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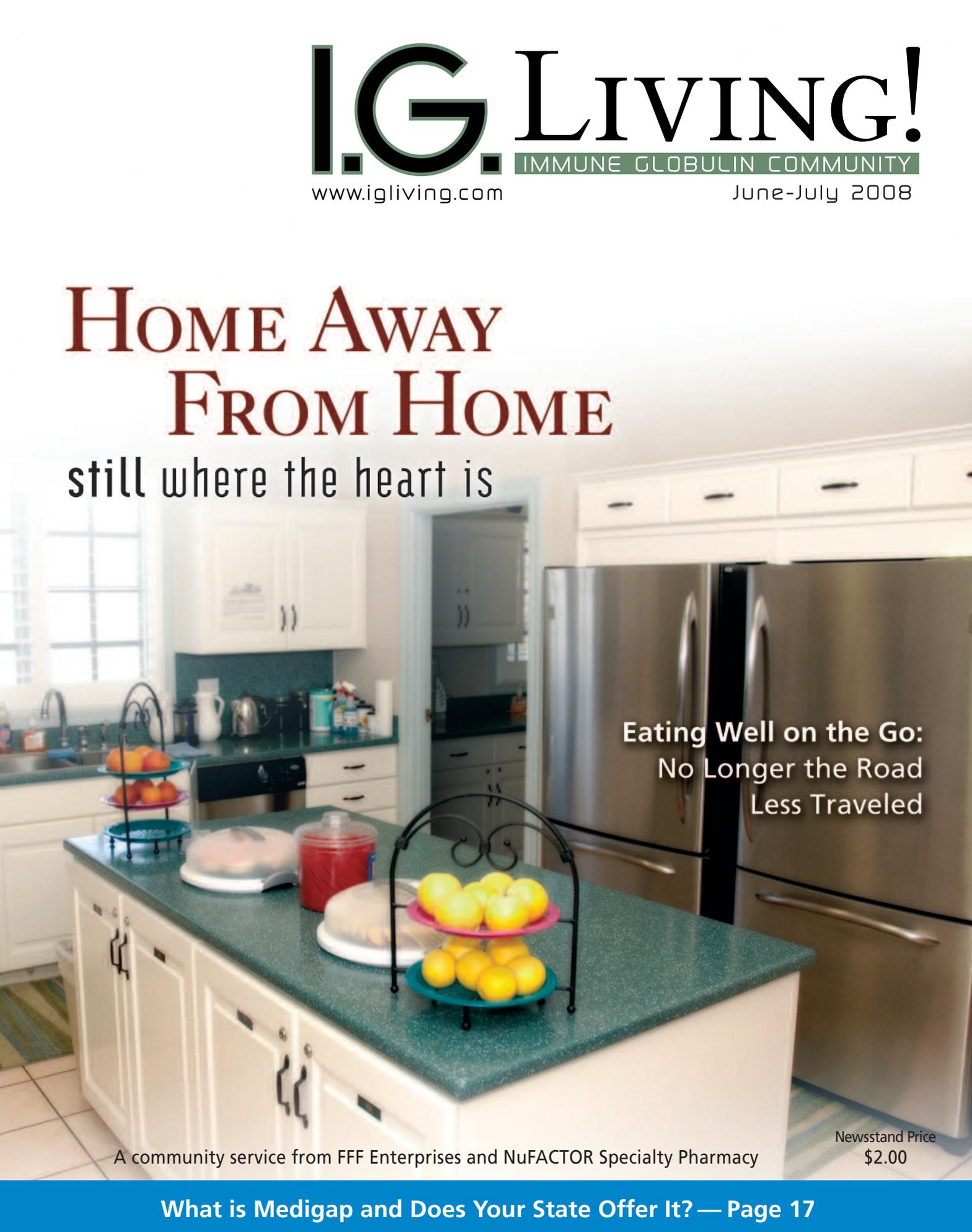
IMMUNE GLOBULIN COMMUNITY

www.igliving.com

June-July 2008

HOME AWAY FROM HOME

still where the heart is



Eating Well on the Go:
No Longer the Road
Less Traveled

A community service from FFF Enterprises and NuFACTOR Specialty Pharmacy

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\$2.00

What is Medigap and Does Your State Offer It? — Page 17

Our mission is to support the IG community through education, communication and advocacy

A community service from FFF Enterprises and NuFACTOR Specialty Pharmacy

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Publisher **Patrick M. Schmidt**

Editor **Amanda M. Traxler**

Assistant Editor **Cheryl Brooks**

Copy Editor and Proofreader **Jackie Logue**

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Graphic Artists **Allan Bean**

Caroline Carlson

Kim Kinman

Contributing Writers

Lauren Gerstmann, MPH

Cheryl L. Haggard

Mark T. Haggard

Michelle Freeman

Zachary Pugh

Jessica Schulman, PhD, MPH, RD

Jim Trageser

Michelle Vogel, MPA

Shirley German Vulpe, EdD

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Please direct editorial, advertising and marketing communications to
41093 County Center Drive
Temecula, CA 92591
Ph: 800-843-7477

Email: editor@igliving.com
www.igliving.com

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About IG Living

IG Living is the only magazine dedicated to bringing comprehensive healthcare information, immune globulin information, community and reimbursement news, and resources for successful living directly to immune globulin consumers and their healthcare providers.

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Amanda M. Traxler takes the helm at *IG Living*. See her editorial on Page 4.



Join an IGL Readers Group!

We hear from many, many patient and family member readers who would like to connect with others in their geographic areas—to share their experiences living with chronic diseases or maybe just to have a cup of coffee with folks who understand.

We can help you determine if there's a patient organization support group in your area or help you start an IG Living Readers Group.

To join a group or start one in your own area, visit www.igliving.com and click on IGL Readers Groups.

Hello, IG Community!

My name is Amanda Traxler, and I am IG Living's new editor.

Coming onboard has been exciting, and I'd like to thank everyone who has helped ease the transition. This includes staff, contributing writers, and the person who has made this entire undertaking possible, our publisher, Patrick M. Schmidt. The compassion that underpins *IG Living* has been apparent from my first day.

To learn about the IG community, I've been reading, and rereading, back issues. The vision, passion and dedication of my predecessor shine through in each issue, and I am grateful for the opportunity to take the wheel from here.

My greatest source of inspiration, however, is you—our readers. The resilience and strength in the IG community is astounding. The challenges faced daily range greatly, from those associated with specific conditions to those dealing with reimbursement. Yet the courage you display is remarkable. It's a valor that I've never had to muster, and which I find deeply admirable. The inspiration I draw from this community is palpable. And with it, I intend to serve you the best I can—sharing not just your stories, but also medical knowledge and information to help you advocate for the best possible care.

It's a privilege to be here, and I look forward to hearing from you—whether it be concerns, feedback, suggestions, or a simple hello—please email me at editor@igliving.com.

Document!

Having to put up a fight to receive needed care when ill is a travesty. And yet, as many of you know all too well, it's also sometimes reality. As stories about appeals continue to increase, the importance of documentation becomes even clearer. Everyone should document everything relevant to their care—even those who are not currently having access or reimbursement issues. It may save a bundle of time if, down the road, reauthorization of treatment is required or your employer changes insurance.

In the upcoming issue, we will feature how to navigate the appeals process. But step number one is not hard to guess: Document! To quote from the December/January 2008 Reimbursement Q&A: "Comprehensive documentation



is an important element of a successful appeal, so all communications with your insurer should be in writing, whenever possible, including your request to appeal a determination." Document every call with insurance representatives: Keep a journal and write down the date and time, name of the person(s) with whom you spoke, his or her title, phone number, and important details of the conversation.

Also, document discussions with your medical providers. In cases where treatment is not considered medically necessary, having evidence of a doctor's support is vital. Further, get looped into the initial authorization process between your doctor and your insurance company. I've been hearing anecdotally that when some doctors request authorization for medical treatment for patients, the insurer sends the doctor (not the patient) the denial. As a result, patients don't realize until time is already lost that a denial has occurred. So if your doctor wants to try a new treatment, stay abreast of the communication between him or her and the insurance company. You can do this by asking for copies of all letters, lab results and communication that go between your doctor and the insurance company. Keep your own copies organized in a binder. This will help ensure that no time is wasted if it becomes clear that an appeal will be necessary. Ideally, having this information at your fingertips will help resolve the issue—and get you the necessary care—as quickly as possible.

Look for more details in the August/September issue. However, if you have immediate concerns or problems, please contact us via the Reimbursement Q&A or Ask Kris on www.igliving.com.

Finally!

On May 1, Congress finally passed the Genetic Information Nondiscrimination Act, which protects against discrimination by health insurance providers or employers based on genes or genetic predisposition to disease or chronic conditions. The bill is now headed to the President, who's expected to sign. ■

Readers Write

Your feedback, opinions, suggestions and anything else you'd like to share are important! Email us at editor@igliving.com or visit www.igliving.com and go to the Contact Us page to send your comments.

Dear Editor,

I'm responding to the letter from Michelle in Florida. I can really relate to her revelation that dealing with polymyositis is a true exercise in patience. I was diagnosed over eight years ago, and the words my doctors used were, "Don't worry, with these high doses of prednisone, we'll have you back to 90 percent of your old self in no time." Considering that I was totally disabled in a wheelchair, this sounded pretty good to me. What they didn't tell me was that, yes, I'd be back to 90 percent—that is, until my first flare-up. My CK levels and strength have been on a constant roller-coaster ride since then. Flare-up; increase or add new meds; get better; taper the meds; then another flare-up. I take prednisone, methotrexate, Imuran and monthly IVIG. Who knows which, or which combination, is currently holding me in my 90 percent mode, but I'll keep taking these meds if they keep letting me be nearly normal.

Two other notes: One is that I'd encourage Michelle to go to The Myositis Association website, www.myositis.org and enter her ZIP code. She might just get my email address and an invitation to join a local Keep In Touch (KIT) group that helps myositis patients stay in touch with others in the same boat. If not me, there are at least two other KITs in Florida that would welcome her.

Second, in all the articles about side effects of IVIG I've read in *IG Living* (a great magazine, by the way), there's one prevention I don't think gets enough attention. After three years of monthly IVIG, and battling subsequent migraines, I've finally hit on what appears to be the way to hold the headaches at bay. When I first started having the headaches, my rheumatologist prescribed Benadryl and Tylenol before the infusion. That seemed to work for a while. Then the headaches returned and he additionally prescribed Decadron and that too seemed to help, but not for long. Then, at a KIT meeting, when I shared this story with another polymyositis patient, she said that we need to not just hydrate but superhydrate, both before and during the infusions. I now glug down many, many bottles of water the day before and during the two days of my infusion. So far, so good! And it's not another drug; it's just something that I probably ought to be doing anyway!

Keep up the good work!
—Marianne, Florida

I've read a few articles from one of your publications, and found them very informative. I would like to receive my own copy, since my daughter is dealing with this condition. I am also a physician specializing in family medicine, and found your publication very impressive!

—A reader in Georgia

I realize that this is a very special 'specialty' publication, but I never ever expected that the staff could be so responsive! Every staff person and consultant I have encountered has been professional and helpful beyond any expectation imaginable. I have tended to ask some questions of the staff that most folks don't tend to think of, as I am a curious person about the whole process of this "stuff" called IVIG that helps me function. I have seen bits of my questions answered in subsequent issues. I am very grateful that my questions and concerns have been heard! Not only that, the support that the staff and consultants provide is extraordinary! They have helped many I know with IG issues that could not ever have been resolved otherwise. I have recommended this publication to many, here in the United States and internationally, as a resource. All responses are positive about the publication and the assistance—the key to happy IG campers. My best to you and all the wonderful staff for this new year! May we all be healthier as a result.

—Joan, Virginia

I saw a copy of *IG Living* in my doctor's office. It provided me with more usable information on my condition than any other source I have been able to find. "Barriers to Effective Support" (*IG Living*, Oct-Nov 2007) was enormously helpful. Bravo! Keep up the good work.

—William, Florida

How About the Well Child?

By Zachary Pugh

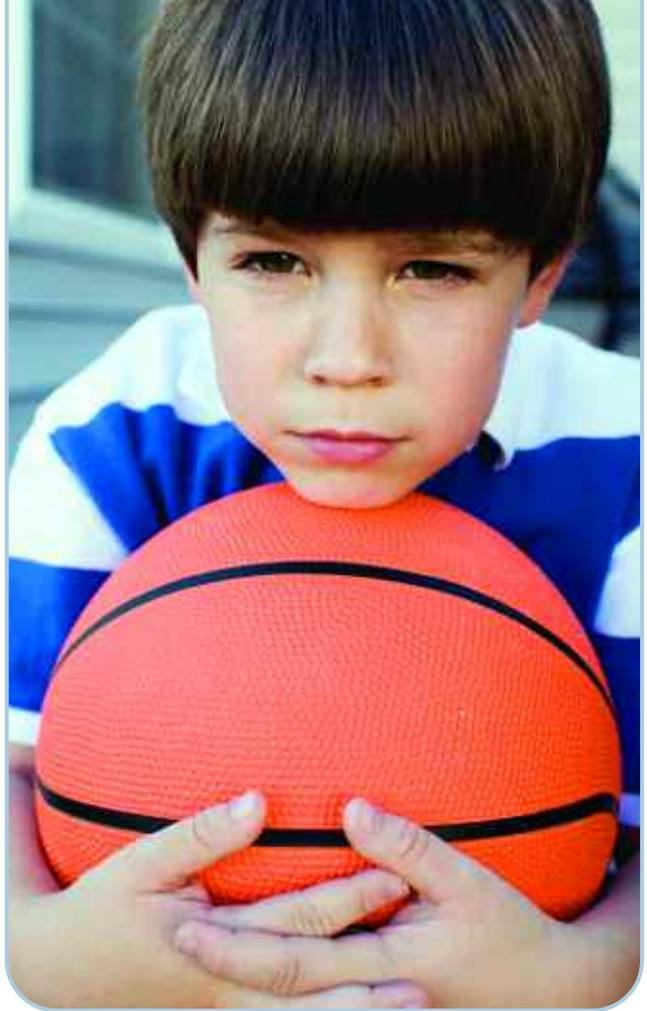
There is a truth that seems to be proven through the ages: In families with more than one child, a delicate balance exists between siblings vying for their parents' attention. When one sibling is chronically ill, the stability of that balance can easily become skewed, leaving one or more child feeling unattended. The result can be an unintentional alienation, which can lead to long-term problems for the well sibling(s).

For these families, and specifically the well siblings, resources are available to help them through this often confusing and chaotic time. One of the most helpful is the Sibling Center in San Francisco.

The Sibling Center was established as part of the California Pacific Medical Center to support the needs of these well sibling(s). "It's a preventive model so that theoretically anytime a child is diagnosed with a serious illness, the siblings are referred to the center, although we're actually getting them at all different points because families find out about us at different points," says Joanna Fanos, PhD, founder and director of the Sibling Center and professor of pediatrics at Dartmouth Medical School.

Dr. Fanos' work exceeds what most others have done in this area. "I've done research the last twenty years on the impact of serious childhood illness on the siblings. After publishing article after article in medical journals saying siblings need help from the time of diagnosis, nothing [was] being done," she says. "So I set up that center back in July of 2002 and got a grant from my hospital in San Francisco."

The center's focus is on the well child and the parents.



"The mission is really to prevent the kind of long-term difficulties that I kept documenting in the research," Dr. Fanos continues. "We don't work with the child who is ill. We work with the parents and their siblings."

The well sibling often feels an array of emotions, which can include anxiety, depression, survivor's guilt and empathy. The siblings of chronically ill children also tend to have a unique maturity and passion born from experiencing something of this magnitude at such a young age. Dr. Fanos observes that there is often "a high development of empathy and wanting to help others."

How can you tell if your child is struggling with these emotions? According to Dr. Fanos, there are tell-tale signs parents can recognize.

"There is anxiety about being or getting sick, or pretending to be sick. In other words, they might be a little hypochondriacal because they've grown up in a family where they know experientially that something bad can go wrong. They don't have any illusions of health that a regular kid would have," she says.

In her experience, Dr. Fanos has seen a decline in the number of cases where children pretend to be sick. Essentially, this is a way to "get love and attention from

the parent, so it's understandable that they might try to sort of exaggerate their illness," she says, adding that she "sees much more of the other, though. They're worried that they might have some horrible thing and they don't articulate their fear to anybody."

Other signs that indicate a well child is struggling include excessive dependency, withdrawal, acting out, school problems, difficulty concentrating, loss of appetite, overeating or regressive behavior, excessive concern for others, frequent accidents, sleeping difficulties (nightmares, which are a trauma response) and, in later years, substance abuse.

Another clear indicator that a well sibling is struggling is when the child "takes on a developmentally inappropriate adult role," Dr. Fanos explains. "In other words, you get the kind of caregiver role where they become parentified when they're just children, so they lose their childhood."

One of the more serious red flags is more subtle: the foreclosure of future goals and hopes. "Often parents won't talk to their children about the future because they want to live one day at a time. And that's great for the child who's sick and [who] may even have a foreshortened life span," Dr. Fanos explains. "But it's not good for the well sibling who will grow up."

The Program

The Sibling Center offers its services free of charge to all families, whether they are receiving healthcare or not. The program comprises four sessions and a follow-up meeting. All sessions are facilitated by advanced trainees who are either doing their post-doctoral clinical hours for their clinical psychology license, or they're doing their post-master's hours. Their skill level tends to be pretty high by the time they get to the center. The center also requires that they undergo various training related to specific sibling issues before they see families at the center.

"[In] the first session, which is one of the longer ones, the therapist will meet with the parents to get a sense of what the disease or the disorder or the condition is and what they [the parents] have told their well children about it," Dr. Fanos explains.

This crucial first session gives the therapist a chance to investigate the issues and concerns the family faces and how they are handling them. Of course, the therapist's analysis keeps the well sibling in the forefront.

"In sessions two and three, the well sibling is seen alone without the parents, and the spacing is usually a couple

weeks in between ... two or three weeks, depending on the schedule of the family," Dr. Fanos says.

After the third session, the parents and siblings are "reconstituted back together," according to Dr. Fanos. "Whatever insights have been garnered during the other sessions [are] discussed within the family, and the family is told that if anything comes up before the three- to six-month follow-up, they should feel free to call the therapist."

Three to six months after the last session, the therapist will phone the family for an update. If things are not going well, the therapist will refer the family to an appropriate professional.

Free Service

Though unusual in this day and age, this high-quality therapy conducted by advanced trainees is offered free of charge. At one point, the services were charged to the family on a fee-for-service basis, but fees were eliminated because families with chronically ill children are often financially burdened.

The no-charge strategy bypasses insurance companies altogether. "We never wanted to deal with insurance companies because we didn't want to pathologize the sibling. We aren't saying these kids have a problem, we're saying often we find that it's difficult for the sibling and so we want to get in there early and see how they're doing," Dr. Fanos urges. "It's psycho education, really ... rather than 'there's something wrong going on here.'" ➤



Advice for Parents

Often, when a family first undergoes the trauma of learning that a child has a chronic illness, the parents explain the illness to the other children. This is where it typically stops. "They don't ever really have that conversation again, when, in fact, they really should be very available and create an environment where it's OK for the sibling to ask questions as they grow older and have higher cognitive abilities to understand what the parents are saying," Dr. Fanos explains.

