or therapies they are taking those days and how they feel in general. At the end of the study, participants will be asked to return the logbook to IDF in a prepaid envelope. Households participating in the study will each receive a $20 coupon code that can be redeemed at Amazon.com.

To determine if a household qualifies for the study, a brief online screening survey must be completed by both individuals in the household. IDF’s goal is to enroll 350 people. For more information, go to primaryimmune.org/news/participate-idf-fever-study.
Medicines

GigaGen Receives Grant to Develop Production Technology for rIVIG

GigaGen Inc. has been awarded a $3 million grant from the National Institutes of Health (NIH) to advance its next-generation plasma therapy for patients with primary immune deficiencies. The funds will be used to develop a natural repertoire antibody protein expression system to lay the foundation for a recombinant intravenous immune globulin (rIVIG) therapy that could overcome many of the limitations of current plasma-based IVIG options such as limited human plasma supply and investment in expensive, large-scale purification facilities.

“Immunoglobulin replacement therapy has, for decades, been a critical treatment for patients with primary immunodeficiency,” said Dave Johnson, PhD, CEO of GigaGen. “In line with our mission to create antibody therapeutics that improve long-term outcomes for patients with severe diseases of immune dysregulation, we aim to significantly improve upon today’s standard plasma therapy with a product that is safer, more consistent and unconstrained by donor supply. This NIH grant will enable us to continue to move swiftly toward our goal.”


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12th Annual Neuropathy Action Awareness Day Is June 22

The 12th Annual Neuropathy Action Awareness Day will be held Friday, June 22, at the InterContinental Hotel in Los Angeles. This largest neuropathy awareness and education event in the U.S. provides an opportunity for patients to interact with other patients, providers and exhibitors, as well as to learn about neuropathy and how to cope with the disease, policy issues and patient advocacy. The day begins with an exhibit area and educational sessions in the morning, followed by a luncheon and additional educational sessions and exhibit area with refreshments in the afternoon. Additional features include a celebrity speaker, elected officials and a silent auction that includes trips, activities and other fun items.

To ensure as many patients can participate as possible, the Neuropathy Action Foundation (NAF) has booked a block of rooms at the hotel for a special, reduced rate. For those with a financial hardship, NAF will pay for up to 10 flights and hotel rooms for the night of the event or the night before the event for patients from outside the Los Angeles area. In addition, those unable to attend in person can listen free of charge via Livestream technology using a computer with an Internet connection. Those individuals will also be able to ask speakers questions and receive answers in real time. The event will be recorded, so individuals can watch the event even after it has ended.

To register, go to the NAF website (neuropathyaction.org) and click on “register today” on the right side of the homepage. Once registered, those watching via Livestream will receive a link to view the event. Registration is $25 for patients and caretakers for the event and luncheon. For nonpatients and noncaretakers, the event is $125 per person. The event is free for Livestream participants. Registration after June 9 is an additional $50.

Research

Study Finds Differences in Gut Bacteria Between CFS Patients and Healthy People

A study has found that individuals with chronic fatigue syndrome (CFS) have higher levels of certain gut bacteria and lower levels of others compared with healthy people. In addition, patients have different patterns of gut bacteria disturbances depending on whether they have only CFS or both CFS and irritable bowel syndrome (IBS).

In the study, researchers analyzed fecal samples from 50 patients with CFS and 50 healthy people, and found nearly half (21) of CFS patients also had IBS. They also found that differences in the levels of six types of gut bacteria — Faecalibacterium, Roseburia, Dorea, Coprococcus, Clostridium, Ruminococcus and Coprobacillus — were strongly linked with CFS. In fact, the abundance of these species in participants’ guts could be used to predict whether patients had CFS. Patients with CFS and IBS also had higher levels of a bacteria called Alistipes and lower levels of Faecalibacterium. And, those with CFS but not IBS had higher levels of a bacteria called Bacteroides but lower amounts of a specific species of the genus called Bacteroides vulgatus. According to the researchers, these findings could aid in the diagnosis and treatment of the disease.

Research

Study to Evaluate New Drug in Myasthenia Gravis Patients

A Phase II proof-of-concept study will evaluate the safety, tolerability, efficacy, impact of quality-of-life and assessment of pharmacokinetics and pharmacodynamic markers of ARGX-113 on up to 24 myasthenia gravis (MG) patients. ARGX-113 will be dosed on top of current standard of care, corticosteroids and/or immunomodulatory agents. Topline data from the study is expected in the second half of 2018.

ARGX-113 is manufactured by argenx, a clinical-stage biopharmaceutical company focused on creating and developing differentiated therapeutic antibodies for the treatment of cancer and severe autoimmune diseases. “MG is a rare and debilitating muscle disease with limited effective and sustainable treatments. ARGX-113 has the potential to eliminate patient symptoms while minimizing common side effects seen with current treatments by reducing the pathogenic IgG levels,” said Nicolas Leupin, CMO at argenx. “The initiation of this Phase II study is an important milestone in understanding how ARGX-113 can be effective in a wide range of IgG-mediated autoimmune diseases, including additional orphan indications and larger indications like multiple sclerosis and lupus.”


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Travel Tips for Those with Chronic Illness

Patients don’t need to be deterred from taking a vacation just because of their illness, as long as they plan accordingly.

By Abbie Cornett and Ronale Tucker Rhodes, MS
VACATIONS ARE A great way to recharge your batteries and relax, but they take planning, even for healthy people. For those who suffer from chronic illness, planning is particularly important. How much planning is needed depends on the severity of your illness and your travel destination. Just be sure to give yourself plenty of time to avoid getting overwhelmed with the logistics. Following these tips can help you have a safe and enjoyable trip.

Before You Go

Before making travel plans, talk with your doctor to make sure it’s safe for you to travel. And, ask whether any travel restrictions are required due to your illness. For instance, depending on the severity of your condition, it might not be safe to travel to certain areas of the world.

If you are planning a trip abroad, the Centers for Disease Control and Prevention (CDC) recommends a visit to a travel medicine specialist at least four to six weeks prior to departure to discuss what steps are needed to prepare. In some instances, you may need to be vaccinated. Most vaccines are inactivated, which means they contain the killed version of the virus that causes the disease. The drawback of inactivated vaccines is they don’t provide immunity (protection) as strong as live vaccines, so several doses are typically required. Further, inactivated vaccines may not be as effective for those with weak or compromised immune systems. Your doctor can perform a blood test to determine whether the vaccine was effective for you.

Individuals with a weakened immune system should not receive live virus vaccines that contain a weakened form of the virus such as the measles, mumps and rubella vaccine and the yellow fever vaccine. This may limit where you can travel. CDC recommends you check its Travelers’ Health website (wwwnc.cdc.gov/travel) to view which vaccines are recommended at your intended destination.

Whether you are planning travel abroad or in the U.S., you should always have a copy of your health history information sheet (HHIS), and it should be kept with you at all times. The HHIS should include:

- Medical diagnosis
- Physician contact information
- Medications and dosages
- Emergency contact information

When packing your medication, use a pillbox. A change in daily routine increases the risk of forgetting to take medications or doubling up on them. A pillbox can also ensure you pack enough for the entire trip. It’s also recommended that you include extra medication to cover delays caused by missed flights, bad weather, etc. A good rule of thumb is to pack for at least two extra days. You should also have a copy of all your prescriptions in case you run out of or lose your medications.

If you’re flying, put all your medications (in their original packaging), supplies and medical equipment in your carry-on bag. Keeping these essentials in your possession minimizes a bad situation if the airline loses your baggage. You can’t count on being able to purchase your medications, particularly if you are traveling outside the U.S. For answers on how to properly transport health-related items when flying, visit www.tsa.gov/travel/special-procedures.

In addition to your medications, insurance is of upmost importance when traveling. Keep in mind that for a trip abroad, most health insurance policies don’t cover you outside the U.S. Before you leave, find out what your policy covers, and then purchase additional travelers health and evacuation insurance if needed. Insurance companies and credit card companies frequently offer these types of policies, and many even include medical flights.

Insurance for trip cancellation should also be considered. Because of the uncertainties involved with chronic illness, you need to allow for the possibility that you will be unable to travel at the last minute. To protect your investment, airlines, cruise lines and tour agencies usually offer this type of insurance.

Transportation and Accommodations

To reduce the stress of traveling, try not to rush. The more tired you are, the less you will be able to do once you get to your destination.

Planes, trains and automobiles can take their toll. If flying, don’t book connecting flights close together. Give yourself ample time to get from one gate to another without running — even if it means longer layovers. And, don’t be afraid to ask for help. If you have mobility issues, contact the airline prior to departure to ask for wheelchair assistance. A wheelchair attendant can take you from the curb through security and to your boarding gate. And, if traveling with others, they can stay with you through security. An attendant can also transport you between gates for connecting flights.

If traveling by train, Amtrak (www.amtrak.com/accessible-travel-services) has accommodations for travelers with disabilities, including special rooms and cars for those who cannot negotiate stairs on the cars.

No matter what your transportation mode, pack healthy snacks. Tired and hungry are a losing combination. And, be sure to drink plenty of water to stay hydrated. You can purchase
water in airports to mix with electrolyte drink such as individually sealed powder packs of Propel, Gatorade or other electrolyte drinks.

If you are doing a lot of traveling, you may be required to sit for extended periods of time. If driving, stop every two hours for a 10-minute stretching break. If flying or taking the train, take a walk to the restroom. You can also perform arm and neck stretches in your seat.4

Also, make sure the people you are traveling with are well aware of your illness. When driving, plan your route in case you may need to make frequent bathroom or rest stops along the way. If you’re prone to car sickness, carry a plastic bag or garbage bag and Kleenex in case you’re nauseous.

