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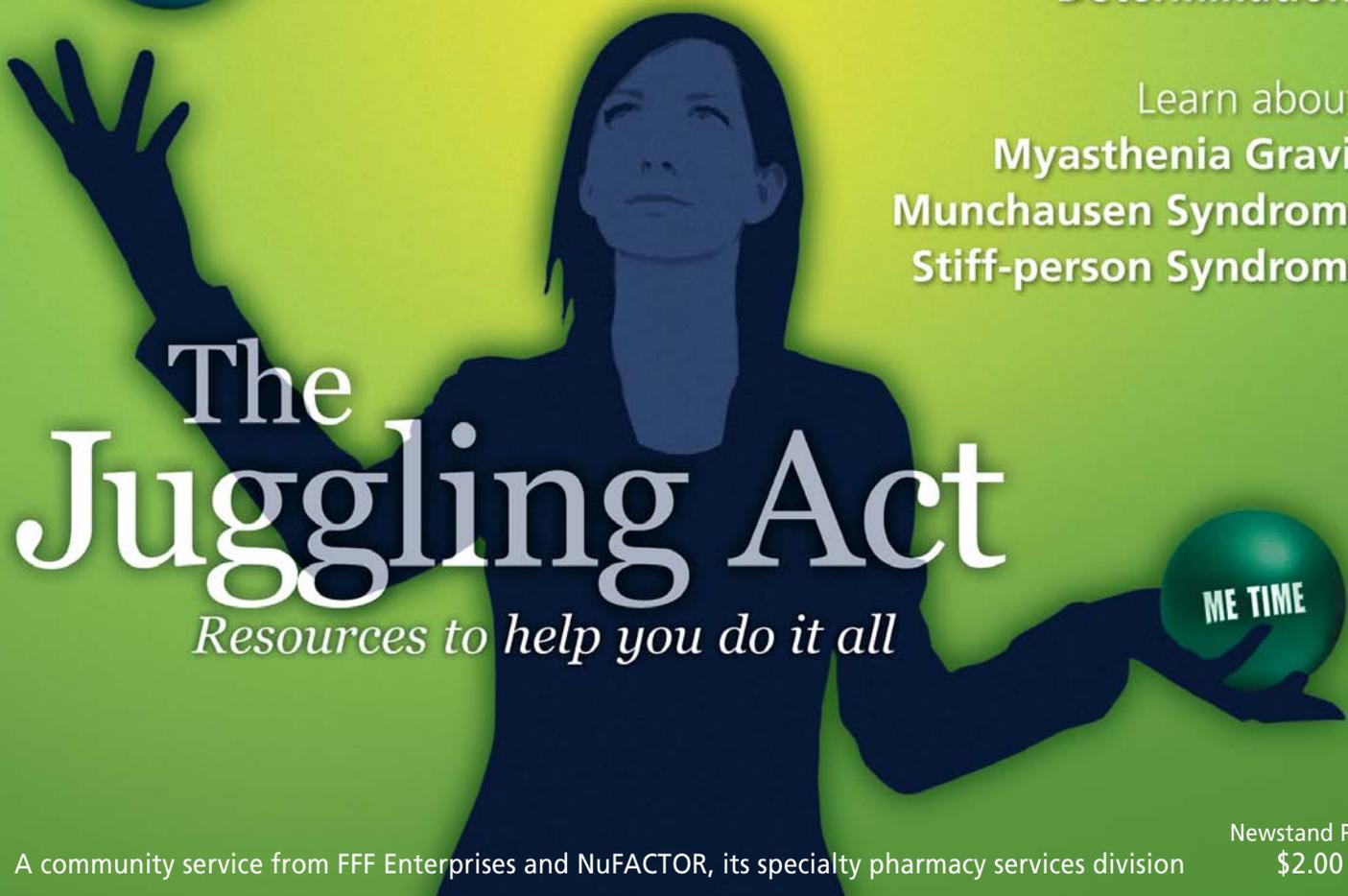
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April-May 2007



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Medicare Local Coverage  
Determinations

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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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# Fair IVIG Reimbursement for All Patients in All Sites of Care



Sounds like a rational, reasonable goal, right? Fair IVIG reimbursement for all patients for whom IVIG is proven efficacious in all sites of care? Sure.

Well, you might be surprised to hear that it's actually a little controversial. Who'd have thunk it!

As the IVIG reimbursement crisis created by the Medicare Modernization Act has persisted since January 2005, various groups concerned with this issue have staked out various positions on various solutions.

The result is a significant void: There is no single, unified voice speaking for the patients, saying, "Hey, guys and gals, here's what will work for the patients, for all of them."

Oh, sure, patient organizations have advocated with dedication for their constituents. Many providers have hung in there in what has proven a protracted battle. And manufacturers also have advocates working the issue.

But, while these groups have been advocating doggedly, their voices are lost in the din of more familiar issues with larger, significantly louder followings.

Divided we do indeed fall. So, how about trying united?

Imagine this: What might happen if all these individual voices were unified in the powerful, passionate surge of a masterful voice, a voice heard far and wide for its common vision, its cohesive consensus on how to fix this problem, its compassion for all patients.

Imagine then, what might happen if the 50 or 60 legislators who have embraced this cause heard that singular voice. Imagine the numbers to which they would grow! Imagine the solution they would achieve!

This image is why we are so touched and humbled by the thousands of readers who've shared their stories with us. Because they continue to hope.

This is why we want to document, through a patient registry of all IVIG users, true demand for this precious product. So manufacturers can effectively plan.

This is why we are so eager for a national alliance representing all patient groups. Because united we do indeed stand.