Misinformation and incorrect perspectives about the illness need to be fixed for the well sibling, who typically does not clearly understand the condition or its cause. Parents of chronically ill children should be proactive about learning about any myths or distortions about the illness a well child may have. "You can't assume your child understands [the illness] because they probably don't," says Dr. Fanos. "The well child might think 'Oh no, little Johnny has cancer because I yelled at him that day, or I hit him in the face that day and that caused the brain tumor.' They don't understand that they had nothing to do with it."

It's important to realize that what the well child wants most is attention and time with their parents. Parents can sometimes mistake their child's desire for interaction, or overcompensate for the lack of it with expensive and elaborate gifts or trips to appease their children. "Often

what the well sibling will want from the parent is something very simple and inexpensive," she explains. "Mainly they want their own special time with the parent because they see that the sick child has their own special time with the parent, even if it's going to doctors. Somehow that looks good because they get their alone time."

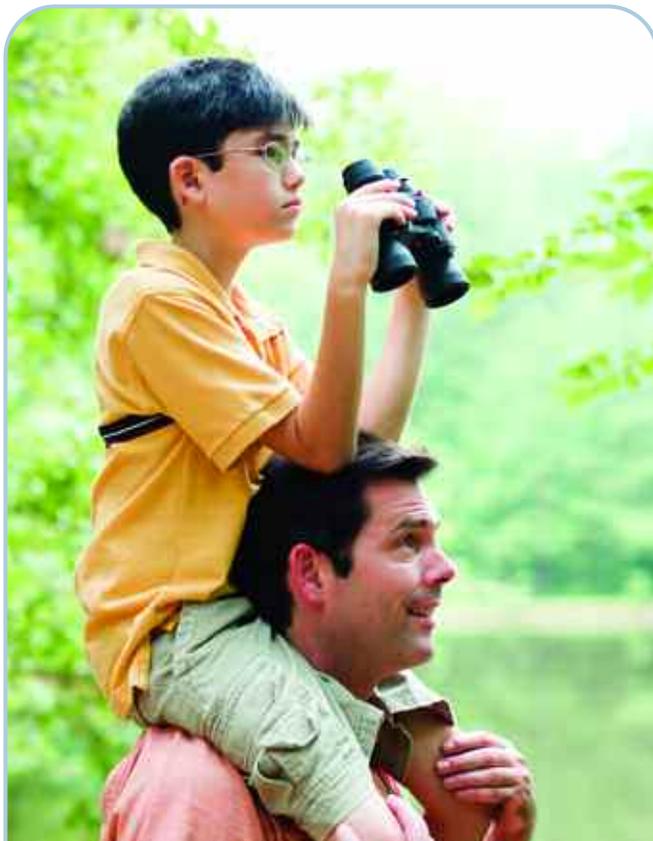
In her experience, Dr. Fanos has seen that the time spent, no matter how simple the event, is time *well* spent—simply because the parent is there. At the Sibling Center, the therapist explores with the child to determine what it is they really want. "It's usually things like 'Gee, I'd love to go have pizza with Dad.' In our experience the parent always says 'Of course we can do that.' It's not that they want a trip to Paris, they want a slice of pizza. Mainly they just want to be alone with Mom or Dad or both so that they get their special time."

Another thing that typically happens in these families is holiday abandonment. "Families sometimes will abandon holidays, depending on how sick the child is. Sometimes they'll be so focused that holidays will go by the wayside, family routines or schedules will go by the wayside, and that's really not a great thing," Dr. Fanos explains. "If the family can keep the routines and schedules, and honor holidays and traditions as much as possible, as they would normally, that would be great."

Parents should also not overlook the importance of nurturing their marriage. They need to care for their own marital relationship or they won't have any energy left to care for the sick child or the well sibling. If they do have problems and they feel they need help, they should seek professional support.

The most important thing, explains Dr. Fanos, is maintaining communication within the family. If the lines of communication are open, and the issues are being addressed in an atmosphere that is open and comfortable for the well sibling, the child is more apt to articulate his or her needs. "Create an atmosphere where it's OK for the sibling to come to the parent and ask questions," she says. "And be pretty proactive about what they understand or not because they may have distortions and myths that aren't true."

"I think those families that have enough energy left over to say, 'Am I doing everything for the well sibling?' are probably doing everything for the well sibling. If it's in their mind-set, then I bet it's fine." ■

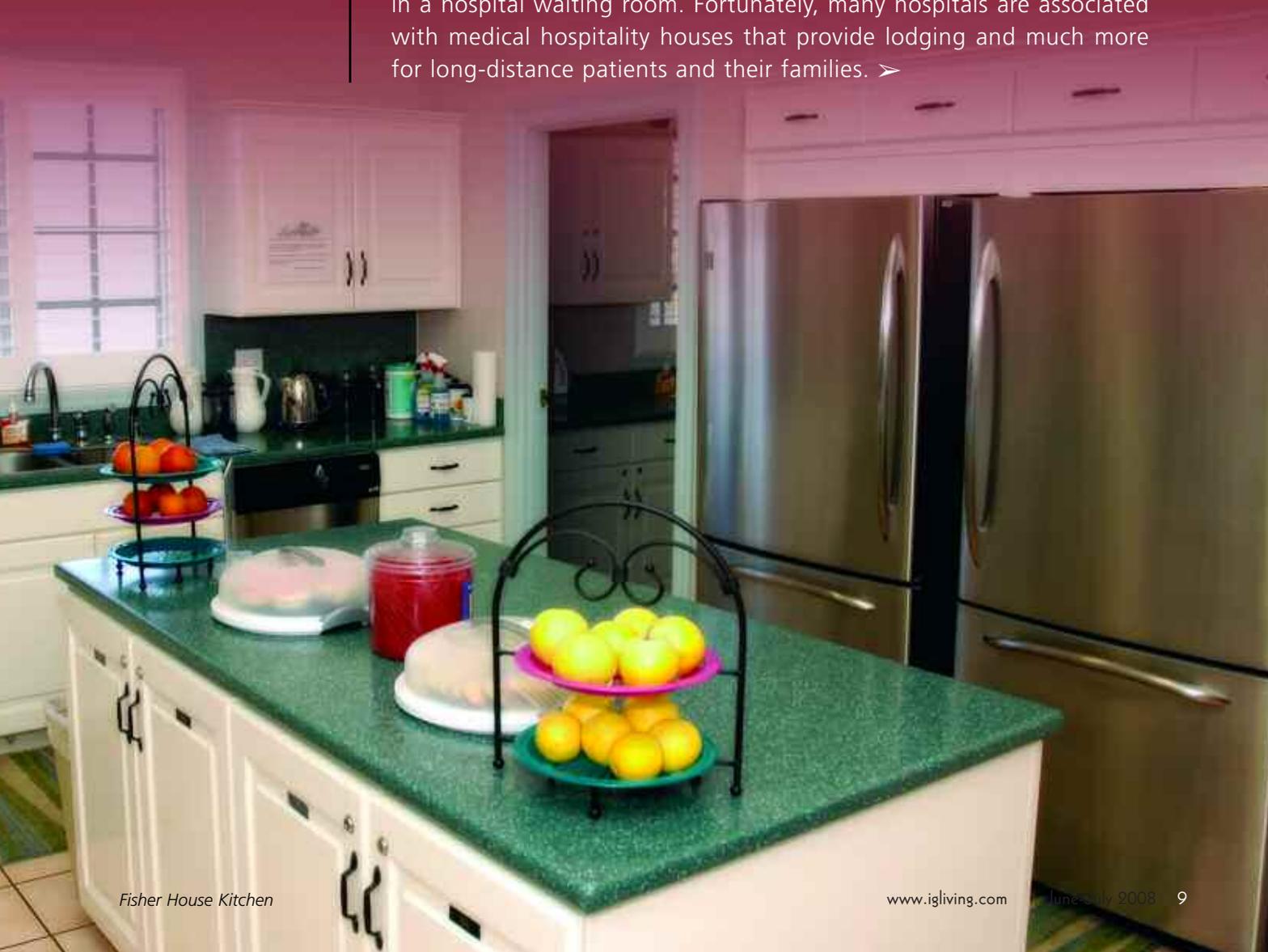


HOME AWAY FROM HOME

By Lauren Gerstmann, MPH

still where the heart is

Hospitality houses are like friends who'll take you in when you need to travel for medical care. It is easy to move heaven and earth to care for a sick loved one, but it is not as easy to move them or yourself. Hotel bills rack up quickly, and you can only spend so many nights on a chair in a hospital waiting room. Fortunately, many hospitals are associated with medical hospitality houses that provide lodging and much more for long-distance patients and their families. ➤



Hospitality houses can be located through the National Association of Hospital Hospitality Houses (NAHHH). More than 150 hospitality houses in the United States are affiliated with NAHHH. Each NAHHH house “assures that a homelike environment is provided to persons who must travel to be with a patient or to receive necessary outpatient care.”¹ Just as helpful, NAHHH keeps a comprehensive database of all the U.S. medical hospitality houses—even those that aren’t affiliated with the association. To view their listings online, visit www.nahhh.org. You can search by state or by house name.

Some of the best-known programs, the Ronald McDonald House Charities and Fisher Houses, are not affiliated with NAHHH. Though these organizations’ houses are listed on the NAHHH website, more extensive information can be obtained by contacting the specific organization or an individual house. Ronald McDonald Houses serve families traveling for medical care for their children, whereas Fisher Houses serve military families.

The Ronald McDonald House Story

The first Ronald McDonald House was founded in Philadelphia in 1974 by Philadelphia Eagles player Fred Hill and Dr. Audrey Evans, a pediatric oncologist and philanthropist. Hill, whose daughter had won her battle against leukemia, wanted to help the hospital that saved her life. Ten years later, the house had made so much of an impact that a larger charity was formed to create similar hospitality homes near other children’s hospitals. The charity was established “in memory of McDonald’s founder and children’s advocate, Ray Kroc ... to help seriously ill children and their families and to continue the success of the first Ronald McDonald House.”² The organization is decentralized, with each house operating independently but following guidelines and standards set by a global headquarters financed by the McDonald’s corporation. Headquarters provide general advertising and fundraising assistance, but each house is a separate entity with its own board of trustees.

The McDonald’s standards ensure that Ronald McDonald Houses, which host families from all over the world, are cheerful spaces. Patients and their siblings have bountiful play space, and guests can share meals and company in

beautiful common rooms. Patients can stay there when they leave the hospital, and families can stay there during their children’s hospital stay. Each Ronald McDonald House is just minutes away from doctors and emergency facilities. The houses are designed with large kitchens that have multiple cooking stations. Since the Boston Ronald McDonald House pioneered bone marrow/stem cell transplant suites in 1985, many of the houses now include special accommodations for the most vulnerable patients and their families. Volunteers bring food and entertainment to the guests. At the Ronald McDonald House in Washington state, volunteers donated so much time that in 2006 their hours totaled the equivalent of 11 full-time employees.³

In Cincinnati, the Ronald McDonald House is within walking distance to the Children’s Hospital and the zoo. On some evenings, zoo staff bring animals to visit families. Jennifer Goodin⁴, executive director of the Ronald McDonald House in Cincinnati, sees the house as a place where she “hopes [guests] would feel as comfortable as they do in their own home,” and that they would “allow staff and volunteers and the whole community to take care of them at this difficult time and that they would find strength in the other families that are going through the exact same thing ... that they will find hope.”

In addition to amenities such as Internet connections, playrooms and communal areas, the emphasis is on creating a safe sense of community. Jennifer successfully campaigned to shut down a dangerous bar across the street. “The police had all these statistics, and there was something like 600 police calls to that site each year; something was going on every day,” including drug deals and gang activities.



Jennifer Goodin plays with Ronald McDonald House children

¹ www.nahhh.org.

² www.rmhc.org/about/history-of-ronald-mcdonald-house-charities.

³ www.rmhcseattle.org/news/house_facts.php.

⁴ Interviewed on 1/31/08.



Fisher House living room, bedroom and common area

Due to her petitioning, the state board of liquor control refused to renew the bar's liquor license.

Emotion is audible in Jennifer's voice when she talks about the house and its occupants. Next to the birth of her own children, the most memorable day in Jennifer's life was the day she helped deliver a baby boy on the lobby sofa. A woman from Indiana with a high-risk pregnancy was staying in the house so that as soon as she gave birth, her son could be taken for heart surgery. The woman went into labor so quickly that she couldn't make it to the hospital next door. The whole staff supported the birth, wiping her forehead, coaching her breathing and calling paramedics to take the baby to the hospital right away. Years later, Jennifer still keeps in touch with the family,

The Cincinnati house, which doesn't have a particular emphasis, works closely with the entire children's hospital next door. The hospital draws patients from all over the globe, and the physicians know that it is critical that their families have a safe and affordable place to stay near the hospital. When Marc Leavitt, a renowned surgeon, was recruited to Cincinnati Children's Hospital, he visited the

Ronald McDonald House before making his decision to join the surgical team.

The house always has a waiting list of about four to five days. Given that families cannot make a reservation in advance of their visit to Cincinnati, most find other accommodations for at least part of their stay. This, however, is about to change. With the support and endorsement of the hospital, a much-needed expansion is about to break ground. The Cincinnati house is adding 30 rooms (bringing the total to 78) with a targeted completion of February 2009. Fifteen of the new rooms will be transplant suites. The goal is that it will still feel like one house: The existing kitchens and dining rooms will expand, maintaining the social heart of the house. The expansion should reduce the waiting list from about 25 to 30 families per night to five to 10.

The support families feel in the Ronald McDonald House cannot be overestimated. One mother says, "From day to day, I did not know whether my 2-year-old son would need more surgery or be able to go home. We were separated from his father and his brother, but he was happy in ➤

the house. Six months later, he asks me when he can return to the 'old McDonald House' to see his friends and ride the elevator!" She continues: "It was incredibly moving and inspiring to meet other families at the house and to experience the caring of staff and volunteers. One Friday night, after we learned we would soon go home, we wanted to express our gratitude with Shabbat candles, wine, and challah [ceremonial bread]. All of our new friends joined us—Christian, Jewish, Muslim. We put our arms around each other and broke bread in honor of our children."

The Fisher House Story

Active-duty military personnel are often on the move. Serving all over the world, some must travel great distances for specialized medical care. To assist military personnel and their families, the Fisher House program was developed in 1990.⁵ Partnering with private philanthropists, Zachary and Elizabeth Fisher originally endowed the organization. Each major military medical center has at least one Fisher House. Designed in a similar fashion, the houses meet the Fishers' standards; most importantly, each feels like a home.

Propelled in part by the increased number of combat casualties in Iraq, the program is now expanding. But, the program is not only for injured military personnel.

A significant portion of military families who use the Fisher House are traveling for treatment of a civilian family member.

As director of the Fisher House as well as of Morale, Welfare and Recreation (MWR), Tim Scully wears two hats. The two programs at the Naval Medical Center in San Diego offer support and respite, with Fisher House guests able to use the workout facilities at MWR to relieve stress.

The Fisher House provides housing that is almost "like grandma's house or a bed and breakfast." Guests have their own rooms, but share a kitchen, living room and dining room. Amenities such as common rooms, secluded areas, televisions, Internet and workout facilities are available. Tim's goal is to "make sure that the [high] standards of the Fisher House are met ... We are not trying to provide something that is basic housing. We are trying to provide the best housing—the most comfortable environment with the most compassion." He continues: "Our role really is to make sure that the families staying here have every care taken care of. If we don't provide it, we find it for them."

The San Diego house follows Navy guidelines, which give priority to active duty soldiers and their dependents. But retired military personnel are also eligible. Although

this house gives priority to family members of combat victims, guests are just as likely to be visiting an ill patient as a wounded patient. According to Tim, the houses were built long before the war started, and will be here long after this war. The house is almost always full.

Despite the high occupancy rates, families are rarely turned away. If they cannot be accommodated at the house, the house will work with other organizations (including the Fisher House Foundation) to make sure that their hotel expenses are paid. But the demand keeps growing. Fortunately, more houses are in the works. Right now, there are 38 houses and five more are under construction. At the San Diego Naval Base, a second Fisher



A Fisher House

⁵ www.fisherhouse.org/aboutUs/aboutUs.shtml.

House is being built next to the first. They will be close enough together to share their outdoor spaces. The new house will add 11 rooms with slightly bigger rooms and kitchens. It was scheduled to be finished just in time for Memorial Day.

Fisher Houses were originally designed to offer short-term respite for acute medical events. In San Diego, the average stay is 12 days. When the program started, families made other arrangements if they needed housing for more than 30 days. According to Tim, this requirement is no longer enforced. Much is based on the needs of the guests and the needs of the patients. At an Army Fisher House in Washington, D.C., some guests stay for years (see Nick's story).

Tim talks about camaraderie at the house, and says it is inherent in the way the homes are designed. "Anytime I go over there, I tend to see at least two families together ... [The design] promotes that, and encourages people to eat together—fairly large kitchen ... incredible yard, gazebo ... areas where people can congregate, share stories, vent—whatever the case may be." It is no accident. From the beginning, Fisher Houses were deliberately designed to be more like private homes than hotels. They offer spaces for seclusion, and places to gather.

Tim's motivation is, in part, personal. He knows firsthand how important it is to have a place to rest and refresh when a loved one is hospitalized. In 1990, Tim's mother passed away from cancer three weeks after he learned of her diagnosis. "I was lucky enough to see her in the hospital before she passed away [but I] slept in the waiting room for a few days." He says that being able to rest, restore, and spend time with his brothers and sisters at a comfortable place would have been invaluable. Offering this type of respite to the Fisher House guests is his passion.

Occasionally, Tim becomes personally involved. "I do photography, so sometimes I offer my photography to them," Tim says. A beautiful photograph can create a special memory even of the most difficult times. "I am very proud to be a part of [this]," says Tim.

One house guest, Misty,⁶ stayed at the San Diego house while her husband had knee surgery. Caught up in the stress, Misty had not thought about where to stay while her husband recovered. His surgeon made arrangements for Misty at the Fisher House. The house was "really homey, I liked it a lot," says Misty. "I was really thankful to have a place to go where I could take a shower; I could rest



Fisher House dining room

if I needed to." It is a beautiful facility with comfortable furnishings, high-quality appliances, and all the current technology. Internet access at the house was a big comfort to Misty, because it meant she could keep in touch with her family. "It was kind of like a home away from home, because you could do your laundry there, and you could cook, and you could relax ... It was really nice."

Fisher Houses are financed through private funding and donations. Volunteers also donate their time. Volunteers at the San Diego house relandscaped the grounds. In Washington, D.C., a volunteer is building new cabinets. Most volunteers have an emotional connection to the houses or the military. But awareness is also increasing among high-profile celebrities and politicians. Larry King and Denzel Washington are major contributors, and California Gov. Arnold Schwarzenegger has visited.

The Ronald McDonald House and Fisher House programs are two examples of places to stay when traveling for medical care. Many other wonderful housing programs are out there that are dedicated to providing the comforts of home. They are easy to locate online through the National Association of Hospital Hospitality Houses, or by talking to a doctor or care coordinator at a hospital. Medical travel is inherently stressful, but small supports can make a big difference. ➤

⁶ Interviewed on 2/24/08.

Radel's Experience at the Ronald McDonald House

George and Corinne Brewer first met Radel Vasquez a few years ago when he came to the United States from the Dominican Republic for orthopedic surgery. The Brewers volunteered for Healing the Children,⁷ an organization that sponsors doctors and volunteers to travel around the world to provide medical care. The organization also sponsors children who cannot find care in their own country, making it possible for them to travel to the United States for medical care. For the past 25 years, the Brewers have hosted children who need acute care. After Radel's surgery, the Brewers thought they might never see him again. But as soon as he returned home, Radel developed signs of Fanconi anemia, a genetic disease that causes problems with a patient's stem cells and eventually leads to leukemia. Radel was accepted into a research study at Cincinnati Children's Hospital. Despite that the Brewers live in New Jersey, they once again stepped in to help. George had recently retired, so they felt that they could provide 8-year-old Radel with the time and intensive support that he needed. George says, "Normally we had kids come for a couple of weeks, a couple of months ... it was in our

home, and they were like one of our children. Going to Cincinnati was a big project."