When booking hotels, consider those that will accommodate your special needs. Some ideas include:

- The convenience of a hotel with a restaurant or room service can conserve precious energy at mealtimes in case you have a flare during your stay.
- A refrigerator in the room can store food, drink and medication.
- A room located near the elevator and on a lower floor allows hotel staff to help you more quickly in case of emergency.

According to the Americans with Disabilities Act, all public accommodations in the U.S. must comply with basic nondiscrimination policies. For hotels, this means they must provide barrier-free rooms and bathrooms and barrier-free access both inside and outside of the building. Other things you may want to request include raised toilet seats, grab bars, tub chairs and walk-in showers.5

Take Care of Yourself

Just because you’re on vacation doesn’t mean you have to do everything. When you’re tired, it’s OK to stay at the hotel and rest while your travel companions go on that extra excursion. And, don’t allow yourself to feel sad you can’t do everything you might have wanted. It’s better to be healthy and do some things than to try to do everything and end up sick and unable to do anything. Importantly, be sure to get your eight hours of sleep.

Eat sensibly. Part of traveling is the adventure, but the best time to experiment is not when you’re out of town. Avoid overindulging and eating anything that could make you sick. For example, if you want to experience new cuisines, only do so one meal a day, and stick with simple fare for the other two meals. If possible, take along your own instant oats, breakfast bars, and nuts and dried fruits for breakfast.6 Look for restaurants, health food stores, supermarkets and other places where you can purchase food that you are used to eating. Carry energy bars for times you are somewhere food is unavailable. And, once again, be sure to drink a lot of water to keep hydrated.

If swimming is on your activities list, follow these guidelines from CDC:6

- Don’t swallow the water you are in or on.
- Don’t swim with open cuts, abrasions or wounds. Breaks in the skin can let harmful germs into your body.
- Don’t swim if you have diarrhea.
- Don’t swim in cloudy water.
- Be careful about swimming or wading in fresh water. In some countries, infections such as schistosomiasis and leptospirosis are spread by contact with fresh water.

Lastly, to be prepared for the unexpected, have a bag ready in case you need to go to the hospital. Your I.C.E (in case of emergency) bag is what most individuals with chronic illness keep packed for emergency trips to the hospital. It usually includes water, snacks, books, pajamas, clean underwear, slippers and a blanket.

Vacations Are Supposed to Be Fun, Not Work

Plan for pleasure by looking at travel brochures before you depart. Choose what looks like the most fun. Don’t feel like you need to see and do everything. And, relax and rest when you need to take a break. It’s not about the quantity of things you do but more about the quality. Using these travel tips will go a long way to ensuring your vacation is both enjoyable and safe!

ABBIE CORNETT is the patient advocate and RONALE TUCKER RHODES, MS, is the editor-in-chief for IG Living magazine.

References
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Strengthening and functional exercises can help improve balance problems related to an immunodeficiency or autoimmune disease.

By Matthew D. Hansen, DPT, MPT, BSPTS

THE BAD NEWS is there isn’t always a clear-cut solution to improving balance. This is because a number of factors can contribute to poor equilibrium, including conditions that impact the inner ear and vestibular system; central nervous system involvement (e.g., cranial nerves, the cerebellum and other regions of the brain); neuropathies of the peripheral nerves; genetic and acquired neuromuscular conditions; impaired vision; alterations in blood sugar and/or blood pressure levels; and reduced muscle strength due to deconditioning. Nonetheless, the good news is even though the solution may not be straightforward, just about everyone can develop better balance.

Improving balance often begins with a visit to a primary care physician to investigate the root cause(s) of the issue. Patients should not be afraid to share their own impressions and ask plenty of questions. A good health interview specific to the complaint should be conducted, including asking if/when dizziness or vertigo occurs and whether the patient can describe any possible patterns or contributing factors. The physician should check the patient’s heart rate and blood pressure both while sitting and standing to test for orthostatic hypotension. The physician may also conduct some simple balance tests in the office, order medical labs or refer patients for diagnostic imaging or other additional
testing. Patients may want to ask their primary care physician if their community has a specialized balance and vestibular clinic. Many medium and large communities do.

Though exercise is not the most appropriate treatment for some health conditions that affect balance, it can have a significant impact on those whose balance problems are related to an immunodeficiency or autoimmune disease. There are four general types of exercise people can perform at home depending on the source of the problem: strengthening exercises that target a muscle group in isolation, functional exercises, habituation activities and gaze stabilization activities.

This article focuses on only strengthening and functional exercises related to balance. It does not include habituation and gaze stabilization activities that are part of vestibular rehabilitation programs designed to improve symptoms related to inner ear disorders (e.g., vertigo/dizziness, imbalance, visual disturbances) and secondary symptoms such as nausea and vomiting. Such activities are customized to address a patient’s specific problems, and should not be prescribed without a thorough clinical exam.

The muscle groups most involved in maintaining balance include the ankle musculature, hip and knee extensors, hip and trunk flexors and hip abductors. Three sets of 10 repetitions should be performed of each exercise unless otherwise indicated. If an individual is unable to perform at least six consecutive repetitions of a strengthening exercise, it’s too difficult. If a person can easily perform 12 or more repetitions, the exercise is too easy. [See “Exercise for CIDP,” IG Living magazine, August-September 2010, for more on modifying exercises at www.igliving.com/magazine/articles/IGL_2010-08_AR_Exercise-for-CIDP.pdf]

While exercising, remember to use upper-extremity support to increase safety by holding onto a stable surface or handrail for all activities. And, breathe properly by inhaling prior to performing the exercise, and exhaling slowly through pursed lips as the exercise is performed. Take another breath between repetitions.

**Ankle Exercises**

The ankle is normally the first part of the body to react when balance is challenged. You can feel this in action by holding onto a stable surface with two hands and then standing on one leg. To maintain balance, you’ll likely feel and see your ankle teeter back and forth. If you don’t, try balancing with just one hand on the surface or without holding on at all, but only if it is safe to do so.

Standing on one leg for up to 10 seconds at a time, three times on each side, is a good way to exercise the ankle musculature. Support can be varied to make it more or less difficult. To exercise the muscles in isolation, you can write the letters of the alphabet with your foot in the air or in a tub of water. A resistive therapy band or surgical tubing can be used to add resistance. Place the looped band around the foot on one end and grasp or loop the other end to stable furniture. Move the foot/ankle opposite to the direction of pull from the band or tubing (Figure 1).

**Figure 1. Ankle Exercise with Band or Tubing**

**Hip and Knee Exercises**

When the body moves far enough outside of the base of support, ankle movement will not be sufficient to remain upright, and the body will subconsciously attempt to maintain balance using the hips. The hip and trunk flexors help to prevent backward falls, while the hip extensors, dominated by the gluteus maximus, assist with preventing forward falls.

**Hip and knee extension.** The quadriceps muscle group (made up of four muscles) is the primary extensor of the knee. The quads are also partially responsible for flexing the hip. Activity in the muscle group isn’t as prominent as in other muscles when balance is challenged, but the quads are key to preventing the knees from collapsing during stance when they are bent even slightly, whether walking or standing still. Together with the hip extensors, the quads are also very important for standing up and stepping up (e.g., climbing stairs).
The following activities are good dynamic exercises for knee and hip extensors:

- Mini-squats: Stand with the legs shoulder-width apart and slowly bend your knees until they completely eclipse the tip of your toes. If you aren’t able to see your toes, position your arms at the side of your body (bent at the elbow to a right angle with closed fists), and bend your knees until they are directly under your fists. It’s important not to bend too deeply because of the pressure that is placed on the joints, as well as the risk of going too far and not being able to get back up.

- Mini-lunges: To make the exercise a mini-lunge, take a half step forward and shift your weight over the leading leg (Figure 2). Repeat the knee bend to the point that the leading knee eclipses the toes beneath it.

- Step-ups: From a standing position below a flight of stairs, step up onto the bottom step and return. Alternate foot leads, so that both sides get a workout.

**Hip and trunk flexion.** The rectus femoris of the quadriceps, as well as a number of other muscles, help to flex (or lift) the leg. When the feet are planted on the ground, these muscles help to stabilize the hip and pelvis. If someone begins to fall backward, the trunk flexors can bend the spine forward in an attempt to regain control, but strengthening the abdominal muscles, which are responsible for the action, is not typically a focus of balance programs. Abdominal crunches (not traditional sit-ups) are the best way to exercise the abdominals if you do decide to make trunk flexion a part of your regular exercise routine.

There aren’t any great balance exercises focusing on the hip flexors that can safely be performed on one’s own. The best activity may simply be walking backward, accompanied by a companion for safety. Walking backward uses the hip extensors to walk, but the hip and trunk flexors are utilized for stabilization. Speed and step length can be varied to make the task more or less difficult.

To exercise the hip flexors in isolation, march in place or perform a straight leg raise by lying flat on your back and raising a straight leg 6 to 12 inches off of the surface (Figure 3). Maintaining a bent knee on the opposite side can help to take strain off of the back.

**Hip abduction.** The hip abductors, highlighted by the gluteus medius (which can be felt over the ball of the hip when they are actively working), are responsible for side-to-side stability. Two of the activities already presented — mini-lunges and step-ups — also exercise the hip abductors. Another great functional activity is side-stepping while holding onto the wall or a countertop for support. Once you’ve made it down the length of the hallway or countertop, reverse direction so the other leg is leading.