Because united, patients will not fall. Instead, they will thrive and go to school and have families and work and live and love.

Because, when it comes right down to it, wouldn't you rather be singing the "Ode to Joy" than the "Worried Life Blues"?

## In This Issue

We are disappointed to have to draw your attention to this issue's reimbursement article beginning on page 9, "Medicare Local Coverage Determinations Limit Access to IVIG." The article introduces a disturbing trend in local IVIG reimbursement decisions that we will continue to report on in subsequent issues. Like the overall reimbursement crisis, this one affects patients across all disease states. So, although the material is a bit dry, it may affect you or someone you love, so please take a look.

We are very pleased to introduce a new feature, "Ask Kris." A member of a patient community, mother to two adult sons with PIDD, patient advocate, and employee of our sponsoring homecare company, Kris has a broad and intimate perspective on issues related to living with a chronic disease and IG therapy—and she also knows how to dig deep to mine answers to your questions. So, send them on, to [editor@igliving.com](mailto:editor@igliving.com).

On a bittersweet note, we must say farewell and thank you to Dr. Jordan Orange, a dedicated immunologist and patient advocate. His service as an IG Living Advisory Board member was exemplary. We will miss his sage counsel. Thank you, Dr. Orange! ■

A handwritten signature in black ink, appearing to be 'Kit-Bacon Gressitt'.

Kit-Bacon Gressitt, Editor

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Please send your letters to the editor to [editor@igliving.com](mailto:editor@igliving.com).

# A Lesson in CVID

By Alma Morales

If you mention “travel soccer” to Ashton Eisenhut, her eyes light up. She began playing recreational soccer when she was 6. Now 13, Ashton finally feels strong and healthy enough to play competitive travel soccer for her community league. She looks forward to playing midfielder, a demanding position that requires both defensive and offensive skills. “Our games started in February, and we will be going to tournaments,” says Ashton with enthusiasm.

Her quest to regain her health was a frustrating one. At countless visits to her pediatrician, she was continuously diagnosed with allergies and administered allergy shots on a regular basis. When she was 11, the right test finally revealed that Ashton has low immunoglobulin levels, and she was diagnosed with common variable immune deficiency (CVID). “I was shocked and, at the same time, sort of relieved,” explains Ashton.

Being diagnosed with CVID at the age of 11 was difficult for Ashton. “The most challenging part about having CVID is getting used to it—and having [immune globulin (IG)] infusions. The hardest thing is accepting the port,” explains Ashton.

Nancy Lloyd, Ashton’s mother, tells how they have learned to manage CVID. “The psychologist helped a lot,” says Nancy. “She has a great doctor who monitors her closely. We concentrate on keeping everything as normal as possible.”

Prior to her diagnosis, Ashton was enrolled in private school, but when the financial strain on her family increased due to the costs of her IG treatments, they decided to homeschool her. “I like homeschooling because I can do things on my own,” says Ashton.

Nancy explains, “We watch DVDs of the teacher teaching the curriculum. We study every subject, every day. The great thing about homeschooling is that it’s very flexible.”

Ashton receives her infusions at home as well. She uses a port for her biweekly treatments because regular infusions were too painful for her. “Now, we use numbing cream and I lay on the couch during my infusion,” explains Ashton. And, she is getting accus-

tomed to her routine treatments, often preparing her infusions on her own. “I pull out my IG from the refrigerator and set up all of the things I will need.”

One aspect of homecare therapy that Ashton really enjoys is the close relationship she has built with her homecare nurse, whom she calls Miss Lisa. “She’s fun; we play games and do crafts during my treatments. It helps pass the time.” Ashton has such a great bond with her nurse that she was invited to be a part of Miss Lisa’s wedding party.

Recently, Ashton has taken on the role of advocate by teaching others about CVID. When approached by her homecare company to participate with her nurse in a billboard advertisement that would be displayed in her hometown, Ashton was very excited. “The company wanted to build awareness of CVID because there aren’t many people in my region with it. We would be at Wal-Mart or Target and people would ask me if I was the girl in the billboard.”

“It was cool!” Ashton exclaims, as she recounts her experience of creating the ad. “The fun part was doing the photos.” But it had its serious side, too.

Her knowledge of CVID and primary immune deficiencies has not only allowed Ashton and her family to teach others about it, it has also resulted in helping a neighbor be properly diagnosed with a peripheral neuropathy.

Although Ashton is only in eighth grade, she thinks about her future. “I plan to go to college in North Carolina.” She would like to study microbiology and genetics. “My grandfather had myasthenia gravis. He was diagnosed with it when he was older. There has been some genetic research that shows primary immune deficiencies may be genetic.”

Through all of the struggles of dealing with CVID, Ashton has managed to see the silver lining. “The most rewarding part is knowing I can overcome it,” says Ashton optimistically. With her positive attitude and inquisitive nature, Ashton is preparing for a bright future. ■

*Note: Some physicians discourage the use of ports. Please always discuss treatment options with your doctor.*



## Catch the Spirit of IDF in St. Louis

From the Immune Deficiency Foundation

Mark your calendars for the 2007 Immune Deficiency Foundation (IDF) National Conference, Catch the Spirit of IDF, on June 28–30, 2007, in the heart of St. Louis, Mo.

It's a biennial event with programs for everyone—patients, their families and friends, and healthcare professionals alike.

An Introductory Workshop will provide an overview of the immune system and primary immune deficiency

diseases. Newly diagnosed families, first-time conferees and anyone who needs a refresher course are encouraged to attend.

Twenty-five world-renowned immunologists will share their expertise. Specific diagnosis sessions will provide individuals and families with a better understanding of the disease, treatment options, research advances, and dealing with their particular disease.