In 2007, the Brewers and Radel spent 10 months at the Ronald McDonald House in Cincinnati. "That place is wonderful ... It was clean, friendly, [everyone was] reaching out," comments George. He felt strong support from the staff and volunteers. "There was a different meal, and a different cook, and a different provider [every night]." The Brewers felt supported not only by the house, but also by the whole community in Cincinnati. Their stay in Cincinnati soon lengthened from an expected 100 days to 250, but the house made it feasible. Radel even thinks of it as a positive experience. He told me, "It was good. [I liked] the people and all," and he enjoyed playing with the kids and toys there. George says that the staff and volunteers made his family feel at home. Radel would often sit behind the registration desk and pal around with the staff.

Radel was discharged from the hospital within a month of getting a curative bone marrow transplant. At the Ronald McDonald House, Radel was able to play with other children between his frequent hospital visits. He

Fisher House Facts

- Serving military families traveling to visit hospitalized relatives
- Whether patients stay in the house varies from house to house
- More than 10,000 military personnel and their families have been served annually since 1990
- All funding is raised privately; houses may not actively solicit donations
- Families do not pay for their stay
- Every major military hospital and many VA centers have houses
- 38 houses currently exist, five more are under construction

For more information or to make a donation, see: www.fisherhouse.org.

To learn about the San Diego Fisher House, see: www.nmcsd.med.navy.mil/service/services_view.cfm?csid=88 or call 619-532-9055. To learn about the Washington, D.C., Fisher Houses, see: www.wranc.army.mil/Visitors/visitservice/meals/fisher/Pages/default.aspx or call 202-545-3218.

Ronald McDonald House Facts

- Serving seriously ill children and their families who are traveling to children's hospitals for medical care
- Children may be traveling for inpatient or outpatient care
- More than 10 million children and their families have been served
- As long as the stay is medically necessary, there is no limit on length of stay
- Houses are not medical facilities, although patients may receive home healthcare services
- Houses are financed by private donations
- Families are asked to pay between \$5 and \$25 per night, but this fee is waived in cases of financial hardship
- 271 houses in 30 countries

For more information or to make a donation, see: www.rmhc.org.

For more information or to donate to the Cincinnati Ronald McDonald House, see: www.rmhcincinnati.org/home or call 513-636-7642.

⁷ For more on Healing the Children, please see www.healingthechildren.org or call 800-992-0324 or 509-327-4281.



Radel Vasquez

wore a mask to protect his recovering immune system, but many of the children in the house wear masks, so his illness did not set him apart or isolate him. The Brewers kept Radel occupied without jeopardizing his health. They took him on outdoor activities—to baseball games and to the zoo—and the Ronald McDonald House helped arrange tickets for everything. The entire time, Radel was in touch with his family back home. His mother has sole care of his siblings and is the only financial support that the family has, but she was able to travel to America to support Radel around the time of his transplant, and the Ronald McDonald House welcomed her as well.

“The people in the house were extremely good to him, and to us as well,” says George. They became close with staff and volunteers. When the Brewers returned to Cincinnati with Radel for a three-month follow-up, they went to dinner with one of the front-desk volunteers and her family. George still speaks with amazement about their experience in Cincinnati. “There is a waiting list for groups to provide meals [to the houseguests].”

Nick's Experience at the Fisher House

Just over a year ago, Nick lost his leg in the war in Iraq. Originally based out of North Carolina, Nick was shipped to the Walter Reed Army Medical Center in Washington, D.C., for medical care. When first out of the hospital, Nick stayed at the Malogne House on base. But the cramped quarters didn't have cooking facilities, and Nick wanted to be somewhere that would help him learn to be back in the “real world.”

The Fisher House was a great improvement. Nick has his own room and bathroom, as well as areas to cook, eat and socialize. Being around other veterans has been invaluable. “It really helps being surrounded by guys who are going through the exact same thing that I have.”

For Nick, the best thing about the Fisher House is that it is a community. The house in Washington, D.C., is small enough that Nick has gotten to know everyone there. Houses vary in character, and the manager of the house in Washington, D.C., discourages personal relationships between staff and guests. Even so, the staff has been warm. Everyone will “bend over backwards to help you.” Nick speaks particularly fondly of one man who “cares about the people here, and he doesn't mind showing it.” Right after Nick's surgery, he would run errands for Nick, knowing that Nick couldn't drive.

For Nick, the Fisher House is home. When he first arrived, his wife, Krystal, was on bed rest while pregnant with their first child, so his mother cared for him. Now his wife is with him. It isn't easy; their two children are staying with Krystal's mother in North Carolina—one is 15 months old and the other (their “happy accident”) is only 3 months old. But Nick is just out of surgery and is not far enough along to have the kids around. Hopefully, they will be able to join them soon. It will be a year before Nick is ready to leave his doctors and the Fisher House. In the meantime, the Fisher House is providing him with a place to create a new community and to reunite with his family while he heals.

Other Housing Options

If a hospitality house is not an option, alternatives exist. Some hospitals work with local hotels, apartment managers and corporate housing offices to provide significantly discounted short- or long-term lodging. A care coordinator can provide details about options. Also, those affiliated with a religious group should check if local resources can provide food, lodging or support. ■

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What Is Medigap and Does Your State Offer It?

By Michelle Vogel, MPA, and Michelle Freeman

Loretta of Arizona was shocked to learn that her disability excluded her from eligibility for Medigap, a federally subsidized private insurance option with the singular purpose of eliminating the difference between what Medicare pays for her treatment and the bill she receives from her provider. According to Arizona regulations, to be eligible for Medigap coverage, Loretta apparently needed to be receiving Medicare as a senior citizen rather than as a person with a disability. Investigating further, Loretta was even more shocked when she learned that Arizona is only one of several states that prohibit this vital gap coverage to the disabled. Moving to a state that allows coverage for persons with disabilities seems to be her only option.

How has the lack of availability of Medigap policies for disabled Medicare beneficiaries affected the immune globulin community?

1. Patients who do not have a secondary policy cannot afford the 20 percent co-pay for their infusions, especially in the hospital outpatient setting where 42 percent of Medicare beneficiaries have been receiving their IVIG since the implementation of the Medicare Modernization Act in 2005.
2. Providers have been forced to deny care to patients without Medigap plans or secondary plans because they cannot waive or wait for payment due to reduced reimbursement under the Medicare program.
3. Patients are going into debt, or going without therapy, due to the financial burden of expensive co-pays.
4. Patients living in states that offer Medigap policies for the disabled are not told that, on average, they must choose a policy within the first six months of being eligible for Medicare, or they lose the ability to purchase a plan until they are 65 years old.

Background on Medigap Policies

When Congress passed legislation creating the Medigap program, policymakers opted to allow states to decide

whether Medigap policies would include the disabled; to set eligibility guidelines; and to determine coverage for medical treatments under the new insurance plans. As a result, geography determines the availability and quality of medical coverage for the disabled.

The number of Medigap program choices, the price of insurance, and the qualifications for coverage vary significantly from state to state. For example, many states require disabled individuals to pay more than non-disabled participants. **Twenty-eight states deny the disabled Medigap coverage entirely.** Sadly, the variance in state standards leads to confusion and often to the mistreatment and exploitation of disabled Medicare recipients.

Congress has recently increased its oversight of state Medigap policies. For instance, Congress has already acknowledged that the 24-month waiting period after a disabled recipient becomes eligible for Medicare, but before medical treatments are subsidized, can be a death sentence for people with certain diseases. As a result, it has eliminated the 24-month waiting period for people with amyotrophic lateral sclerosis (ALS or Lou Gehrig's disease) and end-stage renal disease (ESRD). An act that would eliminate the 24-month waiting period for all disabling conditions was introduced in both the House of Representatives and the Senate in the fall of 2007, but the bill has yet to be voted on in either chamber.

To date, legislation to make Medigap coverage universal for all eligible recipients of Medicare has been introduced only in the House of Representatives. The bill, Medigap Access Improvement Act of 2007, co-sponsored by Representatives Philip English (R-PA) and Bobby Rush (D-IL), seeks to expand access to Medigap policies for all Americans with disabilities. However, the legislation does not equalize the purchase price of the Medigap policy for the disabled and the senior population.

Ultimately, the financial burden generated by healthcare costs for many disabled recipients is detrimental. According to a report published by the AARP Public Policy Institute, "Disabled beneficiaries are less likely to have ➤

Pokémon and CVID

By Mark T. Haggard

"Calvin," I clamored, "turn that thing down!"

I thought I was doing myself a favor when I bought my son a handheld video game for his birthday. I remember the cross-country drives my family endured when I was younger. I'm always amazed that my parents took my two brothers and me on a long vacation in a station wagon with only conversation and AM radio to amuse us. My brothers, however, often amused themselves by giving me repeated noogies and stingers. I'm sure my parents were thrilled when Mattel came out with the handheld Electronic Football, until, of course, their aural senses were barraged by the never-ending "bleep, bleep" of little red dots being tackled.

Today, my son entertains himself with Pokémon for Nintendo DS, and the dings and other noises from his game drive me batty.

"Hey," my son yells from the backseat, "I got a Chimpchar!"

"Oh geez, not again," I reply despondently. "Do I need to take you to Dr. Rusty?"

"It's on my game, Dad. He's a Pokémon."

"On your game?" I answered. "Thank God, I thought you were getting sick again." Admittedly, my knowledge of Pokémon is lacking. I first thought he was a Jamaican proctologist. Now I realize that it's the Japanese mega-franchise that started as a card game, moved to television, and is now in the world of video.

As I listen to the play-by-play of each game, I note how the names of the Pokémon resemble those of all the pathogens, antibiotics and brand names of immune replacements that surround my kids.

"Chimpchar, Dad. He's a chimp Pokémon. His attacks are Ember, Scratch and Growl."

"Ember?" I queried.

"Yeah, he throws little pieces of fire at his opponents." Now my imagination was piqued. "What else do you have?"

What Is Medigap and Does Your State Offer It? (continued)

employee-sponsored insurance coverage than older beneficiaries (19 percent to 33 percent); [they] are four times less likely to have individually purchased insurance to supplement Medicare than their older counterparts; and they are also twice as likely as older beneficiaries to have neither public nor private supplemental insurance." Participation in Medigap coverage programs at a rate comparable to low-risk patients could relieve their burden significantly.

In a speech about universal tolerance, President Jimmy Carter stated, *"The measure of a society is found in how they treat their weakest and most helpless citizens."* The current lack of uniform access to Medigap for disabled Americans denies them a benefit that is open to their non-disabled peers.

If we do less for people who need more, what does it say about our society? ■

States that provide Medigap insurance for the disabled:

California	Minnesota	Oklahoma
Connecticut	Missouri	Oregon
Kansas	Mississippi	Pennsylvania
Louisiana	New Hampshire	South Dakota
Maine	New Jersey	Texas
Maryland	New York	Washington
Massachusetts	North Carolina	Wisconsin
Michigan		

States that deny Medigap insurance to the disabled:

Alabama	Georgia	Nebraska	Tennessee
Alaska	Hawaii	Nevada	Utah
Arizona	Idaho	New Mexico	Vermont
Arkansas	Illinois	North Dakota	Virginia
Colorado	Iowa	Ohio	West Virginia
Delaware	Kentucky	South Carolina	Wyoming
Florida	Montana	Rhode Island	Indiana

"Well, Ghastly is a ghost-type Pokémon. His attacks are Confuse-Ray and Astonish. I also have Torterra. He's a grass-type Pokémon whose attacks are Crunch, Rock Smash, and Razor-Leaf."

"Torterra?" I repeated.

"Yeah, he's the evolution of Grotle."

Now, I'm not a doctor of immunology, nor do I profess to understand much about immune deficiencies. I have listened to doctors as they try to explain, in layman's terms, what is occurring inside my children.

Nevertheless, when my children come to me wondering about their repeated sicknesses, I have been speechless.

That was until I applied the language of Pokémon to immune deficiency. Now it is perfectly clear to all of us.

I came home from the pharmacy one afternoon with a bottle of antibiotic in a bag. "Tonight," I announced while taking the bottle from the bag, "inside my child's lymph nodes: Streptococcus vs. Amoxicillin!"

My wife looked at me with a furrowed brow.

"What's that?" my wide-eyed son asked.

"I'm talking on your level, Son. Imagine inside your body, two dudes show up doing a Pokémon battle. One guy says, 'Streptococcus, I choose you.' The other guy says 'Amoxicillin, I choose you.' The battle is on."

"What's Streptococcus?"

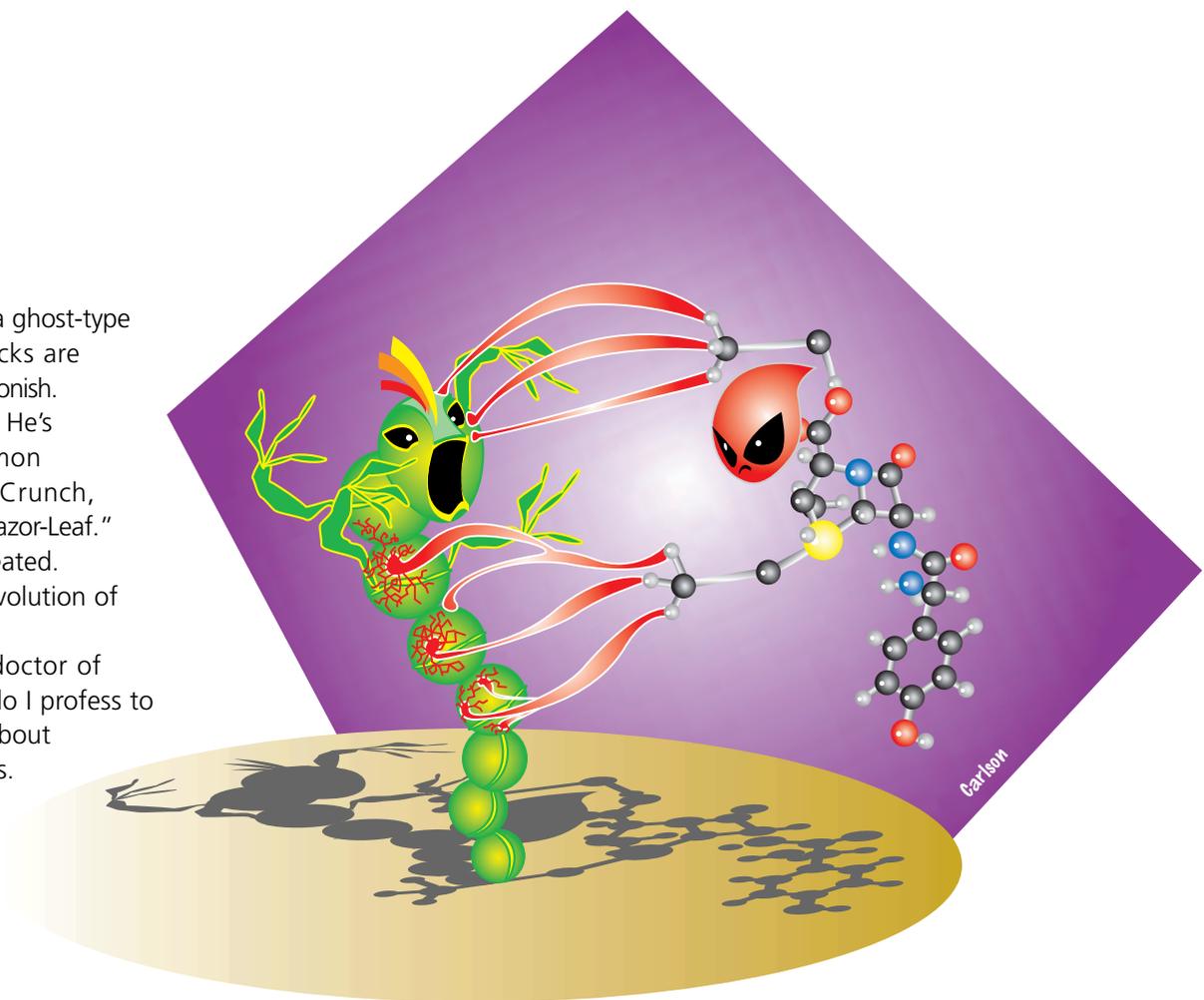
"Streptococcus is the evolution of 'Strepto-coccaceae.'"

"What are his attacks?"

"His attacks are Glandular Swelling and Sore Throat."

"So, what's Amoxicillin?"

"It's the evolution of Penicillin. Its attack is Bactricide. Amoxicillin binds itself to the cell wall of Streptococcus.



When the bacterium tries to expand, it can't, and it explodes! Isn't that cool?"

"Wow," my son replied somewhat downcast. "The guy on my game just faints."

"Your life isn't a game, Son."

On the next fourth Thursday of the month, when Nurse Nancy showed up to infuse my kids, my son assumed the role of Pokémon announcer: "This morning, inside my pancreas: Common Variable Immune Deficiency vs. Immune Globulin."

I smiled at my son. Nancy shot him a cockeyed look. My wife muttered something under her breath.

"Immune Globulin is the evolution of Plasma," my son continued.

Later that morning, I thought I heard the sound of Mattel's Electronic Football. I instinctively covered up, expecting noogies and stingers from my brothers. Then I remembered that they were 800 miles away and regained composure. "Did you score a touchdown?" I asked.

"No, it's my pump. This isn't a game, Dad. This is my life."

"And a good one it is." ■

SubQ at SubZero? No Problem!

By Jim Trageser

Carry on a conversation with Lauren and one of the first things you'll notice is that she's not one for excuses. This 29-year-old Boston native, who resides in Jonesborough, Northern Ireland, is an extreme mountain climber. Her two brothers, Neil and Jim, are veteran marathon runners, and all three have hypogammaglobulinemia, a condition in which the body doesn't produce enough of certain antibodies.



Neil, Jim and Lauren

Lauren and her brothers have been receiving replacement immune globulin treatment since their diagnoses as toddlers, now more than a quarter-century ago. Hypogammaglobulinemia is a disorder that is generally classified under the umbrella of common variable immune deficiency (CVID). The cause of hypogammaglobulinemia is believed to be genetic—and there is currently no cure. Those who have the condition are generally more susceptible to respiratory infections if it is not treated.

"We were initially all on intramuscular injections every two weeks until we were too big to receive an adequate dose this way," Lauren writes in an email interview. "At that stage, we were given intravenous gamma globulin."

Her two brothers still receive their infusions in this manner, but she receives subcutaneous (subQ) immune globulin therapy, the most common technique used in Northern Ireland.

When learning that their children had a permanent medical condition that would require lifelong treatment, Lauren's parents were taken aback.

"My parents obviously found the diagnosis difficult initially. Having three sick children with various injection schedules, etc., was not easy, but they both managed quite well," she continues. "We have all been under the care of an excellent immunologist at Children's Hospital in Boston, and he was very supportive of my family. His name is Dr. Raif Geha. His regular contact with my mother and father and open and encouraging attitude helped them immensely."

Despite their children's diagnoses and ongoing treatment, though, Lauren says her parents held them to the same expectations and standards they would have if the children had been born without ongoing medical issues.