To exercise the hip abductors in isolation, stand at a countertop or lie on your side, slowly lift the leg out to the side and then return to center (Figure 4). When performed while standing, the
exercise actually works both sets of hip abductors, because while one set is working to move the leg, the other side is stabilizing the pelvis to prevent you from falling over to the side.

**Modifications and Other Functional Balance Activities**

All of the static (stationary) or static-dynamic (moving in place) functional activities found in this article can be cautiously modified to challenge balance further and/or to make the exercise more difficult.

It’s a common occurrence that many falls happen at night when lighting is absent or insufficient. This is particularly true for those who experience lost or diminished sensation in their lower extremities (feet and legs) associated with neuropathy. When sensation is impaired, sense of sight becomes more important to help orient a body in space. If vision is also impaired, the risk of missteps and falls increases substantially. Hence the importance of sufficient lighting, including use of night lights when getting up to go to the bathroom.

Obscuring vision can also be used as a technique to challenge and train balance further when it’s used in a controlled environment. To do so, try closing your eyes while holding onto a stable surface or using a friend or family member to serve as a “spotter” while performing one of the static or static-dynamic activities (mini-squats, mini-lunges, hip abduction or marching in place).

The demands on balance can also be enhanced (with or without eyes closed) by performing the exercises on a compliant surface like a foam pad or pillow. This forces stabilization muscles to work harder to prevent deviations from the body’s base of support.

In addition, the following are two higher-level functional balance activities for your consideration:

- **Diagonal stepping (Figure 5):** To perform this activity, place masking tape or Velcro on the floor in the form of a cross to create an area of four equally sized quadrants. Keep the left foot planted in the middle of the cross and take a step forward along the line with the right foot. Return to center with the right foot and take another step, this time to a point midway in the first quadrant. Follow this pattern in a clockwise manner until you have taken a step backward along the tape line directly behind you (i.e., halfway around the cross). Then, switch sides by planting your right foot in the center of the cross, and taking steps clockwise around the rest of the cross with your left foot.

- **Stepping over obstacles:** Set up a line of stacked books (3 to 8 inches tall and spaced 3 to 5 feet apart) down the length of a hallway so you have a wall to support yourself if you begin to lose balance. Walk slowly down the hallway, trying to alternate your leading foot, as you step over the obstacles in the pathway.

**Better Balance Can Be Achieved**

Notably, there is some investigative support to suggest a home exercise program consisting of functional activities is more effective for helping to prevent falls than one that is comprised solely of strengthening exercises that target a muscle in isolation. Considering this, if someone is able to incorporate functional activities into their exercise routine, I would recommend doing so. If there are restrictions on exercising while standing due to safety concerns or increased pain, non-weight-bearing strengthening exercises are a good alternative.

Just about everyone can develop better balance. Proper diagnosis, learning what can be done to make the home environment safer and an appropriately designed exercise program are a good start.

**MATTHEW DAVID HANSEN, DPT, MPT, BSPTS,** is a practicing physical therapist in Utah and president of an allied healthcare staffing and consulting agency named SOMA Health, LLC. He completed his formal education at the University of Utah, Salt Lake City, and has additional training in exercise and sports science, motor development and neurological and pediatric physical therapy.
Specific Antibody Deficiency and/or Impaired Polysaccharide Responsiveness

Much is being learned from research and case studies about this very common primary immunodeficiency that is only rarely treated with immune globulin therapy.

By E. Richard Stiehm, MD

SAD IS a primary immunodeficiency (PI) characterized by recurrent infections, normal immunoglobulin levels and normal antibody responses to protein antigens, but impaired antibody responses to polysaccharide antigens. These antigens are typically those present in the PPSV23 Pneumovax vaccine.

SAD is probably the most common PI. However, because of its mild symptoms and cumbersome diagnostic procedure, the illness is often overlooked, and patients are not referred to an immunology center for detailed immunologic studies. They are also not entered into PI registries.

SAD was first described in 1983 by Ambrosino, et al., under the title impaired antibody response to polysaccharides. The patient was a 30-year-old man with a history of otitis, bronchitis and several bouts of pneumococcal pneumonia. His immunoglobulins were normal except for a slightly elevated IgM, and antibody responses to tetanus and diphtheria vaccines were normal, but he had no detectable antibody to H. influenzae, meningococcal and pneumococcal polysaccharide vaccines.

The diagnosis of SAD should be reserved for patients with no other PI or secondary immunodeficiency (SI). Yet, its signature immune defect, impaired polysaccharide responsiveness (IPR), is common in many other PIs and SIs, which will be discussed later.

The cause of SAD is not established. It is maturational in infants and associated with waning immunity of the elderly. In a few cases, there may be a genetic defect of the BTK (Bruton’s tyrosine kinase) gene associated with Bruton’s agammaglobulinemia, subtle T-cell defects or a defect in generation of memory B cells.

Incidence

A normal response to each serotype of the PPSV23 vaccine was defined as a protective titer of 1.3 ug/ml by an American Academy of Allergy, Asthma and Immunology (AAAAI) working group. Immunized children should respond to at least 50 percent of the serotypes not present in the PCV13 vaccine, and
adults should respond to at least 70 percent of the serotypes not present in the PCV13 vaccine. For simplicity’s sake, the rise in titer or prevaccine titers is not used in the calculation. These arbitrary levels have been questioned, some using 1.0 ug/ml or 1.5 ug/ml as a protective titer, and at least one group using 50 percent of the titers as a normal response in adults.

A diminished response to the PPSV23 vaccine may occur in up to 10 percent of the population that is not clinically ill. Vaccine responsiveness increases with age in children and young adults, and wanes after age 60. Accordingly, children younger than 24 months and adults older than 60 should get the PCV13 vaccine for protection against pneumococcal disease.

Among children older than 2 years with recurrent respiratory infections, SAD is exceedingly common. In five studies, the pooled frequency of SAD was 221 of 825 children or 27 percent, compared to 3 percent to 10 percent of well age-matched controls with the same immune profile.

Among adults with chronic rhinosinusitis (CRS), two large studies showed 200 of 834 (24 percent) had SAD, which is in agreement with a meta-analysis of 13 studies in which 8 percent to 35 percent of adults had SAD. Among adults older than age 60, SAD may be even more frequent as surmised by their weakened responses to the PPSV23 vaccine and by one small study that showed SAD in 12 of 15 (70 percent) older adults with CRS.

The high frequency of CRS, estimated to be present in 31 million persons in the U.S., and the high frequency of SAD in these adults lends credence to the Immune Deficiency Foundation’s estimate that PI may affect one in 1,200 people.

Clinical Features

As in the highlighted vignette, children with SAD characteristically have recurrent episodes of runny nose, cough, sore throat, otitis and low-grade fever. Many have allergic rhinitis, asthma and eczema. Nasal obstruction leads to mouth breathing and nighttime snoring. Physical examination may disclose the A SAD Vignette

Five-year-old Sadie has had several episodes of otitis (middle ear infection) with low-grade fever, runny nose and cough since she started day care at 3 years old. She has had multiple courses of antibiotics, but the respiratory complaints recur after a few weeks. Growth and development is normal, and a family history is unremarkable. Childhood vaccines are up to date, including the 13-valent pneumococcal conjugated vaccine (PCV13, Prevnar).

A physical exam disclosed a pale girl with circles under her eyes. She was in the 20th percentile for height and weight. There were moderately enlarged neck lymph nodes, dull and scarred tympanic membranes (ear drums), enlarged tonsils, posterior pharyngeal cobblestoning (lymphoid nodules) and an absent cough reflex. Her chest was clear, and the rest of the exam was unremarkable.

Hemoglobin was 10.5 gm/dl, and the white blood count was 5,400 cells/ul with a normal differential. The erythrocyte (red blood cell) sedimentation rate was slightly elevated at 18 mm/hr. A throat culture showed normal flora (bacteria). A Waters’ view sinus X-ray showed opacification of the right maxillary sinus and mucosal thickening of the left maxillary sinus (a symptom of acute sinusitis). A lateral pharyngeal X-ray showed enlarged adenoids.

Immunoglobulin levels were 630 mg/dl for IgG, 85 mg/dl for IgA, 42 mg/dl for IgM and 55 IU/ml for IgE. Antibodies to tetanus, H. influenzae and nine of 23 pneumococcal serotypes were protective, all of which were in the protein-conjugated PCV13 vaccine. The 23 valent pneumococcal polysaccharide vaccine (PPSV23, Pneumovax) was given, and repeat antibody titers one month later showed protective titers (1.3 ug/ml or higher) to only three of the 10 serotypes present in the PPSV23 vaccine but absent in the PCV13 vaccine.

A diagnosis of sinusitis, chronic otitis and specific antibody deficiency (SAD) was made. She was given a three-week course of cefdinir, followed by four months of 5 mg/kg of azithromycin prophylaxis three times a week. Repeat sinus films and respiratory symptoms were improved significantly. Prophylaxis was stopped since it was summer, and she will be reevaluated before entering school.
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.

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

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

Hizentra is made from components of human blood. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent, cannot be completely eliminated.
Hizentra®, Immune Globulin Subcutaneous (Human), 20% Liquid
Initial U.S. Approval: 2010

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

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

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

DOSAGE AND ADMINISTRATION
For subcutaneous infusion only.
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) \times 1.37 \div \text{No. of weeks between IGIV doses}
• Biweekly: Start Hizentra 1 or 2 weeks after the last IGIV infusion or 1 week after the last weekly Hizentra/IGSC infusion. Administer twice the calculated weekly dose.
• Frequent dosing (2 to 7 times per week): Start Hizentra 1 week after the last IGIV or Hizentra/IGSC infusion. Divide the calculated weekly dose by the desired number of times per week.
• Adjust the dose based on clinical response and serum IgG trough levels.
Administration
• Infusion sites – 1 to 4 injection sites simultaneously, with at least 2 inches between sites.