Additional scientific sessions will provide in-depth information on specific issues related to diagnosis, treatment and management.

For young people, ages 6 to 17 years, the Youth Program offers age-appropriate educational programs and a trip to the St. Louis Science Center. And for the little ones, onsite childcare for children ages 6 months to 5 years will be available at the hotel.

Life Management sessions will cover an array of subjects: from health insurance to school issues; from family dynamics to travel considerations. There will be plenty of time for questions and answers at all these gatherings. Be sure to check out a complete list of all sessions in the online registration brochure on the IDF website at [www.primaryimmune.org](http://www.primaryimmune.org).

A Professional Education Program will be held on Friday afternoon. The program is designed for immunologists, allergists, primary care physicians, family practitioners,

## in St. Louis



pediatricians and nurses. The sessions include Introduction to Primary Immune Deficiency Disease; Use of Laboratory Tests in Diagnosis of Primary Immune Deficiency Disease; New Developments in the Understanding of Primary Immune Deficiency Disease; Treatment of Primary Immune Deficiency Disease Through Bone Marrow Transplantation and Gene Therapy; and Immune Globulin Therapies.

And it isn't all work—there will be plenty of opportunities to renew old friendships, meet new friends, share ideas and have fun. Thursday will begin with a Welcome Reception for all conference attendees in the historic Crystal Ballroom in the Renaissance Grand Hotel. On Friday night, come Catch the Spirit of IDF! with St. Louis blues music, dining and a celebration of the IDF community. The IDF Family Night Zoobelation on Saturday offers a tour of the No. 1 zoo in the country, the Zooline Railroad and Conservation Carousel, and a barbecue dinner and entertainment.

IDF is grateful to its generous sponsors, Baxter Healthcare Corporation, CSL Behring, Grifols and Talecris Biotherapeutics, for their support in making this exciting conference possible. ■

### Comments from 2005 IDF National Conference Participants

*"It was great, especially helpful to recently diagnosed persons."*

*"It was an amazing experience. The IDF is a lifesaving organization."*

*"This was put together just beautifully. I have never felt so relaxed and yet learned a great deal in one weekend."*

For more information and to register, visit [www.primaryimmune.org](http://www.primaryimmune.org) or contact IDF at 800-296-4433.

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*"Service is high quality and the best I have dealt with since I started taking IVIG 13 years ago."*



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Many IG Living readers are contacting the magazine to report problems acquiring their intravenous immune globulin (IVIG) treatments, and the reasons they report are varying:

- Patients' IVIG dosing is being reduced at hospitals by an average of 50%, contrary to physicians' orders.
- Medicare will not cover their IVIG therapy.
- Their trough levels are too high to warrant IVIG.
- Hospitals are requiring patients to sign documents (Advance Beneficiary Notices or ABNs) indicating that they will assume financial responsibility for their IVIG treatments, in the event that Medicare refuses to cover them.
- Their IVIG treatment is being terminated temporarily until medical necessity is determined.

Our reimbursement consultants have investigated these cases and discovered that the majority of Medicare carriers (these are private insurance companies that implement Medicare benefits at the local level) are implementing "Local Coverage Determinations"<sup>1</sup> that are reducing patient access to IVIG therapy in states across the country. These determinations vary by state and carrier, and are not based on accepted medical guidelines for treatment and dosing.

According to Jordan Orange, MD, PhD, assistant professor of pediatrics at the University of Pennsylvania School of Medicine and attending physician in immunology at Children's Hospital of Philadelphia, "The randomness that underlies the differences in these determinations is truly idiosyncratic and unscientific, and thus the criteria are detrimental to patient care."

We encourage you to read this article, become informed about Local Coverage Determinations, learn how to appeal such determinations, and advocate against this challenge to quality patient care.

<sup>1</sup> Additionally, many private insurance companies have adopted the Milliman Care Guidelines, which have limitations in coverage similar to Local Coverage Determinations and seem to be outdated, arbitrarily applied, and not necessarily based on sound medical evidence. In a subsequent article, we will review the Milliman Care Guidelines.

# Medicare Local Coverage Determinations Limit Access to IVIG

By Melissa Schweitzer and Michelle Vogel

## Local Coverage Determinations

Although Medicare is a federal program, it contracts with private insurance companies (carriers) and fiscal intermediaries (FIs) to pay healthcare bills at the local level. Each Medicare carrier and FI has the discretion to determine which services are reasonable and necessary and, therefore, covered as a Medicare benefit. These coverage policies are issued in a document called a Local Coverage Determination (LCD).

LCDs are drafted by individual Medicare carriers in individual states; consequently, there are distinct differences in them, carrier-to-carrier and state-to-state, rather than having a single set of coverage guidelines defined at the federal level. This creates confusion about coverage and payment policies for providers and patients. Also of concern, the soundness of the individual LCD criteria depends on the capabilities of the local Medicare carrier's medical director and the expertise of the Carrier Advisory Committee members, who provide insurance claims and medical determination review and advice. Clinical expertise is essential when determinations involve conditions that are rare or otherwise difficult to diagnose and/or difficult to treat.

In the February-March 2007 IG Living, Reimbursement Q&A section, we briefly discussed a Rhode Island case in which Linda, a Medicare patient with a primary immune deficiency disease (PIDD), was having trouble accessing her IVIG treatment due to an LCD denial, despite PIDD being an on-label use for IVIG. Specifically, her LCD stated the following reasons in its explanation of the denial of IVIG coverage:

*Documentation provided does not adequately support the medical need for the services rendered. Medicare guidelines require that the services provided are supported by a covered diagnosis that would justify the medical need for treatment of the patient's illness or injury. The medical documentation submitted indicates the laboratory level is 1030; dosage to be adjusted to keep levels between 400–600. There was no evidence of infection or dosage adjustment. We will therefore hold the provider liable.*

Linda's provider, the hospital where she had been regularly receiving her infusions prior to the LCD, was suddenly refusing to treat her unless she signed an Advance Beneficiary Notice.