"My parents never held us back from doing anything as kids. They were so supportive and encouraging. We were all very active in sports and other activities, and my parents made sure that we were never treated differently than other kids. This was good and bad! It meant that we never got to 'play' sick, either. ... They made sure we got to school no matter how bad we pretended to be."

All three were involved in sports growing up. Neil played football; the oldest, Jim, who ran cross-country and track, was captain of both in high school; and Lauren, who was a swimmer, also ran track and was a captain too.

After college, Neil and Jim began running marathons. Neil, who ran the Boston Marathon in April, has also completed the Chicago and NYC races. Jim has run 13 marathons, including eight consecutive Bostons. His best time—3:03—places him in the top 800 of 36,000 finishers in Chicago.

Keeping up with her brothers, Lauren continued to swim competitively, and swam in a two-mile lake race in Northern



Ireland two years in a row. But then she discovered her life's passion—extreme mountain climbing.

Six years ago, while working in Thailand as an English teacher, Lauren tried rock climbing. Never faint of heart, her next adventure was a 75-mile jungle trek on foot into Burma, followed by travel through Southeast Asia. Her expedition culminated in Tibet at the North Face Base Camp of Mount Everest, where she met her future husband, an Irish climber who reached the summit that season. Together, they traveled to Nepal, Northern Ireland, Scotland and Ecuador, building their life around hiking and mountaineering.

"My husband and I took my two brothers on their first ice-climbing adventure [in Scotland] and we all [reached the summit] one night after a 10-hour climb in the dark," Lauren tells us.

"Finally, I accompanied my husband on the Irish K2 expedition in 2005. I was the base camp manager for the trip and spent a month in northern Pakistan with the team."

While hypogammaglobulinemia is associated with respiratory problems, Lauren says she doesn't prepare any differently than other climbers.

"I try and stay in very good shape, which really helps, and other than that, I don't do anything different from anyone else."

Maintaining SCIG treatment on a climb might give others pause, but Lauren claims the only thing she had to do was sleep with the product in her sleeping bag so it wouldn't



Terence, Lauren and Conor

freeze. She successfully infused at K2 base camp, which is about 5,000 meters at an altitude of 16,404 feet, or just over three miles.

"Altitude sickness is a funny thing," she continues. "There is not a lot that you can do to tell who will be affected. Some of the climbs, I have been so strong and not affected by it while others have, and then on other climbs, I have been slow and suffered headaches."

Lately, the chance to even get headaches has been greatly reduced, however, due to a recent addition to Lauren's family—a baby boy named Conor.

Given that hypogammaglobulinemia is believed to be genetically indicated, Lauren has considered what her condition may mean for her child.

"We saw a genetics specialist here prior to his birth who advised us that it was highly unlikely that he would inherit the condition," Lauren writes. "In fact, she felt it was nearly impossible as my husband would have to have the same faulty gene and she did not see this as likely. My son will be tested next month, though, as a precautionary measure, but we are confident he is fine."

Not surprisingly, having a new baby has slowed Lauren's mountaineering far more than her condition ever did.

"At the moment, we do not have any other big climbs or adventures planned, but I am sure we will manage to get away next year on some type of trip!" ■



Lauren and her husband, Terence, after he proposed at the summit of Ben Nevis, Scotland

Editor's note: Since the article was written, Lauren has learned from initial tests that Conor is likely affected by hypogammaglobulinemia. More tests are scheduled.

Make 'Em Laugh

By Cheryl L. Haggard



"I have what?" My voice responds in fear as I almost drop my cell phone into my mocha.

"Severe degenerative scoliosis and nerve damage with two ruptured discs," Dr. Alex repeats.

He knows not to ask, "Are you sitting down?" as the

pure luxury of a couch inflicts indescribable pain. The only position my body will allow is either standing or lying down, neither convenient for a mom with three children, two of whom battle primary immune deficiency disease.

Many of you have come to expect me to be lighthearted when it comes to our community's not-so-funny circumstances living with chronic illness. The most recent question I've received is, "How are you going to make 'em laugh this month?"

Honestly folks, I've been asking myself this same question. Trust me, there's nothing funny about being diagnosed with scoliosis at age late 30-something.

Despite feeble attempts at wrapping my funny bone around pain, whether I'm getting tortured at physical therapy or finding myself at the receiving end of an IV "cocktail" of muscle relaxers and painkillers, I've discovered that I have much to be thankful for.

Yup, you read that right—thankful.

I've discovered that when you're going through hell, you might as well take along a few briquettes and a hibachi. In fact, if our government could harness the burning and electrical power that accompanies one of my muscle spasms, it would solve our nationwide energy crisis.

Pithiness aside, I'd like to share with you a few things I've learned to be thankful for:

- **A Female Massage Therapist**

Aware of alternative lifestyles, I write this with the utmost respect. However, being happily married for 17 years to a wonderful man, I was a bit uncomfortable when my husband, Mark, purchased a massage for me with Kathryn within minutes of my phone conversation with Dr. Alex. I've never experienced another woman "touch" my body other than my mom (that was a few years back when she saw me in the buff; I was probably receiving some "wisdom" in the buttocks region, if you know what I mean). But when I saw Kathryn for the first time days after my injury and in unbearable pain, I told her, "I've never been so happy to take my clothes off for another woman in all my life."

- **Pharmacy Frequent Flier Miles**

Two ruptured discs plus scoliosis equals indescribable pain, and with pain comes lots and lots of drugs. Drugs that have longer names than a spring earthworm emerging from its home in your front yard. Drugs that make you

laugh at your fifth-grader's morning "bed head" when his messy hair used to make you frustrated as all get-out. Drugs that also lead you to interesting conversations with the family's Saturday morning bacon as it sizzles and pops in the skillet. And there's an added bonus: Because I've been visiting my pharmacy daily to pick up a prescription since my injury, my name was entered multiple times into the pharmacy's drawing for a digital camera. Lucky me, I am now the proud owner of a beautiful camera.

- **'Roided-Up PIDD Kid**

My son, Caleb, who has an immune deficiency, recently had a bout of infections that caused him to break out in uncontrollable hives. At the height of his infection and hives, he was on two separate steroids to calm his angry immune system. Our average-sized 8-year-old morphed into Augustus Gloop. When you hugged Caleb, it was like wrapping your arms around a human marshmallow. One frozen Idaho morning, I slipped and fell on the sidewalk. My bloated boy just happened to be on my left, in line with the direction I toppled, and I landed on what felt like that cloud you see on cream cheese commercials. Yes, I'm finally grateful for prednisone side effects.

- **My Family, Friends and Just Plain Ol' Good Folks**

On Dec. 30, my life forever changed. I quickly went from caregiver to patient, from a mom and wife to a woman who appreciated my neighbor's elk stew. I also discovered that all the goofy and mushy emails from my friends just meant they were thinking about me. Most of all, I'm thankful for my family, especially my husband. Even though he's spent more time making "whoopee" to the vacuum cleaner, he's been my hero, taking on the duties of keeping a roof over our heads while making sure the kids have clean undies.

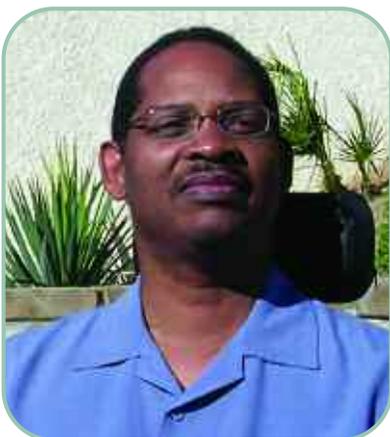
Yes, I think there can be humor amidst the pain of chronic illness. Though it's hard and much like finding that barbecue in hell, it sure makes that "feast" of a good belly laugh mean more to those of us who have earned it.

Even if we do look silly laughing at bacon frying, I'm just thankful for the laughs, wherever I find them. 🍷

Let's Talk!

By Shirley German Vulpe, EdD

If your life depends on immune globulin, this column is for you! It is an opportunity to network and share our experiences, because it behooves us to learn as much as possible about all of the ramifications of our illnesses. This column allows us to learn from one another. If you have a story you'd like to share, please email us at editor@igliving.com.



Ken Hobbs

For this column, I interviewed 53-year-old Ken Hobbs, who lives with two debilitating disorders. Ken suffers from chronic inflammatory demyelinating polyneuropathy (CIDP), an autoimmune disorder characterized by progressive weakness with impaired sensory dysfunction in the legs and arms. In addition, he also has adult onset nemaline myopathy, a rare neuromuscular disorder that causes weakness in the muscles of the face, neck, limbs and in the respiratory muscles.

Shirley: Ken, can you tell us about your illness and how it has impacted your life?

Ken: That is a long, complex situation.

Shirley: I can imagine. Why don't we start with how it began ...

Ken: I was working as a city manager in Northern California in 1999. The first problem I noticed was difficulty walking. It got progressively worse. The doctors ordered multiple tests. The first diagnosis was a spinal tumor followed by prostate cancer.

I had surgery in February and June 2000. Luckily, both tumors were benign. I recovered well, then I hit a wall, literally and figuratively: I couldn't control my walking. Within 30 days I lost 60 pounds and could not lift a glass. Local MDs did many tests, but were unable to establish what was wrong. They suggested I go to the Mayo Clinic. I took a leave of absence from work. My wife and I were there seven weeks. They said I had nemaline myopathy. They got me a wheelchair. They told me there was no cure and no treatment. I was to go home and enjoy what I could of my remaining life. My wife and I tried to stay focused on life. I attempted to go back to work. I was told they did not want someone in a wheelchair in my position. I was let go in spite of begging to stay. I have since received a settlement.

Shirley: How difficult for you and your family. What happened next?

Ken: A miracle. We moved to Southern California for my health. Within a week, a neighbor who noticed I was in a wheelchair and dependent on my wife said his brother was director of neurology at Cedars-Sinai hospital in Los Angeles. He offered to look at my Mayo Clinic records. We agreed. He asked if he could share my file with Dr. King Engel, one of the people who discovered nemaline myopathy. After multiple tests, they concluded that I did indeed have nemaline myopathy, but also CIDP, and that I could be helped with

IVIG. I started it the next month, in December of 2002.

Shirley: How do you receive the IVIG?

Ken: Through a Mediport in my chest. I infuse a total of 85 grams, two times a week.

Shirley: Has this helped your condition?

Ken: Yes. After six months, there was an absolute change. First my energy level, legs and trunk improved. After one year, I was able to dress and feed myself. In a year and a half, I was able to transfer. Now I drive an adapted van. It has a ramp and hand controls. And I have energy to volunteer. I advocate for persons on IVIG for reasonable reimbursement by letter writing and visiting government officials. I helped start a Coachella Valley Neuropathy Association. I am also a volunteer pastor.

Shirley: Have you had any problems receiving or paying for the IVIG?

Ken: Another long story. The short version is yes—many on both accounts. [Because of] the reimbursement difficulties caused by Medicare decreasing reimbursement for IVIG, several places stopped infusing IVIG. I now do it at home.

Shirley: Do you have any final message for our readers?

Ken: Yes. I'm happy that as an able-bodied person I let my wife know how much I appreciate and love her. For example I brought her flowers each payday. I had never expected that I would become fully dependent on her. I also realize

that I'll do whatever it takes to be a positive influence for those in my life. That's why I advocate and volunteer. My wife and I have also written a chronicle of this experience to share with others called "No Cure, No Surrender." ■

For More Information

Nemaline Myopathy

Nemaline myopathy is one of the diseases the Muscular Dystrophy Association addresses. Visit www.mdausa.org/disease/nm.html

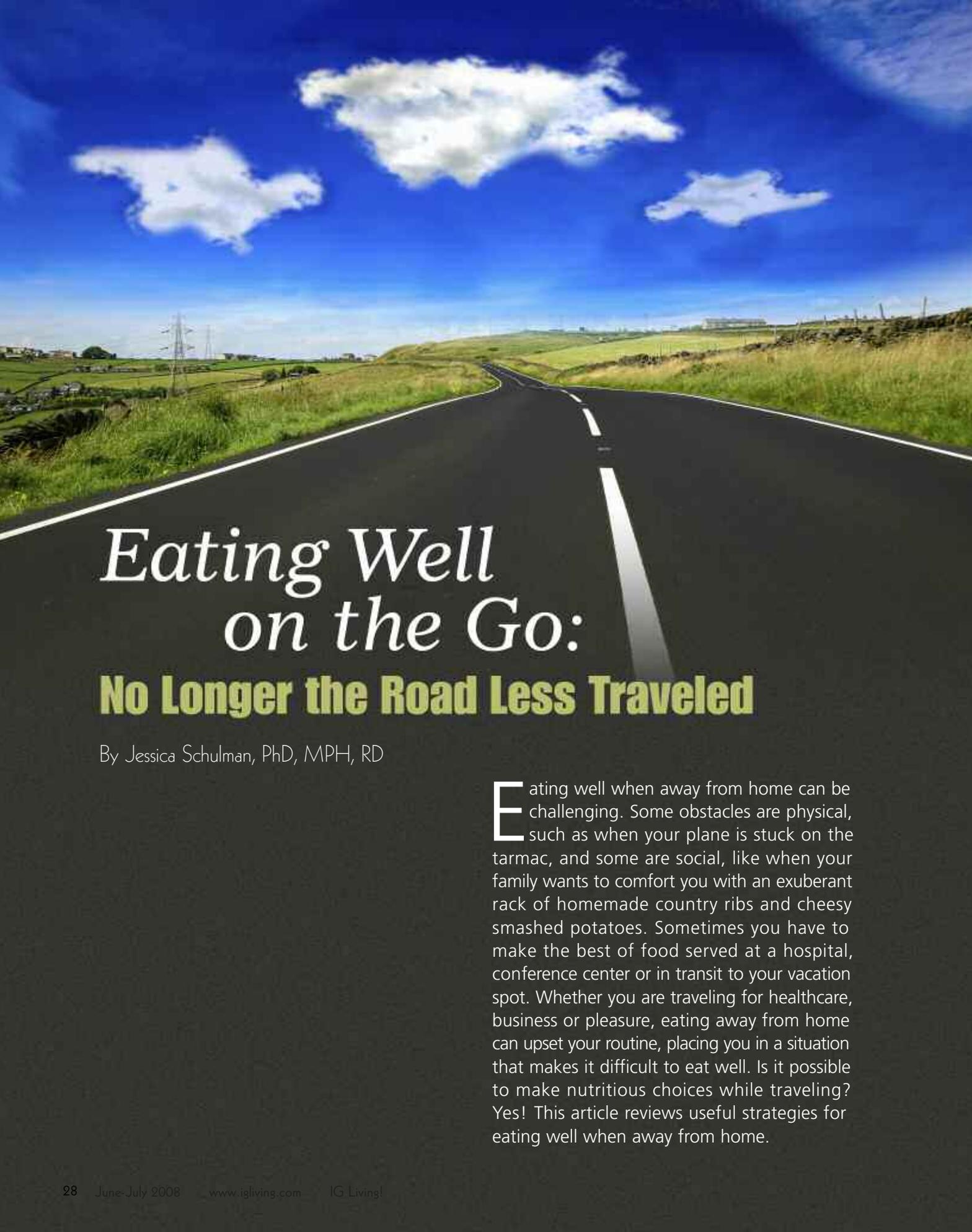
The National Organization for Rare Disorders has other good links at www.rarediseases.org/search/rd_bdetail_abstract.html?disname=Nemaline%20Myopathy

CIDP

National Institute of Neurological Disorders and Stroke www.ninds.nih.gov/disorders/cidp/cidp.htm

GBS/CIDP Foundation International
info@gbsfi.com

American Autoimmune Related Diseases Association
aarda@aarda.org



Eating Well on the Go: **No Longer the Road Less Traveled**

By Jessica Schulman, PhD, MPH, RD

Eating well when away from home can be challenging. Some obstacles are physical, such as when your plane is stuck on the tarmac, and some are social, like when your family wants to comfort you with an exuberant rack of homemade country ribs and cheesy smashed potatoes. Sometimes you have to make the best of food served at a hospital, conference center or in transit to your vacation spot. Whether you are traveling for healthcare, business or pleasure, eating away from home can upset your routine, placing you in a situation that makes it difficult to eat well. Is it possible to make nutritious choices while traveling? Yes! This article reviews useful strategies for eating well when away from home.

Planning Ahead

Americans spend more today than ever on foods eaten away from home. From 1992 to 2002, the total amount spent jumped from \$263 billion to \$415 billion, a 58% increase.¹ In recent years, the public has demanded healthier choices when eating out, and the food service industry has responded by offering a wider variety of menu options. As a result, it is possible to eat nutritious meals while traveling—but it does require that you plan ahead. Before you go, and not after you arrive, is the best time to work out reasonable solutions to the potential challenges of dining out. Answering the following questions can help you prepare to make choices that are consistent with your personal diet plan.

1. What are your nutrition goals or plan?
2. What are examples of foods that are often available and not consistent with your dietary plan?
3. What are examples of foods that are better choices and might actually be available on your travels?
4. What action will you take to ensure that healthy meals are accessible on your travels?

How to Eat Out Well

When you call ahead to make hotel reservations, ask the concierge or receptionist to suggest food service establishments. You can request the restaurant's phone number or website, or even ask that a copy of the menu be sent to you. When in doubt, choose restaurants that have vegetarian options—they are more likely to offer healthy fare and make other food accommodations.

When you arrive at the restaurant, do not hesitate to ask for a special order. Cooks in the United States are accustomed to honoring requests for dressings, sauces or butters on the side, and some will make more complicated substitutions. Remember to be courteous to waitstaff so they will be more considerate about accommodating your needs. Consider starting with something like, "I know that you are busy, but may I ask you some questions about the menu?" The following table provides ideas for restaurant substitutions based on the Dietary Guidelines for Americans (the guidelines do not apply to young children). Always check with your healthcare provider to see what diet is best for you. ➤

Instead of:	Ask for:
Fried Sautéed in butter Stir-fried Anything with or in butter (or hollandaise, béarnaise) Basted	Grilled Broiled Steamed Poached Baked Roasted
Au Gratin Escalloped	Lightly sautéed Lightly stir-fried
White bread Breakfast pastries	Whole grain or whole wheat breads, bagels, cereals
Continental breakfast	Fruit Yogurt Oatmeal Whole grain breads Cereals
Beef	Fish Shrimp Lobster Poultry
<i>(continued next page)</i>	



¹ Stewart H, Blisard N, Bhuyan S, Nayga R Jr., The demand for food away from home: Full service or fast food? *Agricultural Economic Report No.* 829. p.1, Jan. 2004.