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As tolerated

ADVERSE REACTIONS
The most common adverse reactions observed in \geq3% 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.

Based on October 2016 revision

Can IgIQ® help you?
If you answer YES to any of these questions, call 1-877-355-IGIQ (4447) Monday–Friday, 8 AM to 8 PM ET.
allergic facies (pallor, circles under the eyes, open mouth), enlarged tonsils, postnasal drip, pharyngeal cobblestoning (lymphoid nodules) and a diminished gag reflex. Cervical lymphadenopathy may be present. Pectus excavatum (a congenital disorder that causes the chest to have a sunken appearance), rales (rattling noises), wheezing or a Harrison’s groove (a dip or crevice in the chest where the ribcage meets the diaphragm) suggest chronic asthma or other chronic lung disease.

Adults with SAD typically have CRS, as characterized by at least 12 weeks of symptoms, including purulent nasal discharge, nasal congestion, headache, anosmia (loss of smell) and fever, with objective findings of sinusitis by nasal endoscopy or a CAT scan. There is a high frequency of tobacco use, asthma and other chronic lung disease. Physical examination may reveal turbinate (inverted cone) swelling, nasal discharge, polyps, postnasal drip, wheezing, rales, increased anti-posterior diameter and digital clubbing, the latter suggesting chronic lung disease.

Immune system evaluation includes IgG, IgM, IgA, IgE and sometimes IgG subclass levels, all characteristically normal in SAD. Antibody tests to previously administered protein vaccines (tetanus, H. influenzae and PCV13 vaccines) are also normal. Other tests for immune deficiencies, including B and T cell subsets, lymphoproliferative studies to antigens and mitogens, rhodamine dye study for chronic granulomatous disease, complement activity and component assays, and mannose binding lectin, are used to identify other PIs.

The hallmark of SAD is the presence of impaired polysaccharide responsiveness. This requires the administration of the PPSV23 vaccine, which contains 10 serotypes not present in the PCV13 vaccine. After four to six weeks, a blood sample is taken and the antibody levels to 23 pneumococcal serotypes are obtained. These are available in many reference laboratories using ELISA (enzyme-linked immunosorbent assay) or multiplex bead assays. A protective level of pneumococcal antibody is 1.3 ug/ml. The number of serotypes unique to the PPSV23 vaccine that are protective establishes a diagnosis. Normal children should develop a protective response to 50 percent of these serotypes, and normal adults should develop a protective response to 70 percent of them. (If a prevaccine antibody test was performed, the rise in titer to individual serotypes or the prevaccine titers are not used to estimate the response. Thus, a titer that increases from 0.4 to 1.0 ug/ml is considered nonprotective, a titer of 1.6 falling to 1.4 ug/ml is protective, and a titer that falls from 1.4 to 0.8 ug/ml is nonprotective).

The response to the PPSV23 pneumococcal vaccine is also used to determine the severity of SAD. An AAAAI working group has designated these as mild, moderate, severe and memory phenotypes as summarized in Table 1. The mild phenotype

<table>
<thead>
<tr>
<th>Phenotype*</th>
<th>PPV23 response, age &gt;6 y</th>
<th>PPV23 response, age &lt;6 y</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe</td>
<td>≤2 protective titers (≥1.3 μg/mL)</td>
<td>≤2 protective titers (≥1.3 μg/mL)</td>
<td>Protective titers present are low</td>
</tr>
<tr>
<td>Moderate</td>
<td>&lt;70% of serotypes are protective (≥1.3 μg/mL)</td>
<td>&lt;50% of serotypes are protective (≥1.3 μg/mL)</td>
<td>Protective titers present to ≥3 serotypes</td>
</tr>
<tr>
<td>Mild</td>
<td>Failure to generate protective titers to multiple serotypes or failure of a 2-fold increase in 70% of serotypes</td>
<td>Failure to generate protective titers to multiple serotypes or failure of a 2-fold increase in 50% of serotypes</td>
<td>2-Fold increases assume a prevaccination titer of less than cutoff values in Summary Statement 26</td>
</tr>
<tr>
<td>Memory</td>
<td>Loss of response within 6 mo</td>
<td>Loss of response within 6 mo</td>
<td>Adequate initial response to ≥50% of serotypes in children &lt;6 y of age and ≥70% in those &gt;6 y of age</td>
</tr>
</tbody>
</table>

includes patients who respond to some but less than 50 percent of the serotypes, the moderate phenotype responds to two to three serotypes, and the severe phenotype responds to none or one serotype. The memory phenotype includes patients who initially have a protective phenotype but revert to a nonprotective phenotype after six months. Many of these latter patients continue with recurrent infections. Repeating the PPSV23 vaccine immediately after a poor response is not useful or recommended since polysaccharide vaccines do not elicit a T-cell memory cell response.

Other polysaccharide vaccines are available for use in identifying SAD. These include the meningococcal (Menactra) and Typhoid Vi polysaccharide vaccines. The latter vaccine’s advantage is that nearly all patients have no antibodies to it nor are these antibodies present in immune globulin (IG) preparations. Thus, it is of special value in identifying SAD in a patient while on IG replacement therapy.

Management

The first step in managing SAD is to treat the infectious illnesses that brought the patient to the doctor. This screening includes blood tests, cultures, imaging studies and antimicrobial therapy. Then, the immune evaluation outlined above is initiated, including a blood test for antibodies to protein antigens and 23 pneumococcal serotypes. Then, the PPSV23 vaccine is given, and its response is rechecked in four to six weeks.

At that time, a diagnosis of SAD is made, and the plan for continued therapy is established. If chronic infection persists, antibiotics must be continued or changed. Long-term prophylactic antibiotics may be necessary. Other measures may include environmental control of allergies, inhaled steroids for asthma, surgical removal of tonsils and adenoids, and sinus washes.

If the antibody test shows waning immunity to PCV13 serotypes, this vaccine may be repeated. As noted above, an immediate booster of the PPSV23 is not recommended or useful since polysaccharide vaccines do not provide T-cell memory responses.

Depending on the infectious severity, a prolonged course of antibiotics may be necessary. Other children with less severe infections may do well on prophylactic antibiotics given three times a week. Some patients may require tonsillectomy and adenoidectomy. Others may require inhaled steroids, bronchodilators, antihistamines and sinus washes. Repeat titers and imaging are performed after several months if the patient does not do well.

IG therapy is reserved for patients with severe and refractory infections and persistent nonprotective antibody levels.

IPR in Other Conditions

IPR is present at the extremes of life. In infants younger than 2 years of age, this defect led to the development of protein conjugated vaccines for D. pneumoniae and H. influenzae, both serious bacterial infections in infants. IPR may persist beyond age 2 as part of transient hypogammaglobulinemia of infancy.

IPR is also common in the elderly, rendering the recommended PPSV23 vaccines unreliably protective and, thus, the PCV13 vaccine is now also recommended.

IPR has been identified in many other conditions (Table 2 and Table 3). All PIs with global antibody defects, including
agammaglobulinemias, common variable immunodeficiency and severe combined immunodeficiencies, will have IPR. Less severe antibody deficiencies (e.g., selective IgA deficiency, selective IgM deficiency, IgG subclass deficiencies and transient hypogammaglobulinemia of infancy) have a high incidence of chronic respiratory infections and IPR. Several less-severe combined immunodeficiencies (e.g., Wiskott-Aldrich syndrome, ataxia-telangiectasia, DiGeorge syndrome and the hyper-IgM syndromes) often have IPR and frequent respiratory infections.

Many other SIs will have IPR and resultant susceptibility to respiratory infections despite normal or near normal IgG levels. IPR may be associated with several chronic illnesses, medications, trauma, malnutrition and surgery. If an immune workup is deemed necessary, assays for polysaccharide responsiveness should be included. Such information is of use in determining whether vaccines should be given, whether postexposure prophylaxis is needed, whether antibiotics should be used and whether IG therapy is indicated.

The take-home message is that if an immune deficiency is suspected, an IPR must always be considered.

**Summary**

SAD is a PI characterized by normal immunoglobulin levels and normal antibody responses to protein antigens, but impaired antibody responses to polysaccharide antigens in the absence of any other PIs or SIs. It is probably the most common PI, occurring in 15 percent of children with recurrent respiratory infections and 25 percent of adults with CRS. Diagnosis is established by a

**Table 3. Secondary Immunodeficiencies at Risk for Impaired Polysaccharide Responsiveness**

<table>
<thead>
<tr>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infants under 24 months</td>
</tr>
<tr>
<td>Adults over age 60</td>
</tr>
<tr>
<td>Major trauma or surgery</td>
</tr>
<tr>
<td>Splenectomy and asplenia</td>
</tr>
<tr>
<td>Severe burns</td>
</tr>
<tr>
<td>Postorgan transplant with immunosuppression</td>
</tr>
<tr>
<td>Uremia and renal dialysis</td>
</tr>
<tr>
<td>Diabetes</td>
</tr>
<tr>
<td>Cirrhosis</td>
</tr>
<tr>
<td>Malnutrition</td>
</tr>
<tr>
<td>Radiation</td>
</tr>
<tr>
<td>Lymphoma and leukemia</td>
</tr>
<tr>
<td>High-dose corticosteroids</td>
</tr>
<tr>
<td>Immunosuppressive drugs</td>
</tr>
<tr>
<td>Monoclonal antibodies (rituximab, etc.)</td>
</tr>
<tr>
<td>Protein-losing illnesses (nephrosis, intestinal lymphangiectasia, etc.)</td>
</tr>
</tbody>
</table>

**References**

1. Orange, JS, Ballow, M, Stiehm, ER, et al. Use and Interpretation of Diagnostic Vaccination in Primary Immunodeficiency: A Working Group Report of the Basic and Clinical Immunology Interest Section of the American Academy of Allergy, Asthma and Immunology. *Journal of Allergy and Clinical Immunology* 2012; 130:S1-S24.