An Advance Beneficiary Notice (ABN) is a document that providers can require a Medicare beneficiary to sign. The ABN holds the beneficiary responsible for the cost of the treatment, if Medicare does not cover it.

In Linda's case, if she signed the ABN, she would be assuming financial ➤

responsibility for the cost of her IVIG infusions, because the LCD had already indicated Medicare would not cover them. However, Linda is appealing the Local Coverage Determination, so she refused to sign the ABN, to avoid becoming responsible for the cost of her treatments if her appeal fails. Consequently, Linda is going without treatment for her PIDD, and her health is deteriorating.

It is important to note some key elements in the explanation of the carrier's decision: Even though Linda's physician prescribed IVIG to be administered on a monthly basis, and even though Linda's PIDD is normally covered by Medicare, the local carrier can deny coverage, because Linda's serum trough levels do not meet the carrier's lower trough level guidelines.

Linda was prescribed the number of grams of IVIG and the frequency of treatment necessary to keep her as close to infection free as reasonably possible. Her local Medicare carrier is requesting that she have infections in order to be eligible for IVIG, instead of preventing the infections.

In another case, Carol, a Georgia resident, contacted IG Living for assistance. She had been admitted to a hospital and diagnosed with chronic inflammatory demyelinating polyneuropathy (CIDP), normally

a covered indication for IVIG. She received her first infusions as an inpatient, and did well. Her physician subsequently prescribed IVIG infusions in the hospital's outpatient clinic. Carol went to the clinic and was told that Medicare did not cover IVIG.

According to Carol's carrier's LCD:

*IVIG is not covered as an initial therapy for patients with newly diagnosed CIDP nor as a maintenance therapy in patients failing to respond to an initial course of IVIG following therapies with other agents; exception to this rule would be in patients with severe CIDP and in patients who have contraindications for immunosuppressive drugs.*

Carol has not been able to receive her IVIG, and her condition continues to deteriorate even though she showed improvement when she received her infusion as an inpatient. Unfortunately, it is likely that Carol will indeed be determined to be eligible for IVIG by the time this article is published, because her health is deteriorating to the point that she will be admitted to the hospital as an inpatient.

## The Effect of LCDs Across the Country

The following chart shows examples of current provisions by local Medicare contractors that may limit coverage for IVIG.

Primary Immune Deficiency Diseases (PIDD)	
Limitation of Coverage	States Affected
The following limitations are made on patient serum IgG trough levels to qualify for IVIG coverage: <ul style="list-style-type: none"> <li>Serum IgG trough levels must fall within the range of 400-600 mg/dl; this applies to any individual with a primary immune deficiency disease, with no specification on the particular type of PIDD</li> <li>Serum IgG trough levels must be tested every 3 months.</li> </ul>	AK, AL, AZ, AR, CA, CO, CT, DE, FL, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NY, NC, ND, NE, NH, NJ, NM, NV, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY
The following limitations are made on dosage of IVIG: <ul style="list-style-type: none"> <li>IVIG loading dose ranged from 100 mg/kg to 600 mg/kg body weight and maintenance doses ranged from 100 mg/kg to 400 mg/kg body weight administered approximately once per month by intravenous infusion. Maintenance doses exceeding 200 mg/kg may be covered, but only if the desired clinical response to 200 mg/kg is clearly documented as inadequate.</li> </ul>	AK, AL, AZ, CA, CO, CT, DE, FL, GA, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MA, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NV, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY
After a period of 1–2 years and at similar intervals thereafter, attempt must be made to stop IVIG infusion to identify specific immunochemical abnormality; requires periodic monitoring to justify need for continued infusion.	KS, NE
Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)	
Limitation of Coverage	States Affected
IVIG not covered as initial therapy for patients with newly diagnosed CIDP nor as maintenance therapy in patients failing to respond to an initial course of IVIG following therapies with other agents; exception to this rule would be in patients with severe CIDP (Rankin scores of 4 or 5) in whom rapid therapeutic response is deemed medically desirable and in patients who have contraindications for immunosuppressive drugs.	CO, DC, DE, GA, ID, MT, NC, NJ, NM, TN, TX, VA
Covered if intolerant of prednisone or azathioprine over at least 3 months or neurologic function assessment score of at least 3 or greater on Rankin Scale. Covered when difficulty with venous access for plasmapheresis or other therapy has failed or is contraindicated, or for rapidly progressive forms.	AK, AL, AZ, CA, CO, CT, DE, FL, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NY, NV, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY, Puerto Rico, and Virgin Islands
Dosage guidelines: Initial therapy 400 mg/kg body weight for 5 days. Maintenance therapy 250–400 mg/kg body weight no more frequently than every 2 weeks.	AK, AL, AZ, CA, CO, CT, DE, FL, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NV, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY
An attempt must be made to wean the dosage when improvement has occurred. Must be an attempt to stop IVIG if improvement is sustained with dosage reduction.	AZ, AR, KS, LA, MO, MT, NE, NM, ND, OK, RI, SD, UT, WY