Instead of:	Ask for:
Fatty cuts of meat Rib-eye Porterhouse T-bone	Leaner cuts of meat London broil Filet mignon Round or flank steak Sirloin tip Tenderloin
Pre-tossed salad	Dressing on the side Flavored vinegars
Unspecified sandwiches, wraps, burritos, etc.	Dressings or nutrient-poor toppings to be used sparingly
Whole milk (cold or in hot drinks)	Fat-free / low-fat milk
Mayonnaise Butter Margarine Sour cream Sauces (cheese, tartar, etc.) Gravy	Ketchup Mustard Low-fat ranch dressing Yogurt Balsamic vinegar Salsa Cocktail sauce Grated Parmesan cheese (1 tsp)
Soda Fruit drinks or punch	Bottled water with lemon Dilute highly sweetened drinks (1 part water: 1 part drink) Unsweetened tea Flavored waters Broth
Creamed soup	
Salt Mixed unspecified seasonings	Lemon Roasted, fresh or powdered garlic Herbs such as dill, tarragon, cilantro and basil
Cheesecake, ice cream, chocolate chip cookies	Plain flavors (not super premium brands with toppings) Hot milk with a shot of syrup or chocolate cocoa flakes Angel food cake Fruit Biscotti Unspecified: control the portion by sharing dessert

More tips for eating standard or international fare are located at:
www.americanheart.org/presenter.jhtml?identifier=531

Fast Food and Fast Casual

According to the American Public Health Association, consumers are unaware of the high levels of calories, saturated fat, and sodium in fast-food meals.² Additionally, items on fast-food menus are often low in nutritious content such as essential fatty acids, fiber, vitamins and minerals. What are better choices when eating fast or convenience food? Aim for grilled chicken sandwiches, premium salads with low-fat dressing, or even a small (2-ounce) burger with lettuce and tomato. Instead of fries, a baked potato with low-fat dressing or ketchup is a good choice. Grilled vegetarian sandwiches, chicken fajitas and black bean burritos are other decent options, whereas fried fish sandwiches will derail any balanced nutrition plan. Dietary information is available to consumers at most fast-food establishments or online. Here is a selection of websites that list menu items with calories, fat and sodium content that you can print.

Fast food nutrition websites

Baja Fresh®

www.bajafresh.com/jump.jsp?itemType=CATEGORY&itemID=68&iMainCat=4&iSubCat=10&i3Cat=68

Carl's Junior®

www.carlsjr.com/content_svn/downloads/nutrition.pdf

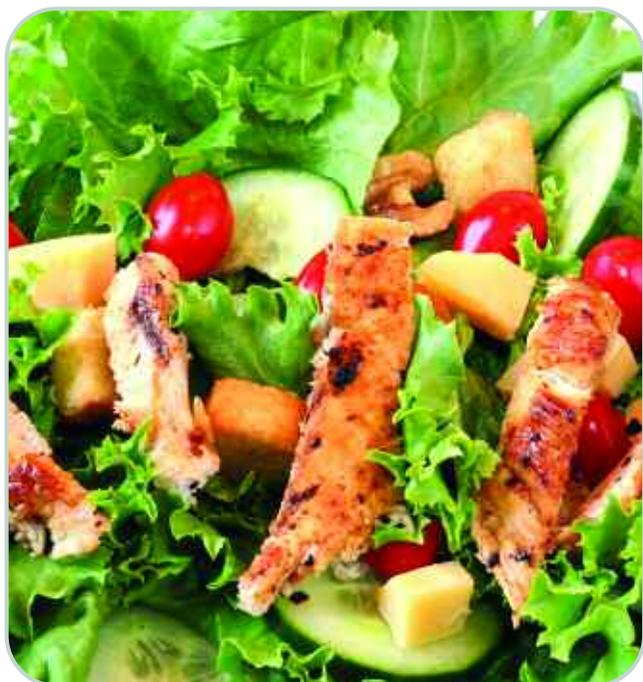
McDonald's®

app.mcdonalds.com/countries/usa/food/nutrition/categories/nutritionfacts.pdf

Taco Bell®

www.yum.com/nutrition/menu.asp?brandID_Abbr=5_TB

² Burton S, Creyer EH, Kees J, Huggins K. Attacking the obesity epidemic: The potential health benefits of providing nutrition information in restaurants. *Am J Public Health* 2006;96:1669-1675.



Traditional sit-down restaurants have made it easier to take hot food to your home, hotel, or hospital room. These “fast casual” places include California Pizza Kitchen, Islands, The Cheesecake Factory, and many others. They allow you to call in your order, and food is brought to your car with curbside to-go service. Though they may offer more healthy choices, along with the traditional ones, be aware that the calories, fat, sodium and serving sizes tend to be larger (even gargantuan) than what is served at home. Nutrition experts Lisa Young, PhD, RD, and Marion Nestle, PhD, MPH, determined that restaurant portions are at least two times—and sometimes eight times—larger than standard serving sizes.³ One slice of Godiva chocolate cheesecake from Cheesecake Factory, for example, contains more than 900 calories, while an actual serving of homemade cheesecake contains about 450 calories.

Budgeting on the Fly

If you think you might have trouble sitting down for a meal, try to at least eat a snack. This way you will not be ravenous when a meal is available. The worst time to walk into a restaurant, or peruse the prepared food aisles, is when you are hungry. Variations in blood sugar, especially hypoglycemia, can affect judgment and make practicing moderation difficult.

Balancing food over the course of a day, or budgeting items that are not part of your plan, does not require precise calorie counting—unless you must follow a rigid diet. The idea is to informally assess what you have consumed and to balance that with what your intake ought to be over the course of the day. Maintaining a healthy weight requires you to balance your “energy in” and “energy out” over the long run. Refer to the Dietary Guidelines for Americans, 2005, for recommendations on food and physical activity choices for health. At MyPyramid (www.mypyramid.gov), you can determine your appropriate energy intake and number of servings for each food category. Because MyPyramid is not a therapeutic diet for any specific health problem, individuals living with medical conditions should ask their physicians about the diet that is right for them.

Convenience Foods and Healthy Snacks

Convenience foods can be packed with your luggage and may come in handy. Items such as instant oatmeal and dehydrated soups (low-fat, low-sodium varieties) can be prepared in a hotel room with hot water from a coffeemaker or water that has been microwaved. Fresh fruit that you can peel, whole grain crackers and trail mix can help you get to the next healthy meal. When traveling by plane, you can pick up essential food items before boarding. Many airlines have cut back on food service options, but most will still provide special meals on request. Be sure to notify the carrier more than 24 hours before your flight. (A low-fat meal will typically offer more balanced and higher-quality protein sources than a “regular” in-flight meal.)

Once you arrive at your destination, you can pick up plenty of items locally to keep in your room. Most hotels can provide a small fridge and microwave, but always check before making reservations. These are a few convenient favorites:

- Whole wheat bread, bagels, crackers, cereals
- Mozzarella cheese sticks
- Fresh fruit (banana, oranges, avocado)
- Juice boxes, Gatorade®, bottled water
- Dried fruits (raisins, apricots, mango, etc.)
- Dehydrated soups
- Nuts and trail mix
- Yogurt
- Hummus by Sabra® ➤

³ Young LR, Nestle M. Expanding portion sizes in the U.S. marketplace: Implications for nutrition counseling, *J Am Diet Assoc.* 2003 Feb;103(2):231-234.

- 100% whole grain Fig Newtons®
- Frozen meals (if a freezer is available)
 - Healthy Choice®, Flavor Adventures
 - Kashi®
 - Amy’s®
- Energy bars with low sugar and moderate protein

Airport Dining

The Physicians Committee for Responsible Medicine (PCRM) surveyed access to healthy food choices in airport terminals across the U.S. It found that more than 80 percent

of restaurants at the 15 busiest U.S. airports surveyed offered at least one breakfast, lunch or dinner entrée that followed the Dietary Guidelines for Americans (e.g., low fat, cholesterol-free, high in fiber).⁴ Following these guidelines has been shown to help manage weight as well as reduce the risk of heart disease; it also may be useful in treating diabetes. In PCRM’s most recent report, airports were ranked based on the percent of restaurants that offered healthy meals. PCRM nutrition experts provide examples of healthful food offerings from each. Airports with the most healthy options are listed first.

Findings from PCRM’s 2007 report about healthful airport food options	
Dallas/Fort Worth International Airport	Veggie noodle bowls and veggie sushi at Blue Bamboo Xpress, the spinach enchiladas at Cantina Laredo, and the black bean burgers at Bennigan’s. Wendy’s, where low-fat choices include baked potatoes, spring mix salads (minus the cheese), and veggie burgers.
Chicago O’Hare International Airport	Eat a Leaf’s roasted vegetable sandwich; Ciao Gourmet Market’s walnut and fruit salad with mixed greens, cranberries, strawberries, mandarin oranges and jicama; and Fox Skybox’s portobello and red pepper brioche with basil and pine nuts (to make this dish low-fat, ask the server to hold the cheese).
Detroit Metropolitan Wayne County Airport	Tailpipe Tap’s Motown veggie sandwich, Max & Erma’s grilled vegetable sandwich on ciabatta bread with tomato-basil spread (sans cheese), and the veggie stir-fry and almond rice salad at Mediterranean Grill.
Los Angeles International Airport	Vegetable fajitas at El Paseo, the vegetable sandwiches (minus the high-fat cheese) at Creative Croissants, and the roasted vegetable wraps at Eaterna.
Newark Liberty International Airport	Mixed vegetable and tofu stir-fry at Asian Chao, the custom-made salads and wraps at GreenLeaf’s Grille, and the veggie burger (without cheese or mayo) at Dick Clark’s Restaurant.
Denver International Airport	Made-to-order salads at Chef Jimmy’s Bistro and Spirits, the beans and rice at Cantina Grill, and the portobello mushroom sandwiches and garden burgers at the various Lefty’s Grille establishments.
Phoenix Sky Harbor International Airport	Oaxaca’s vegetable burritos and taco salad, Yoshi’s Asian Grill’s avocado cucumber sushi and edamame, and Roadhouse 66 Bar’s veggie burgers and veggie pizza.
Minneapolis-St. Paul International Airport	Caribou Coffee, with its grilled portobello mushroom wraps, black bean and tofu salads, and tabouli salads; and French Meadow Bakery & Café, which serves vegan chili, tofu salad and various soups and sandwiches.
San Francisco International Airport	Deli-up Café, which serves Middle Eastern platters; Ebisu, with its udon noodles in veggie broth and veggie sushi; and Max’s Eatz, which serves a roasted veggie sandwich.
John F. Kennedy International Airport	Buddha’s delight and broccoli with garlic sauce at Sky Asian Bistro, vegetable dumplings and grilled vegetable sandwiches at Soup & Kimbob, and vegetarian wraps at the various Cibo Express eateries.

⁴ Physicians Committee for Responsible Medicine. Airport food still healthful: More than 80 percent of restaurants offer low-fat vegetarian options, but some cities lag behind, Winter, 2007. Available at: www.pcrm.org/health/reports/Airport_Food_Review_07.html

Las Vegas McCarran International Airport	Prickly Pear Café, for its vegetable sandwiches; Don Alejandro’s Texan Grill, for its vegetable burritos (hold the cheese); and Sbarro, which offers steamed veggies as well as spaghetti with marinara sauce and salad.
Orlando International Airport	Thai peanut noodles and vegetable dumplings at Zyng. Travelers can create their own pasta at Macaroni Grill, which will leave out the cheese and substitute broccoli or spinach for the meat in any of its dishes. And Miami Subs offers veggie pitas as well as made-to-order vegetable sandwiches.
Hartsfield-Jackson Atlanta International Airport	Vegan burritos at Qdoba Mexican Grill and the veggie wraps (minus the cheese) at Great Wraps. Also notable are the black bean patties at Chili’s Bar & Bites and the vegetable plate with black-eyed peas, green beans, cabbage, yams, corn and rice at Paschal’s Southern Delights.
Miami International Airport	California Pizza Kitchen’s smashed pea and barley soup, Casa Bacardi’s black bean and crispy rice salad, Jose Cuervo Tequileria offers jicama salad, and Café La Carreta features a buffet bar packed with salad, black beans, vegetables and rice.
Reagan National Airport	Matsutake Sushi’s vegetable combination roll, Cosi’s veggie chili, DC Samuel Adams’ vegetable sandwich (minus the cheese), and T.G.I. Friday’s portobello melt (also sans cheese).

Can You Bring Food on Planes?

Though it has become much more difficult for passengers to bring food aboard planes, it is possible to carry on some small servings of nutritious foods as long as the rules of the Transportation Security Administration (TSA) are honored (www.tsa.gov/311/index.shtm). Formula, breast milk or juice will be inspected at the checkpoint, but containers larger than 3 ounces (and in reasonable quantities for the duration of your trip) should be permitted. (Breast milk is considered in the same category as liquid medications.) According to the TSA website, “You are allowed to bring gel or liquid-filled teething rings, canned, jarred or processed baby food in your carry-on baggage and aboard your plane.” For those with special dietary needs, liquids such as water, juice, nutrition supplements and frozen gel-consistency foods may be carried onboard. If they are in volumes larger than three ounces each, they may not be placed in the quart-size bag and instead must be declared to the TSA Security Officer. A separate, insulated bag may be useful for keeping the foods separate from other property submitted for X-ray screening. In addition, properly packed items may be placed in your luggage and checked at your airline counter. Always contact the airline in advance because these rules are subject to change without notice.

Drinks purchased in the secure boarding area can be brought onboard an aircraft. However, given the hectic environment at the airport, the cost of convenience items, and the likelihood that the only brand of juice

your child will drink will not be available, it is important to plan ahead. Stay well-hydrated the day before your flight and set aside important snack items for your trip. In addition, there may be delays and longer wait times at the airport, so if possible try to eat a nourishing meal in advance. For example, the morning of your trip, a peanut butter sandwich or scrambled egg may be a better choice than a bowl of corn flakes. Because many airlines have scaled back food services, a preflight meal that has adequate protein will help satiate you until you arrive at your destination and can scout out higher-quality food establishments.

Summer Camps and Retreats

Most summer camps prohibit families from sending food with their children. According to Roy Oken, CEO and president of River Way Ranch Camp, children should never be sent to camp with food that is not authorized because of the risk of life-threatening food allergies to other campers, food-borne illnesses, and insects. However, special accommodations can usually be made if the child is on a special diet. With a secure fridge and microwave, just about any food can be prepared at camp.

Food and treatment needs must be taken seriously away from home. Oken explains: “If a child lives with diabetes or they have a milk allergy, we have the camp staff—aka camp ‘moms’—talk with the parents and make certain that the counselor is aware of what foods are on the diet plan and what foods are not.” Oken says that in special ➤

cases foods may be kept in the infirmary and “the camper can go there up to four times a day to get what they need.” If the child goes on an outside adventure, or an “outcamp,” the camp director is notified by the nurses to take any special food items along.

Camps or retreats for children or adults must support them in meeting their nutrition needs. Dietary modifications can be complicated, so never assume that the cooks or counselors know as much as your doctor or dietitian. Contact the camp director or nursing staff to find out how you or your child’s health needs will be met. Explain that nutrition is part of the treatment plan so that your child is comfortable and safe during his or her stay. Nancy Oken, executive director of River Way Ranch Camp, explains: “Everything humanly possible will be done at camp to guarantee the health, safety and happiness of your children.”

Eating Away on an Extended Healthcare Stay

Eating a balanced diet is an integral part of a patient’s treatment, recovery and well-being. For this reason, food quality and choices in medical centers have improved. Still,

Seaworthy Dining

Florence (Flo) Schulman, a retired professor of leisure management at Pierce College (and the author’s stepmom), encourages balance and moderation while sailing the Seven Seas. She explains that travelers can offset an unhealthy meal by eating well throughout the rest of the day. When Flo is on a cruise, she eats “healthy for breakfast, like Egg Beaters, oatmeal and fresh orange segments. For lunch, we try to avoid the main restaurant and stick to the salad bar. In the evening, we can really enjoy ourselves, celebrate with our friends, and choose a dessert.”

Still, she tries to make healthy choices in the evening and says, “My husband will order broiled fish almost every night, so that’s even healthier than back at home.”

She also recommends ordering sauces and dressings on the side so an entrée is not “smothered in empty calories.” She admits that it is not a perfect solution and points out that “when I have overdone it at a meal, I try to go easy the next day.” It is hard to have an ideal diet while on vacation, but Flo is realistic and admits, “We often gain a bit, about one pound a week is really what happens, but for others it is so much more than that.”

for many, the choices available in healthcare settings are not as appetizing as one would hope. Alternatives include 24-hour catering services and friends and family bringing in food to encourage proper intake. (Always make sure that foods brought to the patient are consistent with the diet, or “NPO” status, per doctor’s orders.) Nurses often lack the time to pay attention to their patients’ needs for assistance and socialization when eating. Often, caregivers end up helping out and become overwhelmed, thus neglecting their own needs for proper nutrition.

Places such as corporate housing, extended stay hotels, Ronald McDonald Houses and The Children’s Inn at NIH (National Institutes of Health) provide families with kitchen appliances or a stocked pantry. (See *Home Away From Home*, Page 9.) This is very helpful and enables families under stress to eat well. However, when you can hardly leave your loved one’s bedside, there may not be time to shop, cook and clean up.

If you are staying away from home for long stretches of time and dining out or cooking is not feasible, you can request a caregiver tray for a nominal fee at most hospitals. As an alternative, having frozen or dehydrated convenience items on hand may help. With a microwave and refrigerator, almost any balanced meal can be prepared. There are also plenty of Internet food vendors and full-service, personalized grocery shopping companies (e.g., wegoshop.com, peapod.com, parkeastkosher.com) that will deliver items directly to you. These services, or even shipping food from home, may be helpful for finicky children or individuals on restrictive diets.

With enough notice, one can prepare meals, freeze them, and then give a friend instructions on how to ship them to your location. It is essential that items are frozen and packed properly when food is shipped. Use foam or heavy corrugated cardboard and have it delivered overnight—even for smoked or cured items. Pack with frozen gel packs or dry ice. Before purchasing the dry ice, contact the postal carrier for instructions about how to pack it safely and how much you need (e.g., about two pounds of frozen items requires eight pounds of dry ice for overnight delivery). Label the outer package with “Keep Refrigerated” to alert the recipient. Make sure that perishable foods have remained frozen and are held at temperatures below 40 degrees F. As always, when in doubt, throw it out! ■

Editor’s note: This article is intended for general informational purposes only. Individuals with medical conditions—or those seeking nutritional advice—should consult their doctors to determine the diet that is appropriate for them.

ASK KRIS



Kris McFalls has two adult sons with chronic diseases treated with IG and is also receiving IG therapy. Formerly a physical therapist assistant, Kris is IG Living's full-time patient advocate, and she is eager to find answers to your questions. Email them to editor@igliving.com. Your confidential information will not be used for any purpose but communicating with you about your questions.

Ron: My mother is in the early stages of Alzheimer's disease (AD). No current medicines are helping her. After going through the literature, IVIG treatment seems promising and could be given off-label. I consider this a humanitarian treatment. (She may go into a clinical trial for something else, so we're not interested in a clinical trial.) The problem is, how do we find a neurologist who will prescribe IVIG off-label?

Kris: Of course, Medicare will not cover IVIG for Alzheimer's treatment, so your mother's treatment, if prescribed, would have to be on a cash-pay basis. Dr. Richard Schiff responded to your question. He is the global medical director for Baxter Bioscience, and Baxter is the sponsor of a clinical trial testing the efficacy of IVIG for Alzheimer's (see *IG Living* April-May 2008, 'Hope Is In the Blood').

Dr. Schiff: I recommend having the patient's doctor (the one who diagnosed and treated the AD) contact Dr. Norman Relkin at Cornell. He can better direct them as to whether the person would be a good candidate for treatment, the possible risks and possible ways to get treated. They can reach him through the Memory Disorders Program at New York Presbyterian Hospital-Weill Cornell Medical Center.

Mary: Have you ever heard of a doctor prescribing Imuran for the treatment of chronic inflammatory demyelinating polyneuropathy (CIDP)? My neurologist wants to take me off IVIG (which is working for me)

and put me on Imuran because "IVIG is for people who really need it." It is a scary thought to go off my IVIG when it is working. She also wants to perform nerve conduction tests again to see if they are still slowed. If the IVIG is doing its job, do you think these tests would be normal now? Do I need to go through the pain in obtaining these tests again?