Understanding NK Cell Deficiency

Even as more is discovered about the genetics of this group of rare PIs, and the number of NKD diagnoses on the rise, there is still much to learn about how to treat it.

By Jordan S. Orange, MD, PhD

NK CELL DEFICIENCY (NKD) involves a group of primary immunodeficiency diseases (PIs) in which one specific part of the immune system, the natural killer (NK) cells, is defective. Patients with NKD are susceptible to viral infections, most notably infections with two types: papillomaviruses that cause warts and herpes viruses. Common herpes viruses causing problems in patients with NKD are the varicella zoster virus that causes chickenpox, cytomegalovirus (CMV), Epstein Barr virus and herpes simplex virus (HSV). Patients with NKD typically have more of these infections and unusual consequences from them.

Fifty of the more than 350 PIs have some defect in NK cells. For example, in certain types of severe combined immunodeficiency (SCID), NK cells fail to develop. However, it is the absence of T cells rather than NK cells in SCID that represent the greatest challenge to these patients. Whereas in NKD patients, the NK cells are the main defect of immunity. And, while patients and causes of NKD are increasingly being recognized, it is believed NKD is quite rare.

What Are NK Cells?

NK cells are a type of lymphocyte that specializes in the destruction of diseased cells in the body by directly killing them — a process called cytotoxicity (Figure 1). They are especially effective in killing cells that have become infected with a virus or have lost growth control and are in danger of forming a cancer.

NK cells are one of two types of lymphocytes that mediate cytotoxicity; the other is the cytotoxic T cells (CTLs). The main difference between NK cells and CTLs is NK cells are part of the innate immune system, whereas CTLs are part of the adaptive immune system. This means NK cells are ready to function without additional training, but CTLs require education to become active and effective. Thus, NK cells are part of the early defense against viral infection and cells that have lost growth control, while CTLs are called into action to provide more durable defense.

As a result of this duality in cytotoxicity, lacking NK cells leaves a very specific hole in the immune defense, creating
susceptibility to a small number of viruses. The reason for these particular infectious susceptibilities is that the viruses causing difficulty in NKD patients utilize very specific mechanisms to evade and escape CTL responses. Thus, NK cells are somewhat indispensable in these infections.

NK cells also serve other functions besides cytotoxicity, including producing inflammation to help organize other immune defenses and to help control immunity to prevent immune responses from getting out of control. NK cells are found circulating in the blood and also reside in many of the body’s organs. And, while people can survive for periods of time without them, they become susceptible to life-threatening viral infections.

Interestingly, NK cells are currently being investigated as a treatment for certain types of cancer, where they are grown in the laboratory and infused back into patients. While experimental at this point, they hold therapeutic promise for the future.

**Genes Causing NKD**

Since NK cells were first discovered in the 1970s, a number of cases of NKD have been reported in the medical literature. Several notable descriptions of patients with NKD were made in the 1980s, some of which have subsequently been connected with a genetic explanation. The first gene associated with an NKD was found in the 1990s. Today, there are seven gene defects known to cause NKD, and there will undoubtedly be others reported in coming years as this particular diagnostic category is expected grow.

Genes that are known to cause NKD at present are (in alphabetical order) FCGR3A, GATA2, IRF8, MCM4, MCM10, GINS1 and RTEL1. Some of these genes, when defective, can also cause other conditions, but either particular variants in these genes or particular presentations of the defects can result in NKD. Although the specific details of each of these genes, their impact upon NK cells and how they cause NKD is beyond the scope of this overview, other articles can be accessed for additional information. The clinical sequence analysis of many of these genes can be found on currently available PI diagnostic gene panels, and can also be obtained through whole exome sequencing.

**Diagnosing NKD**

Diagnosing NKD requires patients have a deficiency of NK cell number and function, or just function, as well as a clinical history suggesting NKD. Because there are many reasons someone can have slightly low NK cell numbers and function, it is important for laboratory test results to be repeated, significant and in context of a clinical history suggestive of NKD (i.e., with herpes viral or papillomavirus infections).

NK cell number is assessed by flow cytometry of the lymphocytes in peripheral blood. NK cell function is determined by a cytotoxicity assay that measures the ability of NK cells to kill a tumor cell in a culture dish in the laboratory. People with low NK cell numbers can have a defect in their NK cells’ ability to normally develop, which can be a clue to NKD. And, normal development of NK cells must be assessed in context of NK cell function to ensure they are actually abnormal.

Importantly, the normal range for low NK cell numbers from commercial laboratories is one in 20 people (5th percentile). This does not mean having a low number of NK cells equates to an NKD. For some people, having a slightly low number is just their own personal set point (we get especially concerned, however, when someone has less than 1 percent). Therefore, it is again...
important to be sure these low numbers of NK cells are indeed also dysfunctional to determine whether the patient has an NKD. To ensure valid results, tests must be performed by a reputable laboratory and during a time when someone is generally well, because illness can suppress the test result. And, it is essential these abnormalities in the test results be found repeatedly, with a recommendation that there be consistent abnormalities on three separate occasions scheduled at one-month intervals.

Looking to the future, as we gain a greater understanding of the genes underlying NKD, we should be able to increasingly rely upon genetic results for diagnosis.

What Is Not NKD

Unfortunately, the diagnosis of NKD is frequently misapplied. NK cell numbers are low in many people as defined by normal threshold ranges found in laboratories. The percentages and function of NK cells are affected by stress, depression and illness, and can be low because the body is in the midst of fighting a challenge. Thus, individual values in tests do not define NKD. People with NKD have abnormal values repeatedly and in the context of a convincing clinical history. Borderline low results are also a problem. Many patients with NKD have very clear absent or near-absent test values.

For clinical history, there are certain things typically not found in association with NKD. One example is patients who get sick frequently from cold (and related) viruses. While there are likely many reasons for that, we have not found NKD as an explanation. Another is patients who get frequent cold sores or focused outbreaks of HSV. While there are likely reasons people get frequent cold sores, NKD is typically not one of them. Patients with NKD do have problems with HSV, but these typically occur in many places on the body (not just on the lips) or when it is especially severe. Finally, postherpetic neuralgia is a real problem that occurs when there is excess pain after an HSV outbreak, but this is typically not a main feature of patients with NKD.

Treating NKD

Presently, there is precious little known about treatment for NKD. It is hoped more will be learned as further experience is gained and more patients are diagnosed.

There has never been an interventional trial or drug study in patients with NKD, so we really cannot say that particular treatments are proven to work. However, we do know certain medications are effective against the viruses that cause problems in NKD patients, and we prescribe those to help. In particular, continual prophylaxis with the synthetic nucleoside analogs such as Acyclovir can help fill in some of the void left by defective NK cells. Some patients have benefited from immune stimulatory treatments to try to boost NK cell function (like interleukin-2 or interferon), or prophylaxis with intravenous immune globulin to try to provide additional defense against some of the susceptible viruses. While bone marrow transplantation is not a treatment the author has experience with, there have been reports in the literature of its success for NKD in patients with particularly severe consequences.

Virus-specific T cells are one treatment of interest for the future. With this, CTLs directed against a particular virus can be infused into a patient to help control a viral infection. Although only experimental at this point, it could represent a new path for PI and NKD patients who have defective immunity and challenges fighting viruses.

A Hopeful Future

NKD is a rare but emerging PI in which an abnormality in NK cells is the main immune defect. Patients have susceptibility to herpes viruses and papillomaviruses, and anecdotal experience has provided some hope for treatment. Understanding NKD from an immunologic, clinical and scientific standpoint is leading to advances and new identifiable causes. We are optimistic that future and ongoing research will bring new answers and, hopefully, new treatments for patients suffering from deficiencies in NK cell defenses.

JORDAN S. ORANGE, MD, PhD, is chief of the Section of Immunology, Allergy and Rheumatology at Texas Children’s Hospital, and professor of pediatrics-rheumatology at Baylor College of Medicine in Houston, Texas.

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3. Orange, JS. Natural Killer Cell Deficiency Syndrome: Clinical Manifestations and Diagnosis. In: Rose, BD (Ed.), UpToDate, Wellesley, MA.
“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
Profile: 
SamMichael Long

By Trudie Mitschang

TO SEE SamMichael Long today, you would never guess this active and ambitious college student spent much of his childhood in and out of hospitals. Diagnosed with common variable immune deficiency (CVID) at a young age, SamMichael has resisted the temptation to let his illness define him. Drawing on his relentless optimism to excel in life, in spite of chronic illness, SamMichael serves as a mentor and volunteer for the Immune Deficiency Foundation (IDF) Teen and Young Adult Council. When he is not focused on giving back and helping other teens navigate the often-difficult road of primary immunodeficiency (PI), this Minnesota native is busy pursuing a computer science degree at Saint Mary’s University of Minnesota.