## Guillain-Barré Syndrome (GBS)

Limitation of Coverage	States Affected
IVIg therapy is covered as equivalent to plasmapheresis for the treatment of GBS if patient has paralysis that precludes from walking 30 feet without assistance and is within the first 2 weeks of illness.	AK, AL, AZ, CA, CO, CT, DE, FL, GA, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NV, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY
Covered when difficulty with venous access for plasmapheresis or other therapy has failed or is contraindicated, or for rapidly progressive forms.	CT, IL, IN, KY, NY, OH, Puerto Rico, Virgin Islands
At 6 month intervals after therapy has begun, the following elements must be documented in medical records for chronic use of IVIG: clinical summary of functional status, a recent physical exam, copies of objective tests including nerve conduction studies and pulmonary function tests, an assessment of long-term prognosis and anticipated future treatment courses.	CA, MA, ME, NH, VT
Dosage guidelines: 1000 mg/kg body weight daily for 2 days or 400 mg/kg body weight daily for 5 days. In some states, dosage can only be repeated if evidence of continued clinical progression after initial infusion is completed, but not if patient has ongoing, but stable weakness.	AK, AL, AZ, CA, CO, CT, DE, FL, GA, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NV, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY

## Myositis

Limitation of Coverage	States Affected
Individual coverage determination required for coverage of IVIG in dermatomyositis (DM), polymyositis (PM), and inclusive body myositis (IBM); also includes multifocal motor neuropathy (MMN) and Lambert-Eaton in this category.	AK, AL, AZ, CA, CO, CT, DE, FL, GA, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NV, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY
DM is covered when difficulty with venous access for plasmapheresis or other therapy has failed or is contraindicated (second-line agent) and for rapidly progressive forms.	AL, CT, IL, IN, IA, KY, NY, OH, SD
IVIg covered for PM and DM if the following criteria are met: <ul style="list-style-type: none"> <li>• Biopsy-proven disease</li> <li>• Have had at least a 4–6 month trial of prednisone or prednisone combination therapies</li> <li>• May be covered at less than 4 month trial if profound, rapidly progressive and/or potentially life-threatening muscular weakness and is refractory or intolerant to previous therapy</li> <li>• Lack of response to other therapies as reflected by persistently elevated CK (creatinine kinase) levels and/or lack of improvement on muscle strength improvement scales.</li> </ul>	CO, DC, DE, GA, ID, MT, NC, NM, TN, TX, VA
Patient's record must show that there was a measurable response within 6 months of use of IVIG or its use will no longer be considered medically necessary.	CT, NY
Dosage guidelines: Suggested doses of IVIG range from 1000 mg/kg body weight daily for 2 days every 4 weeks or 400 mg/kg body weight for 5 days every 4 weeks in patients intolerant of high dose therapy, to 2000 mg/kg, as a common initial empirical dose, with the division of the dosage left up to the physician.	AR, GA, ID, IL, IN, KS, MO, NC, NE

## Myasthenia Gravis

Limitation of Coverage	States Affected
IVIg is covered when difficulty with venous access for plasmapheresis, if patient is intolerant of or refractory to cholinesterase inhibitors, corticosteroids and azathioprine, and/or if patient has profound rapidly progressive and/or potentially life-threatening muscular weakness.	AK, AL, AZ, AR, CA, CO, CT, DE, FL, GA, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NY, NV, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY, Puerto Rico, Virgin Islands
Hospitalization is required for coverage. IVIG can be used in patients with chronic, severe, MG with a vital capacity of less than 1 litre, dysphagia associated with aspiration and inability to ambulate 100 ft without assistance. Documentation must be submitted after 2 treatments and should record significant improvement in order for continued coverage to be allowed.	LA, MO, MS; MA, ME, NH, VT: Also states that documentation of medical necessity and efficacy of treatment must be provided at each 6-month interval after treatment has started and include: clinical summary of functional status, physical exam, copies of objective tests, and assessment of long-term prognosis and anticipated future treatment courses
Dosage guidelines: No clearly established regimen, although studies have reported success with dose of 400 mg/kg body weight per day for 5 days.	AK, AL, AZ, CA, CO, CT, DE, FL, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NV, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY

Idiopathic Thrombocytopenic Purpura (ITP)	
Limitation of Coverage	States Affected
<p>For acute ITP, IVIG can be recommended in the following situations:</p> <ul style="list-style-type: none"> <li>• Management of acute bleeding due to severe thrombocytopenia (platelet counts less than 30,000/mm<sup>3</sup>)</li> <li>• To increase platelet counts prior to invasive major surgical procedures (splenectomy)</li> <li>• In patients with severe thrombocytopenia (platelet counts less than 20,000/mm<sup>3</sup>) considered to be at risk for intracerebral hemorrhage.</li> </ul> <p>For chronic refractory ITP, IVIG can be recommended in the following situations:</p> <ul style="list-style-type: none"> <li>• Prior treatment with corticosteroids and splenectomy</li> <li>• Age of 10 years or older</li> <li>• Platelet counts persistently at or below 20,000/mm<sup>3</sup></li> <li>• Duration of illness less than 6 months</li> <li>• No concurrent illness/disease explaining thrombocytopenia</li> </ul>	AL, AK, AR, AZ, CA, CO, CT, DC, DE, FL, GA, HI, IA, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NV, NY, OH, OK, OR, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY
<p>For ITP in pregnancy, IVIG can be recommended for the following:</p> <ol style="list-style-type: none"> <li>1. Pregnant women who have previously delivered infants with autoimmune thrombocytopenia</li> <li>2. Pregnant women who have platelet counts less than 75,000/mm<sup>3</sup> during the current pregnancy</li> <li>3. Pregnant women with past history of splenectomy.</li> </ol>	AR, AZ, GA, IL, IN, KY, LA, MO, MT, ND, NJ, NM, OH, OK, RI, SD, TN, UT, WY
<p>There is good scientific evidence that supports this use in a few of the neurological disorders; in others, however, the evidence is either poor or lacking. The literature shows that not all patients with the following diseases need treatment with IVIG:</p> <ul style="list-style-type: none"> <li>• Acute and chronic inflammatory demyelinating polyradiculoneuropathy</li> <li>• Guillain-Barré syndrome</li> <li>• Immune Thrombocytopenic Purpura in pregnancy</li> <li>• Myasthenia gravis</li> </ul> <p>When administered for the above conditions, the medical record must document one of the following situations to constitute appropriate IVIG infusions:</p> <ul style="list-style-type: none"> <li>• Other therapy has failed or is contraindicated</li> <li>• Rapidly progressive forms of these diseases</li> </ul> <p>The beneficiary is not liable for services denied as not reasonable and necessary unless a valid waiver of liability (advance beneficiary notice or ABN) has been signed.</p>	AZ
<p>Reimbursement can be allowed for chronic use as long as the drug is effective in the immunodeficiency states. The usual treatment for ITP should not exceed 90 days.</p> <p>Dosage should range between 100 to 500 mg/kg, I.V., every month.</p>	LA, MO, MS
<p>Dosage guidelines for Acute ITP:</p> <ol style="list-style-type: none"> <li>a. 1,000 mg/kg body weight given on 1 or 2 consecutive days</li> <li>b. 400 mg/kg body weight given on each of 2–5 consecutive days.</li> </ol> <p>Dosage for chronic refractory ITP:</p> <ol style="list-style-type: none"> <li>a. Initial—1 or 2g/kg body weight (total cumulative dose) given in equal amounts over 2–5 days</li> <li>b. Maintenance—800–1,000 mg/kg body weight administered no more frequently than every 2–6 weeks as determined by serial platelet counts</li> </ol>	AK, AL, AR, AZ, CA, CO, CT, DE, FL, GA, HI, ID, IL, IN, KS, KY, LA, MA, MD, ME, MI, MN, MO, MS, MT, NC, ND, NE, NH, NJ, NM, NV, NY, OH, OK, PA, RI, SC, SD, TN, TX, UT, VA, VT, WA, WI, WV, WY

## Appealing LCDs

If your IVIG therapy is denied based on a Medicare LCD, you probably have the right to appeal the denial—and you should.

You can appeal the LCD denial, if you meet the following criteria:

1. If you are entitled to benefits under Medicare Part A, are enrolled under Medicare Part B, or both.
2. If your physician says that you need the denied item(s) or service(s) or if you have already received the item(s) or service(s).

You will know if your IVIG treatment coverage has been denied if you receive a Medicare Summary Notice (MSN), including a Notice of Denial. An MSN indicating a denial will include the following:

1. Explanation of Benefits (EOB): The EOB explains the services you received; the amount billed; any amount covered; and what you owe.
2. Notice of Denial: This explains what service(s) was denied; why it was denied; how much time you have to appeal the denial; and how to file the appeal.
3. Local Coverage Determination: The LCD explains what limitations your local carrier imposes on coverage for your IVIG therapy for your disease state (along with all other disease states for which IVIG coverage is available).

## When can you file an LCD appeal?

If you have not yet received the item or service, you must file your appeal request within six months of the date of the treating physician's written statement that you need the item or service.

If you have already received the item or service, you must file your appeal request within 120 days of the date of the initial denial notice.

Mail your LCD appeal request to:  
 Departmental Appeals Board  
 Civil Remedies Division, Mail Stop 6132  
 Cohen Building, Room G-644  
 330 Independence Avenue, S.W.  
 Washington, DC 20201

## What information must you include in your LCD appeal request?

When you file an LCD appeal request, include:

1. Name of the Medicare beneficiary
2. His or her address
3. Telephone number
4. Health insurance claim number, if applicable

## Current Medical Guidelines

To understand the differences between the LCD guidelines and quality patient care, it is helpful to review current medical guidelines for IVIG therapy by disease state.

Primary Immune Deficiency Diseases <sup>2</sup>		
Dosage Guidelines	Current Medical Guidelines on Serum IgG Trough Levels	
<p>According to American Academy of Asthma, Allergy and Immunology (AAAAI), the usual initial dose of IVIG for antibody replacement is 400–600 mg/kg delivered every 3–4 weeks. However, this may vary with the particular PIDD in question, i.e., patients with extremely low IgG levels at presentation may benefit from a larger loading dose before the initiation of regular maintenance dosing. AAAAI states that an acceptable starting point for maintenance dosing is 400 mg/kg for many patients, which should be adjusted to achieve the optimal clinical result.</p>	<p>According to the AAAAI, the trough level for patients with agammaglobulinemia should be at least greater than 500 mg/dl and preferably greater than 800 mg/dL. Trough levels for patients with common variable immune deficiency disease (CVID) should be equal to pretreatment levels of IgG plus 300 mg/dl. Higher trough levels (&gt;800 mg/dl) have the potential of improving pulmonary outcome. Additionally, IgG trough levels can be unreliable in patients with CVID and should not be used as primary benchmarks for guiding therapy.</p> <p>AAAAI recommends monitoring trough levels at no greater than every 3 months and preferably no greater than every 6 months.</p>	
Neurological Disorders <sup>3</sup>		
Dosage Guidelines		
<p>A standard loading dose of 2 g/kg is usually administered over several days, depending on tolerance and convenience. It is then followed by booster doses of 0.5 to 1 g/kg every 2 weeks, or 1 to 2/kg every month, for a total of 2 to 3 months, at which time the neuropathy is re-evaluated. Improvement may be seen at 2 weeks to 3 months after beginning therapy, but if there is no improvement in 2 to 3 months, the IVIG is discontinued. Maintenance therapy is usually continued until there is maximal improvement, and then discontinued or tapered to see if it is still needed. In approximately one-third of cases, the disease is monophasic, and the improvement persists. In the other two-thirds, however, the disease relapses so that maintenance therapy is needed. The dose is then adjusted as needed. In such cases, the IVIG therapy can be considered a holding action, until a cure is found.</p>		
Idiopathic (Autoimmune) Thrombocytopenia Purpura		
Dosage Guidelines	Current Medical Guidelines	
<p>Recommend initial dose of 1.0 g/kg for 1–2 days</p>	<p><b>Use of IVIG in pregnancy:</b><sup>4</sup> Generally, corticosteroids are used as the initial therapy in ITP, but this can induce or exacerbate gestational diabetes, bone loss, hypertension, and abruption and prematurity. Therefore, in pregnancy, IVIG together with low-dose prednisone is the treatment of choice. Splenectomy should be avoided if possible to avoid abortion.</p>	<p><b>Use of IVIG in Rh-patients:</b><sup>5</sup> IVIG is the preferred treatment for Rh- ITP patients, and anti-D represents a frontline treatment option for Rh+ ITP patients.</p>