Kris: Dr. Todd Levine, co-director of the Peripheral Neuropathy Clinic at Banner Good Samaritan Medical Center in Phoenix, Ariz., responded to your questions, but please continue discussing your treatment concerns with your doctor, Mary. It helps to write down questions as you think of them, and then go through your list at your next visit with your treating physician. And, remember, it is OK to ask for a second opinion.

Dr. Levine: There are many drugs that we use to treat CIDP. Steroids, despite their side effects, are probably the most effective. Because many neurologists do not like to subject their patients to the side effects of IVIG, they use "steroid-sparing" agents. These drugs include Imuran, Cellcept, cyclosporine and methotrexate. Therefore it is very reasonable to treat a patient with IVIG and then once they are better, find one of the above drugs that can treat the disease, and then we can get them off of the IVIG. The biggest issue is that these steroid-sparing drugs often take three to six months to begin working, so for a while, you may need to be on both the IVIG and the steroid-sparing drugs. ➤

Checking the nerve conduction studies from time to time can be helpful. Unfortunately, sometimes the clinical response and the electrophysiological response do not go hand in hand.

Dawn, a mom of a pediatric CIDP patient, sent the following questions:

1. How does IVIG actually work in the body for CIDP?
2. Is there any harm in extending the treatments to every six weeks?
3. How many loading doses are considered normal before moving to a maintenance dose?
4. Is it true that IVIG only sustains the level of function?
5. Is it necessary to use immune suppressant drugs in combination with IVIG? If so, why?

Kris: These are some great questions! Dr. Scott Carlson, a neurologist at Rockwood Clinic in Spokane, Wash., has a number of CIDP patients being treated with IVIG.

Dr. Carlson: Concerning the questions about treatment of childhood CIDP:

1. The mechanism of IVIG is very complex and somewhat controversial at this point. There may be as many as eight to 10 different ways that IVIG can modify the immune response. The bottom line is it modifies the immune response so that tissue injury and inflammation (in this case nerve injury) are diminished.
2. There is no harm in extending treatment intervals to every six weeks. In fact, it is often a goal to extend the IVIG interval out as much as possible to the point that the patient is stable between treatments. This interval varies from patient to patient.
3. Usually a single "loading dose sequence" or initiation dose is given, then maintenance doses are started at an interval of every two to four weeks until the patient proves stable. Then, as addressed above, continuing to extend the interval is attempted, if tolerated.
4. IVIG limits tissue injury in an autoimmune disease by modifying the immune system. Level of function can improve during the treatment course depending on

nerve and myelin healing and regrowth. Everyone's ultimate level of function will be different, depending on how severe the nerve injury was when treatment was started and how easily the immune injury is halted or modified.

5. It is not necessary to use immune suppressant drugs with IVIG treatment. The choice to add immune suppressant drugs (such as prednisone, azathioprine, Cytoxan) is dependent on each patient's response to IVIG, IVIG side effects, co-morbid diseases, ability for financial coverage of treatments, etc. IVIG can be a single treatment approach for some patients, particularly in childhood CIDP.

Kris: With reduced production of Talecris' Gamunex, many patients are asking what to do. We asked Dr. Mel Berger, Director of Allergy-Immunology at Rainbow Babies & Children's Hospital in Cleveland, what he recommends for patients who now have to switch products.

Dr. Berger: When starting infusion therapy with a different IVIG product, patients should start at a low infusion rate and allow longer intervals between rate changes, and/or they should premedicate the first time they use a different product. The treating doctor should review the choice of available products, and/or the infusion provider must ask the doctor if the proposed product is acceptable for that patient. Gamunex is free from sugar, so patients at risk from sucrose or maltose should be sure they are not switched to products containing them. Patients who tolerate the IgA in Gamunex should not have problems with the IgA in other products.

Rachel: Can subcutaneous immune globulin (SCIG) be used to treat dermatomyositis?

Kris: Dr. Levine answered your question about SCIG for dermatomyositis below. If this is something you want to try, it is important that you have a conversation with your physician. Let her or him know about the problems you have had with IVIG in the past and why you would like to try SCIG.

Dr. Levine: At present, there are no published trials on using SubQ immune globulin for dermatomyositis. There is good reason to believe that it would work as well as IVIG treatment. The only issue is whether you would be able to receive the same monthly dose in a SubQ fashion as you would in an IV fashion. But I would think that you could try it for a few months and see how you do.

Nanette: I have recently been diagnosed with chronic inflammatory demyelinating polyneuropathy (CIDP) after 19 years of living with a slow, ascending peripheral neuropathy of both legs. Chronic Lyme disease was suspected after 10 years without a diagnosis, testing positive many times, but now it has been ruled out and my current diagnosis is CIDP. Could Lyme disease still be the actual culprit?

Kris: Dr. Levine answered your question, but, as always, if you have concerns about any of your treatments, make sure to write them down and bring them to your next doctor appointment.

Dr. Levine: The connection between Lyme disease and neuropathy is still poorly understood. In fact, Lyme disease as a whole is poorly understood. As a neurologist who deals with a lot of neuropathy patients, I take the following approach: For Lyme disease to affect the nerves, the spirochete (microscopic bacteria) must have gained entry into the nervous system. If this occurs, you can find evidence of it in the spinal fluid. So I recommend a spinal tap for any patient who has had Lyme disease and develops a neuropathy or who worries that he or she might have Lyme disease causing their neuropathy. In CIDP, there will be no, or very few, cells in the spinal fluid. In Lyme disease, the spinal fluid will look like it is infected and there will be many cells in the fluid.

Connie: I have three boys, one with common variable immune deficiency (CVID), who is on SCIG. Before my son

with CVID started treatment, he and I both had chickenpox twice. My youngest son has not had chickenpox, but there has been an outbreak at his middle school. I'm thinking of having him vaccinated, but is there any risk to my son with CVID if my youngest son has the vaccine?

Kris: I posed Connie's question to Terry Harville, MD, PhD, medical director of three laboratories at the Departments of Pathology and Laboratory Services and Pediatrics at the University of Arkansas for Medical Sciences, in Little Rock.

Dr. Harville: Different physicians have differing opinions about exposure to varicella (the chickenpox virus) and vaccination.

Varicella is neutralized by antibodies so that an infection cannot become established in someone with sufficient antibody levels. People who cannot produce adequate levels of antibodies and who are dependent on replacement therapy, IVIG or SCIG, may have all the protective antibodies absent from their system during the initial exposure and onset of infection. Therefore, additional replacement with IVIG or SCIG is required. Further, acyclovir or Valtrex is required to help treat the infection, and such therapy is typically needed for at least a month.

Varicella vaccine is a live viral vaccine and is shed by the person who has been vaccinated, potentially placing immunodeficient or immunocompromised patients at risk for catching the infection. Shedding may occur for as long as a month or potentially even longer. Therefore, we have recommended that a vaccinated person who lives with an immunodeficient or immunocompromised person remain out of contact with that person for at least a month. If this is impractical, we have given additional IVIG or SCIG and acyclovir or Valtrex for a month.

In Connie's situation, it may be important to check varicella titers in the younger sibling first. If the titers are protective, vaccination is not required. If the titers are not protective, then care should be used with the vaccination, either through physical separation or increased IVIG or SCIG and acyclovir or Valtrex. ■

Myositis Mysteries

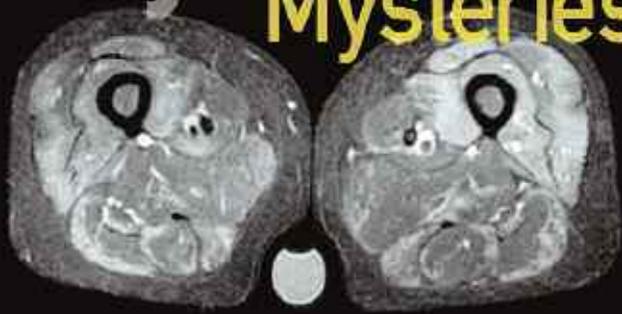


FIGURE 1. Short tau inversion recovery (STIR) image of proximal thighs showing increased signal intensity interpreted as edema in a patient with active polymyositis.

Case Study

A 26-year-old female was referred to the rheumatology service two years after the insidious onset of “walking funny” that progressed to significant proximal muscle weakness over a 10-month period. Early in the course of her illness, her creatine kinase (CK) was 4,000. Subsequent muscle biopsy revealed muscle fiber degeneration and regeneration in addition to numerous phagocytic cells. She was diagnosed with polymyositis but, despite treatment with high-dose corticosteroids, methotrexate (MTX), and monthly intravenous immunoglobulin (IVIG), she did not improve. At the time of presentation to rheumatology, her CK was 8,000, and she reported continued progression of muscle weakness and consequent frequent falls. Her family history was remarkable for a sister with the onset of similar symptoms at 30 years of age. Because of the lack of response to previous appropriate therapies and the family history, we suspected a hereditary myopathy. Her initial muscle biopsy results were consistent with the possibility of a hereditary myopathy, because some dystrophies do show inflammation. Repeat muscle biopsy was recommended for more extensive histopathologic evaluation. Upon immunohistochemical staining, the absence of dysferlin was discovered, consistent with a diagnosis of limb-girdle muscular dystrophy 2B.

Introduction

This case highlights the difficulty in diagnosing polymyositis and the need to think through the appropriate differential diagnosis and to use the appropriate diagnostic modalities when evaluating patients with suspected idiopathic inflammatory myopathy (IIM). Polymyositis (PM), dermatomyositis (DM), and inclusion body myositis (IBM) comprise the major IIM subsets in adults. A further categorization based on associated malignancy or other connective tissue disease features is useful in some circumstances, such as directing therapeutic interventions. Although the IIMs share the characteristics of immune-mediated attacks on skeletal muscle resulting in muscle weakness, they are in fact heterogeneous diseases with varied histopathological and clinical characteristics.¹ Furthermore, these inflammatory myopathies can be confused with other myopathies that sometimes can have an inflammatory component, such as muscular dystrophies.

Differential Diagnosis

The differential diagnosis of a patient with suspected IIM is vast (see Table 1) and highlights the importance of a meticulous history and physical, as well as appropriate and thorough diagnostic testing. The still-useful classification criteria of Bohan and

Why Isn't My Myositis Patient Getting Better?

By Kathleen M. Coyle, MD, Paul H. Plotz, MD, and Mark F. Gourley, MD

Peter require four of the following features for a diagnosis of definite PM or three features plus a characteristic rash for a diagnosis of definite DM:^{2,3}

1. Symmetric, subacute muscle weakness;
2. Abnormal muscle biopsy: fiber size variation, necrosis, regeneration, atrophy, and inflammation;
3. Muscle enzyme elevation: CK, aldolase, alanine transaminase (ALT), aspartate aminotransferase (AST), and lactic dehydrogenase (LDH);



FIGURE 2. Gottron's papules. Raised purple-red areas on the dorsal aspect of metacarpal and interphalangeal joints pathognomonic for dermatomyositis.

TABLE 1.

Myositis Masqueraders⁵

1. Endocrine: Hypothyroidism, Cushing's syndrome, and stiff-person syndrome
2. Toxic: Seven categories
3. Metabolic: McArdle disease, carnitine palmitoyltransferase (CPT) deficiencies, phosphofructokinase deficiency, Pompe disease, and phosphorylase b kinase deficiency
4. Neurologic: Myasthenia gravis, amyotrophic lateral sclerosis, spinal muscular atrophy, Parkinson's disease, and channelopathies (paramyotonia)
5. Muscular dystrophies: Many dominant and recessive forms have been described
6. Electrolyte imbalance: Hypocalcemia/phosphatemia/kalemia/natremia and hypernatremia
7. Infectious: Viral (Coxsackie, Epstein-Barr, human immunodeficiency virus), bacterial (Lyme), fungal, and parasitic (toxoplasmosis, trichinosis)
8. Other: Meningioma, amyloidosis, cystic fibrosis, and Munchausen syndrome

4. Electromyogram abnormality: triad of short, small, polyphasic motor units; insertional irritability, positive sharp waves, fibrillations; and bizarre, high-frequency repetitive discharges; and

5. Typical DM skin abnormalities: Gottron's papules (see Figure 2) or heliotrope rash.

Exclusion criteria include evidence of metabolic, infectious, post-traumatic, and neuromuscular disorders.^{2,3} Although widely cited, Bohan and Peter's criteria were developed prior to the discovery of myositis-specific autoantibodies (MSAs) and have been criticized for their inability to differentiate patients with PM, IBM, the toxic myopathies, and some genetic and metabolic myopathies.¹

Incidence, Demographics, History, and Physical Exam

The collective incidence of the IIM is one in 100,000 per year. The female-to-male ratio for PM and DM is two to one; IBM occurs more frequently in males (three to one). Disease onset in PM is 18 or older; bimodal peaks of onset are seen in DM (5–15 and 45–65 years).^{4,5} IBM is most common in individuals greater than 50 years of age. However, it can occur earlier,

particularly in familial cases. There are many masqueraders of IIM. See Table 2 for a list of characteristics that can point toward or away from a diagnosis of IIM.

Both PM and DM present subacutely over weeks to months with proximal muscle weakness. The photosensitive skin manifestations of DM include facial erythroderma without sparing of the nasolabial folds that is typical in lupus erythematosus; heliotrope rash around the eyes; Gottron's rash/papules (see Figure 2); and shawl and v-neck signs. Patients with MSAs can be categorized by typical features associated with these antibodies. See Table 3 for a list of features associated with particular autoantibodies.

In contrast to the presentation of PM and DM, the onset and progression of IBM is characteristically slow, occurring over months to years. Asymmetric weakness, dysphagia, frequent falling, and distal as well as proximal muscle weakness and wasting are hallmarks of this IIM subset.

IIM can present in overlap with other rheumatic diseases, including systemic lupus erythematosus, progressive systemic sclerosis (scleroderma), Sjögren's syndrome, and RA. The onset of myositis symptoms can occur

at, before, or after diagnosis of these diseases. Overlap with IBM is far less common, but has been reported.

The association of IIM with malignancy is well recognized. In a review of four studies comprising 1,078 myositis patients and a comparable number of controls, the overall odds ratio (OR) for the association of cancer with DM was 4.4 (95%, confidence interval [CI] 3.0, 6.6) and with PM the OR was 2.1 (95% CI 1.4, 3.3).⁶ Malignancies are predominantly adenocarcinomas and usually develop within one year before or after myositis onset, but have been reported to occur up to five years later.⁷ Therefore, age-appropriate cancer screening and ongoing surveillance with a lowered threshold for further investigation is mandated.

Approximately 30% of DM cases present without clinically apparent muscle weakness, the majority of which progress months or years later to clinical myositis.⁸ Patients without evidence of muscle weakness or serum muscle enzyme abnormalities are classified as dermatomyositis sine myositis or amyopathic DM. These patients can develop interstitial lung disease (ILD) which can be rapidly progressive, heralding a poor prognosis.^{9,10}

The importance of evaluation for toxic myopathies cannot be stressed enough, for as a group toxins represent the most common etiology of myopathy. In an elegant review of this topic, Walsh and Amato classified the toxic myopathies to date according to presumed pathogenic mechanism into seven categories:¹¹

1. **Necrotizing myopathy:** Alcohol, antilipemic agents (statins and fibrates), cyclosporin, labetalol, propofol, illicit drugs (cocaine, heroin, amphetamines), and controlled substances (meperidine, pentazocine);
2. **Amphiphilic:** Amiodarone, chloroquine, and hydroxychloroquine;
3. **Antimicrotubular:** Colchicine and vincristine; ➤

TABLE 2. Evaluation of Suspected IIM⁵

Characteristics Suggesting IIM	Characteristics Suggesting Myositis Masquerader
Symmetric, proximal weakness	Temporal relationship of weakness to exercise, eating, or fasting
Rashes: heliotrope, Gottron's, shawl	Neurologic: sensory, weakness, or fasciculations
Pulmonary involvement: IPF, infiltrates	Severe muscle cramping
Features of other autoimmune diseases (e.g., arthritis, Raynaud's)	Family history suggestive of similar illness
CK >2 X or <100 X elevated	CK <2 X or >100 X elevated
Clinical response to immunosuppressives	No clinical response to immunosuppressives
	Hypertrophy at any time or early atrophy

4. Mitochondrial myopathy: Zidovudine;

5. Inflammatory myopathy: L-tryptophan, Dpenicillamine, cimetidine, L-dopa, phenytoin, lamotrigine, interferon- α , hydroxyurea, and imatinib;

6. Hypokalemic myopathy: Diuretics, laxatives, amphotericin, toluene abuse, licorice, corticosteroids, and alcohol abuse; and

7. Unknown: Critical illness myopathy (corticosteroids, nondepolarizing neuromuscular blocking agents, and sepsis), omeprazole, isotretinoin, finasteride, and emetine.

Obtaining a family history of similar symptoms and neuromuscular disease is important; a positive response is most likely indicative of pathology other than IIM, such as in the muscular dystrophy in the case study above. A notable exception is IBM, because both idiopathic and familial forms exist.

Laboratory Tests

Muscle enzymes: As noted above, elevations of serum concentration of intracellular muscle components (CK, aldolase, AST, ALT, LDH, and myoglobin) generally accompany myositis. In IBM, serum CK correlates poorly with disease acuity and severity. It is nor-

mal in 25% of patients and, if abnormal, it is generally only slightly elevated.¹²

Tests commonly called "liver function tests" actually test for liver-associated enzymes such as AST, ALT, and LDH—which are also found in muscle. Unfortunately, it is common for patients to present to rheumatologists after a liver biopsy is performed due to elevated "liver function tests" resulting from active myositis. In this case, the liver biopsy is normal. Also remember that, when monitoring a patient with active myositis and elevated CK being treated with MTX, increased AST and ALT are more likely to reflect active disease than MTX-induced hepatic toxicity.

Autoantibodies: A positive antinuclear antibody titer is present in 60% to 80% of IIM patients; rheumatoid factor is positive in 8%.¹³ See Table 3 for a list of autoantibodies and associated features. These autoantibodies each are generally associated with a characteristic IIM phenotype. As is the case with all autoantibodies, positive and negative results must be interpreted in light of the clinical context, but a positive result can be useful prognostically as well as for guiding further evaluation and treatment.

EMG: As noted above, a myopathic EMG

has a characteristic triad of features. New-onset disease can also result in an added feature of early recruitment. Although the EMG will not point to the specific etiologic entity (disease or toxin), it is helpful in ruling out neuropathic processes.

MRI: MRI has become an important diagnostic modality in IIM and serially may be useful in evaluating response to treatment in individual patients. Short tau inversion recovery images can demonstrate areas of edema that can be interpreted as active myositis (see Figure 1). Although MRI should not substitute for muscle biopsy, it can guide muscle biopsy sampling by identifying sites of active disease. T1 MRI imaging is used to evaluate muscle atrophy and fatty replacement and thus serves as a measure of chronic muscle damage.



FIGURE 3. Mechanic's hands. Roughening of the radial sides of the fingers seen in patients with anti-aminoacyl-tRNA synthetase (anti-Jo-1) autoantibodies.

Muscle biopsy: Muscle biopsy is the essential and definitive diagnostic modality for IIM. Only very rarely should therapy proceed without a biopsy (one case is when a characteristic DM rash is present in a patient with a typical pattern of weakness but biopsy is unavailable). If immediate therapy is clinically deemed wise, a brief delay

before biopsy will not significantly reduce the diagnostic usefulness because most biopsy signs recede slowly.