SamMichael Long has thrived since being diagnosed with CVID in 2005 and is now pursuing a computer science degree and working with the Immune Deficiency Foundation Teen Council.

Trudie: When were you diagnosed with CVID?
SamMichael: I was in and out of the hospital for the first seven years of my life. I was diagnosed with CVID in 2005, and during the first year, I was treated with Xolair (omalizumab). After that, I was switched to intravenous immune globulin (IVIG) therapy for six years, and then was switched again to subcutaneous IG (SCIG) therapy. I was treated with SCIG for four years, but I discontinued IG therapy almost four years ago.

Trudie: What is your current treatment plan?
SamMichael: I am taking an antibiotic supplement called azithromycin in place of IG therapy, and I have never felt better. I go in once a year for labs to check my immunoglobulin levels, and I am currently doing great.

Trudie: How has living with chronic illness impacted your life?
SamMichael: Living with a PI has helped me develop into the person I am today. One of the best pieces of advice I have ever received was when I first learned about IDF from a wonderful lady named Kathy Antilla. She told me: “You cannot let your disease control you; you have to control it.” I live by that motto every day. I don’t let my PI stop me from doing anything I want to do. Just because my body doesn’t work the same as everyone else’s doesn’t mean I can’t do what they are doing. Granted, I have to be more careful about certain things, but my PI has never held me back. I played sports all through high school, I go hiking, camping and swimming, and I live a pretty normal life. I cannot thank IDF for all it has done for me and my family over the years.

Trudie: What is your role on the IDF Teen Council, and how did you become involved?
SamMichael: My role with the IDF Teen Council is to be a mentor and an advocate. I have been blessed to travel around the country and speak to further educate people on PIs and what IDF does. I started becoming really involved after I attended my first IDF Teen Escape event in 2011, which helped me figure out that working with IDF is something I wanted to pursue. First, I started volunteering at local...
family conferences. When I turned 16, I was put in touch with Dan Antilla, the head of the Teen Council, and that’s when things really took off. That same year, I was sent to California to attend a Teen Escape, but as a leader. That was a surreal moment for me because it was like everything had come full circle. Since then, I have attended numerous Teen Escapes and Family Conference Days, as well as spoken on Capitol Hill for IDF Advocacy Day in April 2016.

Trudie: What does the Teen Council do?
SamMichael: We work in conjunction with IDF staff to advocate for youth living with PI to let them know they are not alone, and to help educate them on their specific disease. All of the members of the Teen Council come from different walks of life, and each of us has our own unique story. We serve as mentors for those who want to be educated, but mostly we serve as a family. We welcome each kid we meet and interact with into our little family. Because the PI community is so small, knowing there are people out there just like you means the world. We are just like a family, and we are always here for each other.

Trudie: What are some of the unique challenges teens face living with PI?
SamMichael: The challenges vary depending on the exact diagnosis, as well as its severity. Speaking on my own behalf, I know it was hard for me growing up to keep a high energy level. I would go in for tri-weekly infusions, and by the end of that third week, my energy level would be depleted. Once I received treatment, my energy level would return. When I switched to SCIG, my energy level was more consistent because I was receiving treatments twice a week.

Trudie: How do you maintain a positive attitude?
SamMichael: Family and friends really keep me going. They are a constant motivator for me to keep striving for my goals. I also use music to maintain a positive attitude. Music is an escape for me; I find it to be the best tool to use. I just strap on my headphones and listen. I met one of my closest friends, Keegan, at an IDF event, and ever since that day, he has been like my brother. We always motivate each other and call each other when something is up.

Trudie: What role does music play in your life?
SamMichael: I have been playing music since I was little. I started piano when I was in second grade, and I am mostly self-taught. In middle school, I picked up percussion and have been doing that ever since. When I started college, I really wanted to learn something new, so I taught myself how to play the ukulele. By the end of the semester, I was playing at gigs around campus and for variety shows! Then, just this past summer, I decided to teach myself guitar, and started playing around campus and at church mass. Music has really embodied my whole life. One of my favorite quotes of all time goes like this, “Music speaks what cannot be expressed, soothes the mind and gives it rest; heals the heart and makes it whole, flows from heaven to the soul.” Music has really helped me to express myself over the years, and really helped me to find a center in times of trouble and hardship. It is something that I use as a tool to connect with people, and help them cope in situations. I also use it as a tool to praise Jesus and spread his message. Music has literally become my whole life.

Trudie: What lessons have you learned that you can pass on?
SamMichael: Be who you want to be, and don’t let anyone tell you no. Even with your PI, you can do anything that you set your mind to. There are no limits; the world is your playground. Do not let your disease control you; you have to control it. Once you do that, nothing can stand in your way. If I have learned anything over the years, it is to always ask questions about things you don’t understand, and treat every day like a fresh start. Live in the present. There is no point in living in the past because it already happened, you can’t change it and the future is unknown. But you have full control over the present, so seize it and make it your own.

TRUDIE MITSCHANG is a contributing writer for IG Living magazine.
ABOUT EIGHT WEEKS ago, my body descended into a fiery hell, the depths of which I’d never known. My joints were ablaze. Infections, like wildfires over dry ground, spread throughout my body. The lymph node under my right arm swelled to the size of a golf ball. For the first time, I couldn’t lift my arms to dress myself because of my rheumatoid arthritis (RA). I iced my joints, and then I put heat on them, yet nothing worked. My right knee filled with fluid and grew to twice its normal size. My stomach revolted, and I was quickly losing weight. Almost as troubling as the symptoms was that I couldn’t identify a trigger. What was my body so angry about? What had I done?

Just as I was ready to wave the white flag in surrender to my body, I noticed an unusual pain in my big toe. It throbbed when I tried to sleep. Underneath the edges of my pretty pink gel nail polish, I could see dark shades of purple and black. Naturally, I assumed my toe turning colors was a bad sign. So, the next day, my dear friend was kind enough to remove the polish on the offending toe for me. What we discovered beneath will haunt my dreams for the rest of my days. I sat in stunned silence for a few moments before repeating: "Wow. Wow. Wow. This is really bad." The dermatologist I saw two days later agreed. She said I had bacterial and fungus infections, and I would need to have my entire nail removed.

There was something else I learned that day. My body, in revolt for so many weeks, had been desperately trying to fight this infection and had, instead, been fighting me. Once we removed the nail and got the infection under control, I was told I would most likely succeed in getting the RA flares under control. The wildfires, it seemed, could be squelched.

Having my nail removed was a bit more horrendous than I expected, but almost immediately, the inflammation in my body went down. By the next day, I was able to dress myself. Isn’t it interesting how we oftentimes must do things we don’t want to do so we can do things we do want to do?

Caring for my toe post-avulsion was no walk in the park either. In fact, I hated it. I hated looking at it. I hated caring for it. I hated that such a little thing had caused such an enormous impact on my body. One morning, sitting on the side of the bathtub crying because my toe had gotten wet when it wasn’t supposed to, and I now had to clean it again, I realized my swollen, messy and wounded toe looked a lot like my heart was feeling. I didn’t want to look at my heart. I didn’t want to care for it. The truth is, my heart was messy and wounded. I paused for a moment. Why did I feel so wounded and angry?

Beneath the anger and the hurt was another familiar feeling: injustice. I felt I didn’t deserve this suffering. Expressing my pain and this sense of unfairness had seemed pointless. Yet, refusing to address the wounds of my heart was about as successful as ignoring the pain beneath my pretty pink gel polish.

Sometimes we hide great pain behind shiny strong exteriors because we believe that’s what is expected of us. Or, perhaps, we stuff our feelings deep inside because we believe our pain and our story don’t matter. But, let me remind you that hidden things left unresolved have a tremendous impact on our health, both physically and emotionally.

Let’s commit to caring for the wounds — both seen and unseen.
I Wish I’d Known What PI Could Do to My Teeth

By Ilana Jacqueline

ABOUT A MONTH ago, I made myself the most fabulous looking Greek gyro. I was so excited to dig into this soft-baked naan I’d bought, but when I took the first bite, something just felt wrong. I cut my lip. How did I manage to cut my lip on soft-baked naan? Then, I looked in the mirror, and to my immense horror, I saw my own personal nightmare staring back at me.

Now let me stop here and preface this by saying my body has been through some incredible things. I’ve had more procedures and surgeries than I can count. I’ve been cut open and rearranged and had bones shaved down and organs pulled apart. I’ve had surgeries without anesthesia. I’ve had stitches pulled out of my eyeballs. But, dental work is my nightmare.

One of my front teeth had split right up the middle, and had cut my lip. I looked like a hillbilly. It was the fourth front tooth that had broken this year, and I now had a row of completely different-sized front teeth. There was no way of hiding it or telling myself it could be shaved down or matched to the worst one of the set. No, this was the nail in the coffin. These teeth were now going to have to be replaced.

I stood in front of the mirror for a half hour thinking: “No, this isn’t happening. This isn’t happening.” Then, I called my mom, tossed my gyro and made several dental appointments. After seeing two specialists, I was told my teeth were much worse off than I could have imagined. I needed six teeth to be removed right up to the root and covered with caps. If I didn’t get this done, and soon, my six front teeth would continue splitting, cracking and potentially even falling out on their own.

During the early months of 2017, I’d had a major infection that required treatment with intravenous antibiotics for several months. I’m sure there was some mention of “this may cause cavities or enamel weakening” by an immunologist at some point. But I was a little more worried about the infection putting a complete and total stop to my daily life. I didn’t realize how at risk I was as a primary immune deficiency (PI) patient. I didn’t really think the years of antibiotics and medications taken for my disease would actually cause dental damage.