5. Title of the LCD that is being challenged
6. Specific provision(s) of the LCD affecting the Medicare beneficiary
7. Name of the private company (carrier or FI) that used the LCD

In addition to this, the request should explain:

1. What item or service is needed
2. Why the LCD is incorrect
3. Why the appeal request is being made

Include with your LCD appeal request a letter from the doctor treating you that states:

1. Item or service that you need
2. Any medical literature that supports the treatment of IVIG for your diagnosis, and any treatment guidelines available.

## Now, It's Time for Action!

It is critically important to keep your elected representatives informed of your LCD appeal—and to seek their help. Send them copies of your appeal packet, including a cover letter asking the representatives to support your appeal. Then follow up with phone calls to them. Remember to keep copies of everything for yourself!

The more frequently patients report such problems, the more likely the negative effects of LCDs on patient care will be reduced. Like the rest of us, many of our representatives are unaware of LCDs, and they would be shocked to know how LCDs inhibit quality patient care.

To find your elected representatives' contact information and to download a sample cover letter, visit [www.igliving.com](http://www.igliving.com) and click on Take Action.

If you have a question or need assistance, please email us at [editor@igliving.com](mailto:editor@igliving.com). 📧

<sup>2</sup> *The Journal of Allergy and Clinical Immunology*, Volume 117, Number 4, April 2006: *Use of Intravenous Immunoglobulin in Human Disease: A Review of Evidence by Members of the Primary Immunodeficiency Committee of the American Academy of Allergy, Asthma and Immunology.*

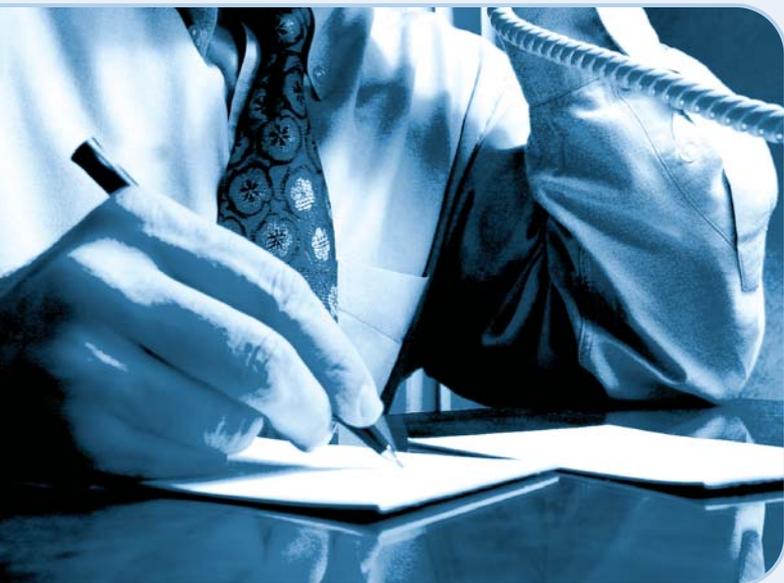
<sup>3</sup> *American Academy of Neurology Press, Demos Publications, New York 2006: Peripheral Neuropathy; When the numbness, weakness and pain won't stop.* Norman Latov, MD, PhD.

<sup>4</sup> *Blood*, The American Society of Hematology; October 1, 2005; Volume 106, Number 7: *How I treat idiopathic thrombocytopenic purpura (ITP);* Douglas B. Cines and James B. Bussel.

<sup>5</sup> *B Journal of Thrombosis and Haemostasis 2005; Volume 3, Supplement 1: Anti-D Compared to IVIG for the Treatment of Immune Thrombocytopenic Purpura: A Systematic Review.* Bussel JB, Provan D, Sinclair CJ, Genereux MG.

# How to Fight Your Insurance Company When Coverage Is Denied

From the Neuropathy Action Foundation



**M**any patients find themselves having to do battle with their health insurance companies over denials of coverage. There are steps you can take to attempt to avoid health insurance denials before they occur, but, if that does not work, there are still ways to win the battle if you have to appeal a denial.

The key is organization:

1. Have all your paperwork in order.
2. Take detailed notes of your interactions with everyone in the process.
3. Know your coverage.

## Information Is Power

Information is power, and this is never truer than when battling a healthcare system. The winner may be the side with the better-organized, more-detailed information. The best bet when dealing with insurers is to minimize the risk of denial, and then, if one does come your way, to solve the problem in the early stages.