The pathogenesis of IIM appears to be reflected in the histochemical characteristics of each subset. DM is thought to result from humorally mediated vascular injury (see Figure 4A). Perifascicular and perivascular inflammation and atrophy are most characteristic of DM, and vessel thrombosis, fiber necrosis, and regeneration can also be seen. In contrast, PM and IBM are thought to involve cytotoxic T cells in their pathogenesis. In PM, CD8-positive lymphocytes surround and invade muscle fibers expressing Major Histocompatibility (MHC) class I antigens (see Figure 4B). IBM is likely to be present if the biopsy shows rimmed vacuoles containing basophilic granules and amyloid deposits in addition to inflammation (see Figure 4C).

Treatment

An integral component of IIM treatment is rehabilitation medicine. Patients should be referred for evaluation and treatment by a physiatrist or physical therapist upon diagnosis. Active disease does not preclude patient-specific graded physical and occupational therapy that is combined with appropriate pharmacologic treatment.

Prednisone remains the first-line pharmacologic therapy for PM and DM. We advocate an initial dose of 1 mg/kg/day, with a pulse of methylprednisolone (typically 1 gm) at the outset in significantly affected patients. We favor disease modifying antirheumatic drug (DMARD) initiation (most often MTX at usual rheumatologic doses of up to 25 to 30 mg/week or azathioprine at up to 2 mg/kg/day) within a month of initiating prednisone in order to avoid disease flares while attempting to taper prednisone. If prednisone taper is not tolerated with single therapy of 25 to 30 mg MTX/week or 100 to 150 mg azathioprine/day, then consider a combination of azathioprine and MTX.¹⁴ In our experience using MTX as adjunct therapy

for IIM, MTX-related pulmonary toxicity has been very uncommon.

Opinions vary considerably regarding the role of IVIG in IIM treatment. A controlled clinical trial has demonstrated efficacy in treatment-resistant DM and, in an open-label study, significant muscle strength improvement was reported in treatment-refractory PM.^{15,16} In our opinion, monthly IVIG infusions (1–2 gm/kg/month) should be considered for both DM and PM. Although many patients may benefit substantially, this effect often

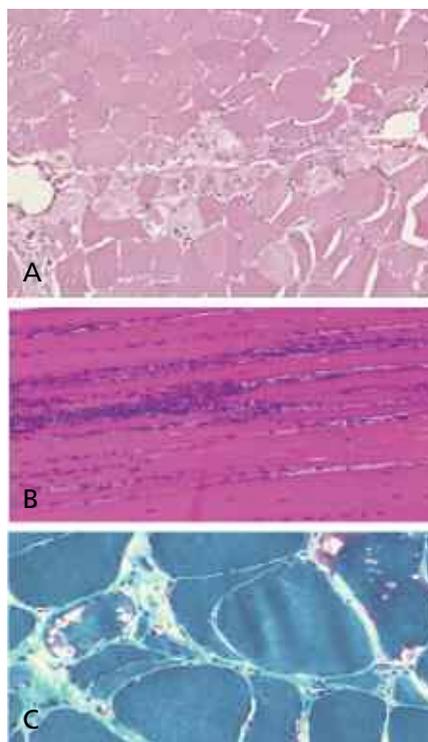


FIGURE 4. Hematoxylin and eosin (H&E) and Gomori trichrome stains of muscle: A) H&E stain demonstrating perivascular and perifascicular inflammation characteristic of DM; B) H&E stain demonstrating foci of endomysial infiltrates by mononuclear cells characteristic of PM; and C) Gomori trichrome stain demonstrating red-rimmed vacuoles characteristic of IBM.

diminishes with a prolonged course of infusions. In cancer-associated myositis, when chemotherapy is used for the treatment of the tumor, IVIG can be a sensible option.

Added options in refractory DM and PM include cyclosporine and tacrolimus, particularly if there is co-existing interstitial lung disease.^{17,18} Early uncontrolled reports using mycophenylate mofetil have shown mixed results and no controlled trial has been reported. Because reports of cyclophosphamide efficacy in IIM are conflicting, this medication should be reserved for cases resistant to other immunosuppressants and IVIG.¹⁹

Despite some evidence for an immune contribution to the pathogenesis of IBM, and although some patients respond transiently to conventional IIM treatment, no therapy substantially affects the progression of this disease. However, no rigorous long-term trial has been reported.²⁰ Significant improvement in dysphagia symptoms following IVIG has been reported.²¹

The role of anti-tumor necrosis factor or B cell-depleting therapies in IIM is unknown. Published uncontrolled case reports describe varied outcomes. Ongoing controlled clinical trials investigating the role of these agents in PM/DM may clarify the potential efficacy of these therapies.

Monitoring indices of active disease is important in guiding therapy and should not be limited to periodic measurement of serum muscle enzymes. The International Myositis Assessment and Clinical Studies (IMACS) international collaboration has developed a core set of three outcomes measures for PM and DM: myositis disease activity, damage, and health-related quality of life.^{22,23} Although developed specifically for use in PM and DM clinical trials, these outcomes measures can be useful in clinical practice. The principle of comprehensive disease assessment is also applicable to IBM.

One of the most important principles of IIM treatment is that the question, “Is this IIM?,” should not be dismissed following initial diagnosis. Maintain a high index of suspicion and reassess the question in the following settings. When in doubt, re-biopsy. ➤

1. Persistent elevated muscle enzymes despite appropriate treatment and improving symptoms: Treat the patient, not the number! MRI can identify active inflammation in patients with conflicting indices of disease activity, such as persistent CK elevations without corresponding muscle weakness or vice versa;
2. Persistent or increasing muscle weakness with or without CK change: This could represent active IIM, but a high index of suspicion for steroid myopathy or a high degree of muscle damage must be maintained; and
3. Evolving symptoms suggestive of IBM in a patient whose muscle biopsy was read as PM: This probably is IBM, particularly in concert with appropriate clinical characteristics.

Conclusion

The IIMs are a heterogenous group of immune-mediated myopathies. Thorough consideration of the potential differential diagnosis and careful examination of a muscle biopsy are essential for accurate diagnosis and choice of appropriate therapy. Cornerstones of therapy are corticosteroids, DMARDs, and physiatry. If your myositis patient is not responding, rethink the diagnosis and consider appropriate consultations, laboratory studies, imaging, or re-biopsy. ■

Dr. Coyle is a clinical fellow in rheumatology at the National Institute of Arthritis, Skin and Musculoskeletal Diseases (NIAMS), National Institutes of Health (NIH) in Bethesda, Md. Dr. Plotz is chief of the arthritis and rheumatism branch of NIAMS/NIH. Dr. Gourley is program director of rheumatology fellowship training at NIAMS/NIH. This article was supported by the Intramural Research Program of NIAMS/NIH.

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TABLE 3. Autoantibodies in IIM

Autoantibody	Features
Anti-aminoacyl-tRNA synthetases (Anti-: Jo-1, PL7, PL-12, OJ, EJ, KS)	PM or DM Acute onset ILD, arthritis, fever, Raynaud's syndrome Mechanic's hands (See Figure 3)
Anti-Mi-2	DM Shawl and v-neck signs Cuticular overgrowth
Anti-SRP	PM Seasonal onset (often in the fall), abrupt onset Rash rare Often treatment refractory
Anti-PM-Scl (myositis-associated autoantibody)	PM or DM Scleroderma PM or DM overlap with scleroderma

References

1. Di Martino SJ, Kagen LJ. Newer therapeutic approaches: Inflammatory muscle disorders. *Rheum Dis Clin North Am.* 2006;32:121-128, ix.
2. Bohan A, Peter J. Polymyositis and dermatomyositis (first of two parts). *New Engl J Med.* 1975;(292):344-347.
3. Bohan A, Peter J. Polymyositis and dermatomyositis (second of two parts). *New Engl J of Med.* 1975;(292):403-407.
4. Flachenecker P. Epidemiology of neuroimmunological diseases. *J Neurol* 2006;253 [Suppl 5], V/2-V/8.
5. Christopher-Stine L, Plotz PH. Adult inflammatory myopathies. *Best Pract Res Clin Rheumatol.* 2004;18(3):331-344.
6. Zantos D, Zhang Y, Felson D. The overall and temporal association of cancer with polymyositis and dermatomyositis. *J Rheumatol.* 1994;21(10):1855-1859.
7. Levine SM. Cancer and myositis: New insights into an old association. *Curr Opin Rheumatol.* 2006;18(6):620-624.
8. Gerami P, Schope JM, McDonald L, Walling HW, Sontheimer RD. A systematic review of adult-onset clinically amyopathic dermatomyositis (dermatomyositis sine myositis): A missing link within the spectrum of the idiopathic inflammatory myopathies. *J Am Acad Dermatol.* 2006;54(4):597-613.
9. Suda T, Fujisawa T, Enomoto N, et al. Interstitial lung diseases associated with amyopathic dermatomyositis. *Eur Respir J.* 2006;28(5):1005-1012.
10. Ideura G, Hanaoka M, Koizumi T, et al. Interstitial lung disease associated with amyopathic dermatomyositis: Review of 18 cases. *Respir Med.* 2007;101(7):1406-1411.
11. Walsh RJ, Amato AA. Toxic myopathies. *Neurol Clin.* 2005;23(2):397-428.
12. Wortmann RL. Inflammatory Diseases of Muscle and Other Myopathies. In: Harris ED, Ruddy S, Budd RC, et al., eds. *Kelly's Textbook of Rheumatology, Seventh Edition.* 2, 1309-1335. 2005. Philadelphia PA, Elsevier Saunders.
13. Targoff IN. Laboratory testing in the diagnosis and management of idiopathic inflammatory myopathies. *Rheum Dis Clin North Am.* 2002;28(4):859-890, viii.
14. Villalba L, Hicks JE, Adams EM, et al. Treatment of refractory myositis: A randomized crossover study of two new cytotoxic regimens. *Arthritis Rheum.* 1998; 41(3):392-399.
15. Dalakas MC, Illa A, Dambrosia JM, et al. A controlled trial of high dose intravenous immune globulin infusions as treatment for dermatomyositis. *New Engl J Med.* 1993;329:1993-2000.
16. Cherin P, Herson S, Wechsler B, et al. Efficacy of intravenous gammaglobulin therapy in chronic refractory polymyositis and dermatomyositis: An open study with 20 adult patients. *Am J Med.* 1991;91(2):162-168.
17. Oddis CV, Sciruba FC, Elmagd KA, Starzl TE. Tacrolimus in refractory polymyositis with interstitial lung disease. *Lancet.* 1999;22;353(9166):1762-1763.
18. Ytterberg SR. Treatment of refractory polymyositis and dermatomyositis. *Curr Rheumatol Rep.* 2006;8(3):167-173.
19. Cordeiro AC, Isenberg DA. Treatment of inflammatory myopathies. *Postgrad Med J.* 2006;82(969):417-424.
20. Griggs RC. The current status of treatment for inclusion body myositis. *Neurology.* 2006;24;66(2 Suppl 1):S30-S32.
21. Cherin P, Pelletier S, Teixeira A, et al. Intravenous immunoglobulin for dysphagia of inclusion body myositis. *Neurology.* 2002;58(2):326.
22. Miller FW, Rider LG, Chung YL, et al. Proposed preliminary core set measures for disease outcome assessment in adult and juvenile idiopathic inflammatory myopathies. *Rheumatology (Oxford).* 2001;40:1262-1273.
23. IMACS Web site. <https://dir-apps.niehs.nih.gov/imacs/index.cfm?action=home.main>. Last accessed December 4, 2007.

Resource Directory

Ataxia Telangiectasia (A-T)

Websites

- A-T Children's Project: www.atcp.org
- NINDS A-T Information Page: www.ninds.nih.gov/disorders/a_t/a-t.htm

Chronic Inflammatory Demyelinating Polyneuropathy (CIPD)

Websites

- New!** • GBS/CIDP Foundation International: www.gbs-cidp.org
- National Institute of Neurological Disorders and Stroke (NINDS) Chronic Inflammatory Demyelinating Polyneuropathy (CIPD) Information Page www.ninds.nih.gov/disorders/cidp
- Neurology Muscular Dystrophy and Neuropathy Institute Illustration of a damaged myelin sheath on a nerve: www.beverlyhillsneurology.com/cidp

Online Peer Support Links

- The Neuropathy Association: www.neuropathy.org
- Barbara's CIDP/GBS Site (This is a personal website) www.geocities.com/HotSprings/Falls/3420

Evans Syndrome

Websites

- Evans Syndrome Research and Support Group: www.evanssyndrome.org
- Office of Rare Diseases (catalog of online resources) <http://rarediseases.info.nih.gov/asp/diseases/diseaseinfo.asp?ID=6389>

Guillain-Barré Syndrome (GBS)

Websites and Chat Rooms

- New!** • The GBS/CIDP Foundation International, www.gbs-cidp.org, has 23,000 members in 160 chapters on five continents. 610-667-0131
- New!** • The GBS/CIDP Foundation International Discussion Forums provide the opportunity to talk to other GBS patients and learn more about ways to manage the illness: www.gbs-cidp.org/forums.
- Miller Fisher Syndrome, considered to be a variant of GBS, is explained on the National Institute of Neurological Disorders and Stroke, NINDS Miller Fisher Syndrome Information Page: www.ninds.nih.gov/disorders/miller_fisher/miller_fisher.htm.

Online Pamphlets

- The National Institute of Neurological Disorders and Stroke has a GBS Fact Sheet at www.ninds.nih.gov/disorders/gbs/detail_gbs.htm.
- The Mayo Clinic has an overview of Guillain-Barré Syndrome at www.mayoclinic.com/health/guillain-barre-syndrome/DS00413.
- The National Institute of Neurological Disorders and Stroke has an information page about CIDP: www.ninds.nih.gov/disorders/cidp/cidp.htm.

Online Peer Support

- GBS & CIDP Discussion Forum – UK Bulletin Board – For Ireland and England www.gbs.org.uk/cgi-bin/ikonboard3/ikonboard.cgi
- GBS Support Group and Chat Room, UK www.jsmarcussen.com/gbs/uk/chat.htm

- GBS Foundation Discussion Forums: www.guillain-barre.com/forums
- Yahoo Support Group Discussion Board http://health.groups.yahoo.com/group/GBS_CIDP

Books and Articles

- "A Handbook for Care Givers," by Helen Rummelsburg, gives an account of caring for her husband and offers helpful hints for patients and caregivers. Available at the GBS website bookstore at www.gbsfi.com.
- "Bed Number Ten," by Sue Baier, provides a view of long-term care through the eyes of a patient totally paralyzed with GBS.
- "Caring for a Child With GBS," by Patricia Schardt, is a short guide written by a mother of a child with CIDP. Available at the GBS website bookstore at www.gbsfi.com.
- "No Laughing Matter," by Joseph Heller (the best-selling author of Catch-22), who teamed up with Speed Vogel, his best friend, to describe Heller's battle with and triumph over GBS.

ITP (Idiopathic Thrombocytopenic Purpura)

Websites

- ITP Support Association, UK: www.itpsupport.org.uk
- Platelet Disorder Support Association: www.pdsa.org
- National Heart, Lung and Blood Institute www.nhlbi.nih.gov/health/dci/Diseases/Itip/ITP_Whats.html

Online References

- New!** • Idiopathic thrombocytopenic purpura www.mayoclinic.com/health/idiopathic-thrombocytopenic-purpura/DS00844
- Idiopathic Thrombocytopenic Purpura: Michael A. Silverman, MD www.emedicine.com/EMERG/topic282.htm
- Immune Thrombocytopenia: Current Understanding and Future Research, Robert S. Mocharnuk, MD www.medscape.com/viewarticle/459249
- ITP: Idiopathic Thrombocytopenic Purpura: <http://familydoctor.org/113.xml>

Kawasaki Disease

Websites

- Kawasaki Disease Foundation: www.kdfoundation.org
PO Box 45 • Boxford, MA 01921
Tel: 978-356-2070 • Fax: 978-356-2079 • Email: info@kdfoundation.org
- Kids Health discusses Kawasaki disease www.kidshealth.org/parent/medical/heart/kawasaki.html.
- Overview from the American Heart Association focuses on how the disease affects the heart www.americanheart.org/presenter.jhtml?identifier=4634

Mitochondrial Disease

Websites

- United Mitochondrial Disease Foundation promotes research and education for the diagnosis, treatment and cure of mitochondrial disorders and provides support to affected individuals and families. www.umdf.org

- The Cleveland Clinic website provides many articles when searched by the topic, "mitochondrial disease." www.clevelandclinic.org/health
- The National Institute of Neurological Disorders and Stroke provides an informative Mitochondrial Myopathies Information Page. www.ninds.nih.gov/disorders/mitochondrial_myopathy/mitochondrial_myopathy.htm

Multifocal Motor Neuropathy (MMN)

Websites

- National Institute of Neurological Disorders and Strokes (NINDS) provides a Multifocal Motor Neuropathy Information Page: www.ninds.nih.gov/disorders/multifocal_neuropathy/multifocal_neuropathy.htm
- Multifocal Motor Neuropathy Center at Johns Hopkins Department of Neurology www.neuro.jhmi.edu/MMN/index.html
- The Neuromuscular Center at Washington University in St. Louis, Mo. Neuromuscular Home Page www.neuro.wustl.edu/neuromuscular
- The Neuropathy Association is dedicated to helping those with conditions affecting peripheral nerves. www.neuropathy.org

Multiple Sclerosis (MS)

Websites and Chat Rooms

- The mission of the National Multiple Sclerosis Society is to end the devastating effects of MS. www.nationalmssociety.org/
- All About Multiple Sclerosis provides accurate and comprehensive medical information about MS written in plain English by people living with the disease and its symptoms. www.mult-sclerosis.org/index.html
- Multiple Sclerosis Foundation works for a brighter tomorrow for those affected by MS. www.msfacts.org
- Multiple Sclerosis Association of America seeks to enrich the quality of life for individuals with multiple sclerosis. www.msaa.com
- MSWorld's Chat and Message Board features patients helping patients. www.msworld.org

Online Peer Support

- Friends with MS: <http://friendswithms.com>
Forum: <http://health.groups.yahoo.com/group/FriendsWithMS>
- My MSViews: www.mymviews.org
Forum: http://health.groups.yahoo.com/group/MSViews_Multiple_Sclerosis
- MS Support Group: <http://health.groups.yahoo.com/group/mscured>
- The MS Carousel—A Place to Meet With People Who Understand MS!
<http://health.groups.yahoo.com/group/themscarousel>

Myasthenia Gravis (MG)

Websites and Chat Rooms

- The Myasthenia Gravis Foundation of America (MGFA) is the only national volunteer health agency dedicated solely to the fight against (MG). www.myasthenia.org
- Myasthenia Gravis Fact Sheet prepared by National Institute of Neurological Disorders and Strokes. www.ninds.nih.gov/disorders/myasthenia_gravis/myasthenia_gravis.htm
- Mayo Clinic's overview of myasthenia gravis: www.mayoclinic.com/health/myasthenia-gravis/DS00375

Online Peer Support

- MGFA's Forum: <http://health.groups.yahoo.com/group/MGnet>

- Bette's Myasthenia Gravis Support: <http://health.groups.yahoo.com/group/bettesmyastheniagravisupport>
- Maddy's MG Support: <http://health.groups.yahoo.com/group/maddysmgsupport>
- Autoimmune Information Network Inc.: www.aininc.org
PO Box 4121 • Brick, NJ 08723 • 877-246-4900 • Email: autoimmunehelp@aol.com

Myositis

Websites

 <p>THE MYOSITIS ASSOCIATION</p>	<p>The mission of The Myositis Association, www.myositis.org, is to find a cure for inflammatory and other related myopathies, while serving those affected by these diseases. 202-887-0088</p>
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- International Myositis Assessment and Clinical Studies Group is a coalition of healthcare providers and researchers with global approaches to improved treatments and understanding of myositis: <https://dir-apps.niehs.nih.gov/imacs/index.cfm?action=home.main>
- The Cure JM Foundation was created specifically to find a cure for Juvenile Myositis (JM), while also providing support and information for families affected by JM. <http://curejm.com>
- Johns Hopkins Myositis Center is a new patient treatment center that brings the expertise of rheumatologists and neurologists into a single clinic for patients with inflammatory (autoimmune) and toxic (drug induced) muscle conditions. www.hopkinsmedicine.org/rheumatology/clinics/myositis_center.html

Online Peer Support

- Juvenile Myositis Family Support Network: www.curejm.com/family_support/index.htm
- Myositis Association Community Forum: www.myositis.org
- Myositis Support Group: www.myositisupportgroup.org
- Myositis Support Group UK: www.myositis.org.uk
- Yahoo Myositis Support Group Discussion Board: <http://health.groups.yahoo.com/group/OurMyositis>
- The California Myositis Symposium held in 2005 was captured on DVD. It contains information about polymyositis, dermatomyositis and inclusion body myositis, including doctors' discussions and detailed slides and explanations of muscle biopsies, skin rash, and tools used to diagnose these diseases. Other presentations offer valuable lessons in maintaining a positive attitude, exercises for physical therapy and innovative tools to aid in everyday activities. The DVD is available at no charge by sending an email to Richard Gay at rgay@socal.rr.com.