Even so, I was diligent about my dental care. I’d had three cleanings last year. Could my dentist actually miss a sign that my teeth were in serious crisis? As it turns out, he could, and boy did he. I was told we could have caught the damage even earlier if my previous dentist had been aware of my condition, knew how much medication I had been on and had taken regular X-rays — more regular than just once a year.

I was fortunate to be able to save the roots of my teeth, so I did not need full implants. This saved me an astronomical amount of money, pain and potential complications. However, for most PI patients, the destruction to their teeth and gums doesn’t get attention until it becomes an unsalvageable, bankrupting mess.

Consider this your cautionary tale. Pick up the phone, and make an appointment to get an extra cleaning and X-rays this month. Even if your teeth aren’t falling out of your mouth, the damage can happen quickly, especially after bad flares that require high doses of medication. You can prevent and prepare for dental disaster by:

• Alerting your dentist to your condition;
• Getting frequent exams, X-rays and cleanings;
• Looking into financing and credit options for high-cost procedures such as veneers, caps and implants that you may need in the future; and
• Asking your dentist for specialty and prescription mouthwashes and toothpaste to help prevent decay.

It’s more common than you think for PI patients to have major dental work, including dentures. There are many dental-based charities and financing options for extreme medical work due to disease progression. Preventive care is your best option for keeping your teeth healthy and strong. Invest in cleanings, good brushes and high-end toothpastes and mouthwashes. And, remember, you are not alone.

ILANA JACQUELINE is a 28-year-old dysautonomia and primary immune deficiency disease patient from South Florida. She’s been writing professionally since 2004 on everything from health and wellness to celebrities and beauty. Her blog www.letsfeelbetter.com is both a personal collection of anecdotes about life with chronic illness, as well as a resource for patients of all ages.
PERHAPS YOU’VE SEEN them in hospital rooms, guitar in hand, leading a family in a familiar song while gathered around the bedside of a sick loved one. Or maybe parading around a room in the pediatric oncology ward, banging on a tambourine with a young cancer patient. These music therapists are not just merry minstrels strolling up and down hospital halls, tooting on flutes and strumming on lyres. They’re highly trained, board-certified professionals who hold a bachelor’s degree or higher in music therapy from one of more than 70 American Music Therapy Association (AMTA)-approved colleges and universities.1

The field of music therapy is growing and gaining more recognition among members of the conventional medical community. With quality of life and patient choice now key issues in the national healthcare agenda, music therapy is being increasingly acknowledged for its unique contribution.2 The medical applications of music therapy are far-reaching and can be very beneficial to chronically ill children alongside their existing treatment regimen, both in and out of the hospital setting.

What Is Music Therapy?
Music therapy is the practice of using music to aid in healing. In a hospital setting, professionally trained music therapists work in conjunction with doctors to develop and deliver individualized music experiences to assess, treat and evaluate patients.2 Treatment plans are tailored specifically to fit each patient’s medical diagnosis, course of treatment and discharge timeline.

Music therapy unofficially began in the 20th century, after musicians were asked to play for World War I and World War II veterans at U.S. hospitals. According to AMTA, there are approximately 5,000 board-certified music therapists in the U.S. today,2 with the association’s numbers growing particularly during the last decade.

Studies show music therapy can be an effective and valid treatment option for patients with a variety of diagnoses and conditions, including respiration difficulties, chronic pain, headaches, cardiac conditions, diabetes and many others. Research also suggests that patients who have resisted more traditional treatment approaches are accepting of music therapy, and benefit from it.

With a carefully planned and strategic combination of music and music activities, therapists can help patients with anxiety, stress reduction, nondrug pain and discomfort management, which can lead to positive changes in mood and emotional state, active patient participation in treatment and decreased length of hospital stays. Along with relaxation and reduced stress and anxiety come measurable physiological changes, including improved respiration, lower blood pressure, reduced heart rate, relaxed muscle tension and improved cardiac function.

In addition to benefiting patients, music therapy can also provide positive experiences for family members by fostering emotional intimacy, relaxation for the entire family and meaningful time spent together in a positive, creative way.2

Why It Works
Although some may question the validity of using music in the world of medicine, there is science behind it. Research in music therapy supports the idea that music has healing properties. In a 2013 study led by Daniel Levitin, a prominent psychologist who studies the neuroscience of music at Montreal’s McGill University, a variety of evidence points to music’s antianxiety properties. Another study revealed music’s association with higher levels of immunoglobulin A.3 Music’s healing properties are a result of the brain’s reward center responding to it by releasing dopamine, a chemical associated with pleasure — much like the effect that comes from eating certain foods.3 Of course, a more simple theory is that music heals because almost everyone relates to music; it’s a familiar tool that is nonthreatening. When used with chronically ill children, who often have to endure painful and scary procedures, the calming effects of music can combat the negative side effects of fear and emotional distress.

Because music therapists often work nonverbally, this approach is particularly effective for patients with difficulties expressing themselves verbally, such as children with autism.3
Music therapy has also proven beneficial in cancer treatment. Walter Quan, Jr., MD, a hematologist-oncologist at St. Luke’s Medical Center in Cleveland, Ohio, says, “The mind/body relationship is particularly important in terms of looking at the immune system to treat cancer. We believe that patients who are under less stress, who are in a brighter mood, appear to do better in terms of their anticancer therapy. A study done just relatively recently on cancer patients showed that approximately three-quarters of cancer patients [who] had their usual pain medicines but also had the additional music therapy experienced less pain than previously…. Music therapy in helping patients relax could possibly be beneficial in raising the innate immune system, which could have therapeutic implications for cancer.”

Music Therapy for Chronically Ill Children

According to Elizabeth Fawcett, MT-BC, a music therapist at North Carolina Children’s Hospital, music therapy can help chronically ill children cope with their diseases. At her hospital, Fawcett spends 20 hours a week playing the guitar and piano for her pediatric patients, while also helping them write their own songs. According to her, music therapy provides a time for her patients to express their feelings in a safe way.

Unlike the doctors and nurses these children see daily, music therapists aren’t there to draw blood or deliver good or bad news. They are simply there to play or create music. But, music therapists like Fawcett play a larger role than merely providing a good time for their patients. They work alongside each child’s medical team to assess the patient’s needs and abilities, and to set attainable treatment goals that will speed up the healing process and help him or her return home more quickly.

In children’s hospitals across the country, where music therapy is practiced, music rooms are being added. These rooms contain a variety of games like Wii and PlayStation, and musical instruments like drums, keyboards and electric guitars. Fawcett’s music room even comes equipped with a digital recording studio, where patients can sing karaoke and learn to play instruments.

Fawcett says that, in some cases, music therapy involves what she calls “well-intentioned trickery.” For example, she encourages children with cystic fibrosis, a genetic disease that can cause lung infections due to thick secretions, to sing. When the karaoke machine is playing and the microphone is in their hands, these children think they are only singing, but they are actually working their lungs — and having fun doing it.

Music Therapy at Home

When a child comes home from the hospital, where music therapy aided in his or her healing, the benefits don’t have to end. Music therapists can be seen in outpatient settings, and children can practice the techniques they learned even while at home. Some simple practices that may aid in healing or maintaining good health include listening to calming music and breathing deeply. Parents may need to experiment with several styles of music to find which type best helps their child relax. “Slow music with 60 or fewer beats per minute may help you reduce anxiety,” says Fawcett. Parents can try playing music during home infusion time to help relax a young child who is typically resistant to the idea of being poked by a needle. Also, having a child sing a song in his or her head can be a means of relaxation when in a public setting, such as an infusion center or during laboratory procedures.

Music therapy may not be beneficial to everyone, and it certainly should not be used in place of mainstream medicine. But when used to complement an existing treatment, it provides another weapon for parents of chronically ill children to keep in their medical arsenal. Anything that can aid in the healing process and reduce the stress and anxiety of living with chronic illness is something worth considering.

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
Mobility Management
By Trudie Mitschang

Walking canes are designed to help people with mobility issues improve balance and walk safely. One innovation in the pipeline is the new Dring Smartcane (dring.io). The device debuted in France last year and features sensors that detect any unusual situation (fall detection, diminished activity, etc.). If that happens, the cane automatically alerts caregivers and family, without any action required by the user.

For a more low-tech solution, there are canes designed to meet very specific needs. Some offer adjustable features and can be customized based on height and left- or right-handedness. Others fold compactly for travel and come equipped with built-in LED lights. Many canes come in a rainbow of color options and with ergonomically-designed grips for added comfort.

A transport wheelchair is a mobility chair designed for convenience, short-distance use and easy handling by a caregiver. Some come equipped with rugged wheels for outdoor use, while others are specially designed for use in the bath or shower. Lightweight and foldable, these wheelchairs are easily moved and typically fit in the trunk of most vehicles. There is even a model designed to fold into a bag to be carried over the shoulder.

Travel scooters not only provide powered mobility, but also transportability. Many are compact, lightweight and can easily be disassembled into separate components to load into a vehicle. Four-wheeled models allow for enhanced stability on rough terrain, and an ergonomic delta tiller can provide increased usability for those with limited hand strength. For all models, consideration must be given to how long the scooter can operate on a single battery charge.

For those who have difficulty traversing household stairs, a stairlift could offer the ideal solution. The most recognizable type is the straight stairlift that attaches to the stair treads by way of a rail on which a chair can glide up and down. These lifts are suitable for those who can walk but not climb, and who don’t need to transport anything up and down except themselves and small items that can be held on a lap. A variation would be a curved model that can be utilized with a winding staircase.