Books and Articles

- "Coping With a Myositis Disease," by James R. Kilpatrick, is written by myositis patients telling their personal stories.
- "Inclusion-Body Myositis and Myopathies," by Valerie Askanas (Editor), Georges Serratrice (Editor) and W. King Engel (Editor), is devoted to discussing the two forms of inclusion-body myositis.
- "Living With Myositis," edited by Jenny Fenton, is an accessible, realistic and sympathetic guide to facts, feelings and future hopes.
- "Myositis—A Medical Dictionary, Bibliography, and Annotated Research Guide to Internet References," by ICON Health Publications, is a three-in-one reference book: a complete dictionary of terms relating to myositis, a list of bibliographic citations about the disorder and a guide to Internet resources.
- "Myositis and You: A Guide to Juvenile Dermatomyositis for Patients, Families, and

Healthcare Providers,” an indispensable resource for families of children with myositis and their healthcare teams, is available at www.myositis.org.

- “The Official Patient’s Sourcebook on Inclusion Body Myositis,” by James N. Parker (Editor) and Philip M. Parker (Editor), is a reference manual for self-directed patient research.

Pemphigus and Pemphigoid

Websites

- The International Pemphigus and Pemphigoid Foundation provides information and support to people living with the autoimmune diseases. www.pemphigus.org
- Information from the National Institutes of Health: www.niams.nih.gov/hi/topics/pemphigus/pemphigus.htm
www.nlm.nih.gov/medlineplus/ency/article/000882.htm
- Rare disease report: <http://rarediseases.about.com/od/rarediseases/p/pemphigus05.htm>

Peripheral Neuropathy (PN)

Websites



The Neuropathy Association, www.neuropathy.org, is devoted exclusively to all types of neuropathy, which affects upwards of 20 million Americans. The Association’s mission is to increase public awareness of the nature and extent of PN, facilitate information exchanges about the disease, advocate the need for early intervention and support research into the causes and treatment of neuropathies. 212-692-0662

- To learn about PN, how it is classified, the symptoms, causes and treatments, see the Peripheral Neuropathy Fact Sheet available at www.ninds.nih.gov/disorders/peripheralneuropathy/peripheralneuropathy.htm.
- The Neuropathy Action Foundation, at www.neuropathyaction.org, educates, empowers and informs patients and physicians about neuropathy.

Support Groups

- Click on the Member Services tab of the website, www.neuropathy.org, for listings of support groups across the nation.

Online Peer Support

- Calgary Neuropathy Support Group: www.calgarypn.org
- MSN Support Group Discussion Board: <http://groups.msn.com/PNPARTNERS>
- The Neuropathy Association Bulletin Board: www.neuropathy.org
- Yahoo Neuropathy Support Group Discussion Board: <http://health.groups.yahoo.com/group/neuropathy>
- Yahoo Support Group – Australia Discussion Board: <http://au.groups.yahoo.com/group/LifeWithPN>

Books and Articles

- “If You’re Having a Crummy Day, Brush Off the Crumbs!,” by Mims Cushing, is a how-to book that offers more than 75 ways to help people get through the days when neuropathy (or other ailments) is particularly difficult.
- “Medifocus Guide to Peripheral Neuropathy,” is a guide to current and relevant PN research, organized into categories for easy reading.
- “Numb Toes and Aching Soles,” by John Senneff, discusses the symptoms, causes, tests, treatments and coping strategies for peripheral neuropathy.
- “Numb Toes and Other Woes,” by John Senneff, is the second in a series of

three books. It focuses on clinical findings and treatment strategies for PN.

- “Nutrients for Neuropathy,” by John Senneff, the third in the Numb Toes series, is focused exclusively on nutrient supplementation as a means for managing PN.
- “Peripheral Neuropathy: When the Numbness, Weakness, and Pain Won’t Stop” by Dr. Norman Latov, MD, PhD, published 2007, Weill Medical College, Cornell University, provides practical information on all the neuropathies, causes and treatments.

Primary Immune Deficiency Disease (PID)

Websites and Chat Rooms



The Immune Deficiency Foundation (IDF), www.primaryimmune.org, is dedicated to improving the diagnosis and treatment of PID through research and education. 800-296-4433

Jeffrey Modell  Foundation

The Jeffrey Modell Foundation, www.info4pi.org, is dedicated to early and precise diagnosis, meaningful treatments and, ultimately, cures for primary immunodeficiency. 212-819-0200

- The National Institute of Child Health and Human Development (NICHD), www.nichd.nih.gov, is part of the National Institutes of Health. Go to the “Health Information and Media” tab on the website and do a search under “primary immunodeficiency.”
- The American Academy of Allergy, Asthma & Immunology, www.aaaai.org, has a helpful Q&A section on its website, with resources and tips for those with various immune deficiencies.
- The Michigan Immunodeficiency Foundation, www.midf.org, seeks to improve the quality of life for Michigan residents affected by PID.
- The International Patient Organization for Primary Immunodeficiencies (IPOPI), www.ipopi.org, promotes the worldwide improvement in the care and treatment of PID patients.
- To connect to a PID message board, go to www.info4pi.org.
- To chat with peers on IDF’s Forum, go to www.primaryimmune.org/forums/forum_intro.htm.
- Chat with parents of children affected by primary immune deficiency at <http://health.groups.yahoo.com/group/PedPID>.
- Chat with peers with PID at <http://health.groups.yahoo.com/group/PIDsupport>.
- A group of family and friends of patients with primary immune deficiencies maintains a nonprofit network in the New England area: www.nepin.org
- Baxter’s website, www.immunedisease.com, offers in-depth information on immunology, PID and treatment with intravenous immune globulin. Click on “European” to see SCIG information.
- New!** • Rainbow Allergy-Immunology, www.rainbowbabies.org/immunology, provides comprehensive diagnostic, therapeutic and consultative services for children and adults with immunologic diseases. For patient information about subcutaneous IG therapy: www.rainbowbabies.org/subcu.

Online Pamphlets and Education

- Go to the National Institute of Allergy and Infectious Diseases site at www.niaid.nih.gov and search for “primary immune deficiency.”
- “Understanding the Immune System: How It Works,” by the U.S. Department of Health and Human Services, is found at www3.niaid.nih.gov/healthscience/healthtopics/immuneSystem/PDF/theImmuneSystem.pdf

- “NIAID Initiative Addresses Primary Immune Deficiency Diseases by National Institute of Allergy and Infectious Diseases” is located at <http://www3.niaid.nih.gov/news/newsreleases/2003/pirc.htm>
- The “Immunodeficiency in Pediatrics” program (PREP®) audio series is a new pediatrician education program that can be obtained by contacting the American Academy of Pediatrics at 866-843-2271 or visiting www.prepaudio.org.

Online Peer Support

- Chat with parents of children affected by PIDD <http://health.groups.yahoo.com/group/PedPID/>
- Chat with peers with PIDD: <http://health.groups.yahoo.com/group/PIDsupport/>
- Immune Deficiency Foundation Forum www.primaryimmune.org/forums/forum_intro.htm
- Jeffrey Modell Foundation Message Board: www.info4pi.org

New! • Rhode Island peer group <http://health.groups.yahoo.com/group/RhodelslandPID/>

Books and Articles

- “21st Century Complete Medical Guide to Primary Immune Deficiency, Severe Combined Immunodeficiency (SCID), Chronic Granulomatous Disease (CGD), for Patients and Physicians,” by PM Medical Health News, contains federal government clinical data and practical information for patients and physicians.

Scleroderma

Websites

- Johns Hopkins Medicine Scleroderma Center: scleroderma.jhmi.edu
- Scleroderma Research Foundation: www.srfcure.org
- Scleroderma Foundation: www.scleroderma.org

Online Peer Support

- Educating instead of medicating CureZone.com <http://curezone.com/forums/f.asp?f=404>
- International Scleroderma Network: www.sclero.org/support/forums/a-to-z.html

Stiff-Person Syndrome (SPS)

Websites

- American Autoimmune Related Diseases Association Inc., www.aarda.org, is the only national organization dedicated to addressing the problem of autoimmunity. 800-598-4668 aarda@aarda.org
- Autoimmune Information Network Inc., www.aininc.org, helps patients and family cope with the disabling effects of autoimmune diseases. 732-262-0450 autoimmunehelp@aol.com
- National Association for Rare Disorders (NORD), www.rarediseases.org, promote awareness of rare diseases and the need for research. 800-999-6673 orphan@rarediseases.org
- National Institute of Neurological Disorders and Stroke (NINDS), www.ninds.nih.gov, offers treatment, diagnosis and research information for rare diseases. 800-352-9424 braininfo@ninds.nih.gov
- Mayo Clinic — Stiff-Person Syndrome: Can it be treated? www.mayoclinic.com/health/stiff-person-syndrome/AN01377
- Diagnosed with SPS in 1994, Debra Kemery recounts her experience and offers practical information about coping with the disease at www.stiffman.org.

Books and Articles

New! • “A Flower Grows in Stone: The Diary of a Life in Progress,” a firsthand account of her struggle with Stiff-Person/Moersch Woltman Syndrome, is written by

first-time author Anisah Hassan. Available from Llumina Press, Ingram, and Baker & Taylor, www.llumina.com.

General Resources

Product Information

- Influenza and the influenza vaccine www.cdc.gov/flu or call 800-CDC-INFO (800-232-4636)
- IVIG Carimune NF www.carimune.com
- IVIG Flebogamma www.grifolsusa.com/pdfs/flebo_14Jun05.pdf
- IVIG Gammagard Liquid www.gammagardliquid.com
- IVIG Gammagard S/D www.immunedisease.com/US/therapies/gammagard/index.html
- IVIG Gamunex www.gamunex.com
- IVIG Octagam www.octapharma.com/corporate/03_products_and_therapeutic_areas/01_immunoglobulin_product_line/03_octagam.php
- SCIG (subcutaneous immune globulin) Vivaglobin www.vivaglobin.com

Other Organizations

- Alliance for Plasma Therapies is a unified, powerful voice of patient organizations, healthcare providers and industry to advocate for fair access to plasma therapies. www.plasmaalliance.org
- For suggestions on how to deal with the medical and emotional impact of caring for an ill child, go to www.kidshealth.org/parent/system/ill/seriously_ill.html.
- The National Committee for Quality Assurance provides free access to detailed report cards on health plans, clinical performance, member satisfaction, access to care and overall quality on its Health Plan Report Cards Online at www.ncqa.org.
- The nonprofit Patient Advocate Foundation, www.patientadvocate.org, seeks to assure patient access to care, maintenance of employment and financial stability. 800-532-5274



The nonprofit Patient Services Incorporated, www.uneedpsi.org, specializes in health insurance premium, pharmacy co-payment and co-payment waiver assistance for people with chronic illnesses. 800-366-7741

- WebMD, www.webmd.com, is a handy medical reference that helps consumers take an active role in managing their health by providing objective healthcare and lifestyle information.
- For a pediatrician’s guide to your child’s health and safety, visit www.keepkidshealthy.com.
- The National Organization for Rare Diseases, at www.rarediseases.org, provides links to numerous other organizations that have disease-specific support groups and virtual communities for patients and caregivers.
- American Autoimmune Related Diseases Association (AARDA) www.aarda.org brings national focus to autoimmunity through research, education and patient services. 800-598-4668
- Band-Aides and Blackboards, www.lehman.cuny.edu/faculty/jfleitas/bandaides, started by a nurse working on her dissertation, focuses on children with chronic illness.
- American Chronic Pain Association (ACPA) was founded in 1980 to provide resources for people coping with chronic pain. www.theacpa.org

Education and Disability Resources

- Continuation of Health Coverage—Consolidated Omnibus Budget Reconciliation Act (COBRA): www.dol.gov/dol/topic/health-plans/cobra.htm

- Social Security: www.ssa.gov/disability
- California State Disability Insurance (SDI): www.edd.ca.gov (Please note that each state has a different disability program.)
- IDEA 2004 Resources: www.ed.gov/policy/speced/guid/idea/idea2004.html
News and information on the Individuals with Disabilities Education Improvement Act of 2004 (IDEA), the nation's law that works to improve results for infants, toddlers, children and youth with disabilities.
- The National Disabilities Rights Network: www.ndrn.org. This website offers a search tool to find resources in your state to assist with school rights and advocacy.
- U.S. Department of Education Website: www.ed.gov. This website, a U.S. federal government website, offers a parents section that has a subsection titled "My Child's Special Needs" that can be most helpful.
- U.S. Department of Health and Human Services, Office of Civil Rights, www.hhs.gov/ocr/504.html, spells out your rights under Section 504 of the Rehabilitation Act.
- Wrightslaw: www.wrightslaw.com. Parents, educators, advocates, and attorneys come to Wrightslaw for accurate, reliable information about special education law and advocacy for children with disabilities.
- The Americans with Disabilities Act of 1990
Provides protection for people with disabilities from certain types of discrimination and requires employers to provide some accommodations of the disability. For more information, visit www.usdoj.gov/crt/ada/adahom1.htm.

Additional Reading

- "Anatomy of an Illness," by Norman Cousins, is a best-seller about overcoming illness and the triumph of the human spirit. The premise is that the human mind is capable of promoting the body's capacity for combating illness and healing itself even when faced with a seemingly hopeless medical predicament.
- "A Parent's Guide to Special Education: Insider Advice on How to Navigate the System and Help Your Child Succeed," by Linda Wilmschurst and Alan W. Brue, is available on Amazon.com.
- "The Confused Consumer's Guide to Choosing a Health Care Plan: Everything You Need to Know," by Martin Gottlieb, helps consumers through the confusing maze of choosing a healthcare plan.
- "The Everyday Guide to Special Education Law," by Randy Chapman, Esq., makes the law accessible to parents so they can be more effective advocates for their children. Available at www.thelegalcenter.org/thelegalcenter-cgi-bin/shop?item=15.
- "Living Creatively With Chronic Illness: Developing Skills for Transcending the Loss, Pain and Frustration," by Eugenia G. Wheeler, is a self-help book specifically designed to help the chronically ill, their families, friends, counselors, medical personnel and the clergy.
- "Managing Pain Before It Manages You," by Dr. Margaret A. Caudill, is a wellspring of wisdom and practical approaches that can help transform your life and your pain.
- "Not Dead Yet: A Long Strange Trip From Doctor to Patient and Back Again," by Dr. Robert Buckman, an oncologist and comic writer, is a witty account of his life as a doctor and autoimmune disease survivor.
- "Pride and the Daily Marathon," by Jonathan Cole, describes how Ian Waterman was suddenly struck down at work by a rare neurological illness that deprived him of all sensation below the neck, and how he reclaimed a life of full mobility.
- "Pronoia Is the Antidote for Paranoia," by Rob Breznsy, explores the best way to attract the blessings that the world is conspiring to give us.
- "When You're Ill or Incapacitated" comprises one-half the booklet it shares with "When You're the Caregiver," both written by James E. Miller, suggesting 12 things to remember or do in each role.
- "YOU the Smart Patient: An Insider's Handbook for Getting the Best Treatment,"

by Michael F. Roizen, MD, and Mehmet C. Oz, MD, with the Joint Commission on Accreditation of Healthcare Organizations, shows you how to tackle such healthcare decisions as picking the best doctors and hospitals for you, knowing when to get a second opinion, and more.

IG Manufacturer Websites

- Baxter: www.baxter.com
- CSL Behring: www.cslbehring.com
- Grifols: www.grifolsusa.com
- Octapharma: www.octapharma.com
- Talecris: www.talecris.com

Pump and Infusion Sets Websites

- EMED Corporation: www.safetymedicalproducts.com
- Graseby Marcal Medical: www.marcalmedical.com
- Intra Pump Infusion Systems: www.intrapump.com
- Repro Med Systems, Inc: www.repro-med.com
- Norfolk Medical: www.norfolkmedical.com

Medical Research Studies

- Check out the official website for the National Institutes of Health patient recruitment program. This site provides summaries and criteria for studies as well as the ability to search for studies being conducted for a specific disease or disorder. <http://clinicalstudies.info.nih.gov>
- This website provides a wealth of information about clinical trials and volunteer participation. It gives you the ability to specify the disorder you are interested in, the location of the study, and the medication names or research protocols. www.centerwatch.com
- This site has a registration form to request that you be notified about recruitment for future studies. www.clinicaltrials.com
- WebMD has a service that matches volunteers with trials. There is an online questionnaire to complete and you will be notified via email of upcoming studies that match the criteria of your questionnaire. You can also search for specific studies. www.webmd.com

Food Allergies

- Allergic Disorders: Promoting Best Practice
www.theallergyreport.com/reportindex.html
- American Partnership for Eosinophilic Disorders: www.apfed.org
- National Institutes of Health, National Institute of Allergy and Infectious Diseases (2004). Food Allergy: An Overview (NIH Publication No. 04-5518). www.niaid.nih.gov/publications/pdf/foodallergy.pdf
- Food Allergy and Anaphylaxis Network: 800-929-4040 www.foodallergy.org
- World Allergy Organization: www.worldallergy.org
- Sicherer, S.H. (2006). "The Complete Peanut Allergy Handbook: Understanding and Managing Your Child's Food Allergies," Johns Hopkins Press.

Reading Just for Kids

- "Germs Make Me Sick," by Melvin Berger, explains with colorful illustrations how your body fights germs.
- "Little Tree: A Story for Children With Serious Medical Illness," by Joyce C. Mills, is a comforting fable for young children facing serious life challenges.
- "My IVIG Book," written from a 3-year-old's perspective about his infusions, comes with a kit for other children to create their own personalized book. Free from Baxter at www.immunedisease.com/US.
- "Our Immune System," enables children who are immune deficient and their families to explore together the immune system. Available from the Immune Deficiency Foundation at www.primaryimmune.org.

Have something to add to these pages? Please send your suggestions for additions to the IG Living Resource Directory to editor@igliving.com. In this case, more is indeed better!

When it comes to flu vaccine,
timing is everything!



June – Time to place your order
on MyFluVaccine.com

- Now, no prepayment required!
- Order online, by phone or by fax.



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