Those with a chronic illness do not need to let mobility issues prevent them from doing the things they love. Freedom in movement is possible with the right device. For help making the best device choice, consult with a physician or physical therapist who can offer recommendations.

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

The HurryCane all-terrain cane features a freestanding design and pivoting base for enhanced stability. The device adjusts to fit height and folds to store easily in a purse or beneath a chair. It supports up to 350 pounds. $39.96; hurrcane.com

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**Style in Motion**

The Rascal 8 Mobility Scooter from Windermere Motion is an easy-to-navigate model that has a 275-pound weight capacity and easily disassembles into six lightweight pieces. It features black, non-scuffing tires and a maximum speed of up to 4 mph. $599; walgreens.com

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**Shopping Guide to Mobility Products**

**Confident Climbing**

The Easy Climber Stair Lift offers a custom-designed fit for either straight or curved and long or short staircases. The swivel design comes with foot and armrests to make getting in and out safe and easy. Custom pricing; easyclimber.com

**Easy Transport**

The Drive Aluminum Transport Chair folds down for easy storage and weighs just 19 pounds. It includes a seat belt for added safety, height adjustments and swing-away footrests. $115; justwalkers.com

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**Improve Your App-titude**

Phones can be updated with a number of popular apps for the mobility-impaired:

- **WheelMap** categorizes buildings and public spaces as fully accessible, partially accessible or not accessible for wheelchair users.
- **WheelMate** lists whether toilets and parking spaces are wheelchair-accessible.
- **Uber** lets individuals choose a wheelchair-accessible “taxi” ride.
- **Tecla Access** enables users to control their devices, electric wheelchair and smart home through motions, eye blinks and puffing.

Apple App or Google Play stores
The IC\-10-CM Code Book for Physicians, 2018 1st Edition
Author: Anne Casto
Publisher: American Health Information Management Association (AHIMA)

This book explores the effects a challenging disability or illness can have on the mind and personal relationships, and how friends, family and professionals can help. It takes a candid look at how discomfort caused by an illness can strain a relationship between partners, families and professionals, as well as how understanding feelings of guilt or shame can transform a situation or relationship. The insights and advice are intended to help children and adolescents overcome anxiousness caused by a parent’s condition, improve communication between partners and family members, and increase professionals’ awareness of how a client feels about his or her situation.

New and Useful Reading

Lubkin’s Chronic Illness: Impact and Intervention, 10th Edition
Author: Patricia D. Larsen
Publisher: Jones & Bartlett Learning

This is intended as a teaching tool to help nursing students understand the impact of chronic Illness on both patients and families. Now in its 10th edition, the book takes a practice-based approach by covering impact and issues, as well as interventions and outcomes. Each chapter employs a theoretical approach to the concept followed by an overview of the impact, nursing interventions and potential outcomes. To bring content to life, it takes a personalized approach featuring real-life stories and scenarios focused on the individual’s experience with chronic illness. The journey of the author and her husband through cancer is chronicled throughout a number of chapters. The book is mapped to all three levels of competencies (BSN, MSN and DNP) and includes updated evidence-based practice boxes, case studies and discussion questions.

Thriving in the Workplace with Autoimmune Disease: Know Your Rights, Resolve Conflict, and Reduce Stress
Author: Holly Bertone
Publisher: Amazon Digital Services

This book explores the effects a challenging disability or illness can have on the mind and personal relationships, and how friends, family and professionals can help. It takes a candid look at how discomfort caused by an illness can strain a relationship between partners, families and professionals, as well as how understanding feelings of guilt or shame can transform a situation or relationship. The insights and advice are intended to help children and adolescents overcome anxiousness caused by a parent’s condition, improve communication between partners and family members, and increase professionals’ awareness of how a client feels about his or her situation.
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- Read the issues anywhere at any time on all of your digital devices (smartphone, computer, iPad, tablet)
- Quickly access all published articles
### Ataxia Telangiectasia (A-T)
**WEBSITES**
- A-T Children’s Project: [www.atcp.org](http://www.atcp.org)

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

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

### Guillain-Barré Syndrome (GBS)
**WEBSITES**
- GBS/CIDP Foundation International: [www.gbs-cidp.org](http://www.gbs-cidp.org)
- The Foundation for Peripheral Neuropathy: [www.foundationforpn.com](http://www.foundationforpn.com)
**ONLINE PEER SUPPORT**
- GBS Support Group: [www.gaincharity.org.uk](http://www.gaincharity.org.uk)
- GBS/CIDP Foundation International Discussion Forums: [forum.gbs-cidp.org/forum/main-forum](http://forum.gbs-cidp.org/forum/main-forum)

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

### Kawasaki Disease
**WEBSITES**
- American Heart Association: [www.heart.org/HEARTORG/Conditions/More/CardiovascularConditions/Childhood/Kawasaki-Disease_UCM_308777_Article.jsp#T1T2boePWE0](http://www.heart.org/HEARTORG/Conditions/More/CardiovascularConditions/Childhood/Kawasaki-Disease_UCM_308777_Article.jsp#T1T2boePWE0)
- Kawasaki Disease Foundation: [www.kdfoundation.org](http://www.kdfoundation.org)
- KidsHealth: [kidshealth.org/parent/medical/heart/kawasaki.html](http://kidshealth.org/parent/medical/heart/kawasaki.html)

### Myositis
**WEBSITES**
- The Myositis Association: [www.myositis.org](http://www.myositis.org)
- International Myositis Assessment and Clinical Studies Group: [www.niehs.nih.gov/research/resources/imacs](http://www.niehs.nih.gov/research/resources/imacs)
**ONLINE PEER SUPPORT**
- The Cure JM Foundation: [www.curejm.org](http://www.curejm.org)
- Myositis Association Community Forum: [tmacommunityforum.ning.com](http://tmacommunityforum.ning.com)
- Myositis Support Group – UK: [www.myositis.org.uk](http://www.myositis.org.uk)

### Multiple Sclerosis (MS)
**WEBSITES**
- Multiple Sclerosis Association of America: [mymsaa.org](http://mymsaa.org)
- Multiple Sclerosis Foundation: [www.msfocus.org](http://www.msfocus.org)
- National Multiple Sclerosis Society: [www.nationalmsociety.org](http://www.nationalmsociety.org)
**ONLINE PEER SUPPORT**
- Friends with MS: [www.FriendsWithMS.com](http://www.FriendsWithMS.com)
- MSWorld’s Chat and Message Board: [www.msworld.org](http://www.msworld.org)

### Myasthenia Gravis (MG)
**WEBSITES AND CHAT ROOMS**
- Myasthenia Gravis Foundation of America (MGFA): [www.myasthenia.org](http://www.myasthenia.org)
**ONLINE PEER SUPPORT**
- Genetic Alliance: [www.geneticalliance.org](http://www.geneticalliance.org)

### Myositis Support Group – UK
**WEBSITES**
- [www.myositis.org.uk](http://www.myositis.org.uk)

### Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)
**WEBSITES**
- PANDAS/PANS Advocacy and Support: [www.pas.care](http://www.pas.care)
- PANDAS Network: [www.pandasnnetwork.org](http://www.pandasnnetwork.org)
- Midwest PANS/PANDAS Support Group: [www.midwestpans.com](http://www.midwestpans.com)

### Peripheral Neuropathy (PN)
**WEBSITES**
- Neuropathy Action Foundation: [www.neuropathyaction.org](http://www.neuropathyaction.org)
- Western Neuropathy Association: [www.pnhelp.org](http://www.pnhelp.org)
- Neuropathy Alliance of Texas: [neuropathyaliancebx.org](http://neuropathyaliancebx.org)
- The Foundation for Peripheral Neuropathy: [www.foundationforpn.com](http://www.foundationforpn.com)

### Primary Immune Deficiency Disease (PIDs)
**WEBSITES**
- Immune Deficiency Foundation: [www.primaryimmune.org](http://www.primaryimmune.org)
- The National Institute of Child Health and Human Development (NICHD): [www.nichd.nih.gov/Pages/index.aspx](http://www.nichd.nih.gov/Pages/index.aspx)
- American Academy of Allergy, Asthma & Immunology: [www.aaaai.org](http://www.aaaai.org)
- International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: [www.ipopi.org](http://www.ipopi.org)
- New England Primary Immunodeficiency Network: [www.nepin.org](http://www.nepin.org)
- Rainbow Allergy-Immunology: [www.uhospitals.org/rainbow/services/allergy-immunology](http://www.uhospitals.org/rainbow/services/allergy-immunology)
**ONLINE PEER SUPPORT**
- IDF Common Ground: [www.idfcommonground.org](http://www.idfcommonground.org)
- IDF Discussion Forum: [idffriends.org/forum](http://idffriends.org/forum)
- IDF Friends: [idffriends.org](http://idffriends.org)
- Jeffrey Modell Foundation Facebook Page: [www.facebook.com/JMFworld](http://www.facebook.com/JMFworld)

### Scleroderma
**WEBSITES**
- Scleroderma Foundation: [www.scleroderma.org](http://www.scleroderma.org)
- Scleroderma Research Foundation: [www.srfcure.org](http://www.srfcure.org)
- Johns Hopkins Scleroderma Center: [www.hopkinsscleroderma.org](http://www.hopkinsscleroderma.org)
**ONLINE PEER SUPPORT**
- International Scleroderma Network: [www.sclero.org/support/forums/a-to-z.html](http://www.sclero.org/support/forums/a-to-z.html)

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