- Understand your health insurance policy thoroughly. Review it on a regular basis, and ensure that you know exactly what is covered and what is not covered.

If you have questions or do not understand any aspect of your coverage, call your insurance company and make them explain it in lay terms. Make sure you understand the exclusions and limitations of the policy—and the section on how to appeal.

- When receiving medical care, make sure your healthcare provider understands what is covered and what is not. Remember, doctors deal with many patients and their staff deal with many insurance companies. Do not assume they will remember the particulars of your situation.
- Take your policy provisions seriously. If it dictates that prior authorization is required, then do not receive care without obtaining that authorization. Assuming that the company will cover you and you can obtain coverage later, even if that is what your doctor tells you, could lead you into a world of bureaucratic nightmares, and it can lead to a denial of coverage.

## Avoid Denials of Care: Maintain Complete Records and Documentation

You are your own best advocate. You know best what ails you and what questions and concerns you have about your treatment. Take yourself seriously—be your own advocate at all times. Here are some steps to help avoid denials of care and coverage by your healthcare provider before they occur:

- Maintain an ongoing medication and infusion log, documenting all medications and treatments you are currently using, including dosage and frequency.
- Always try to bring another person to your doctor appointments. He or she can listen and take notes to help you remember what the doctor tells you. Even your own list of questions can fail you if you do not feel well or the questions are not addressed in the order you've written them.

- Create a file folder to keep all documents, logs, test results and medication lists so that all your pertinent health information is in one place. Save copies of all paperwork from your doctor and your insurance company. Keep these records in chronological order so you can find them easily.
- Maintain a detailed log of all healthcare services and communications (phone conversations, in person meetings, mail, email, etc.) that you have with your physicians, health insurance company and any other healthcare entity. This cannot be overstated. This log

will greatly benefit you should you ever encounter access issues for medications, treatments or procedures.

- If using an out-of-network provider, establish **before care is provided** that he or she will accept your health insurer's payment as payment in full.
- If there is a claim for which your insurance company will reimburse you only after you've paid your provider out of pocket, be sure to file the claim immediately.
- If there is a delay in payment, call your insurance company immediately.

## When Care Is Denied

Assuming you have taken all the above-mentioned steps and are still denied coverage, do the following:

- Review all the paperwork regarding the case immediately, making sure you understand every aspect. Then, with your paperwork in front of you, call your insurance company. Use the customer service number.
- The insurance company representative should be able to tell you why you were denied coverage. Make sure you take detailed notes of the conversation!
- Denial of coverage is often a result of administrative error. If this is the case, you may be able to resolve it on the first call or with just some minor communication thereafter.
- Assuming the problem continues, request an itemized bill from the doctor or hospital, and review every charge. There are often charges on these bills for services not delivered. If you find any, notify the doctor or hospital immediately to get the bill adjusted. Then notify your insurer.
- Often, however, the denial has been legitimately issued. The insurance company may not consider your medical procedure necessary, may consider it experimental or outside their coverage evidence-based guidelines. If this is the case, it is now time to take additional steps.
- Request a formal review by the insurance company. The customer service representative can tell you the specific procedures required. Then, state your case for appeal in writing, and send the letter via certified mail with return receipt requested. Make sure to do

this immediately. Some companies have time limits for appealing denials. Don't wait!

- If the insurance company claims that the cost of your care was above their customary cost, request the doctor's or surgeon's notes. They may show that there were mitigating circumstances in your case that justify that cost. Also, request any other information you need from your doctor to prove your case, and make sure you have it all in writing. Your doctor may also be able to provide a letter and other documentation validating your need for the challenged treatment.

### Contact the Appeals Entity in Your State

If you feel you are in over your head, your appeal is denied, or your insurer does not respond in a timely fashion, contact your state's department of insurance (in some states known as the department of managed care). Every state has different ways of assisting consumers with health insurance appeals. In addition, the Kaiser Family Foundation provides information on every state's healthcare rules ([www.kff.org](http://www.kff.org)).

Some states have an ombudsman who can provide detailed guidance through the process. Some have special offices for HMO issues. Some have only administrative assistance, taking the complaint and investigating. Your state's department of insurance will be able to tell you exactly how much assistance they can provide. Be sure to ask, and take advantage of all that is there. ■

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*Editor's note: Reprinted with the permission of the Neuropathy Action Foundation, [www.neuropathyaction.org](http://www.neuropathyaction.org).*

# Myasthenia Gravis and

By Jeff Siegel

When Lisa Langhals was diagnosed with myasthenia gravis (MG), she knew what treatments she didn't want. This meant no steroids and no immune suppressants, two of the most common therapies for MG. MG is a muscular disease characterized by weakness in the body's voluntary muscles, such as those that control eye movement and swallowing. Langhals, who had been participating in triathlons, first noticed blurry vision and some muscle weakness when she was competing.

"I wasn't sick otherwise, except that I had myasthenia gravis," says Langhals, who was diagnosed almost two years ago.

Langhals and her physicians decided to try intravenous immune globulin (IVIG)—although, for MG, it is an off-label use. The results have been more than encouraging.

"It was like being in a dark room and then turning on the light," says Langhals, who receives IVIG treatments every six weeks. "The first day or so, you don't feel well. But then, it's like it's all gone. The change is that drastic."

Langhals' experience isn't unusual, even though many physicians still regard IVIG as something to try only when nothing else works. More and more, myasthenia gravis patients are turning to IVIG when more traditional therapies aren't helping them.

Says Lynn Waltz, who was diagnosed with MG 16 years ago: "IVIG has been a blessing for a lot of us. I talk to people in my support group, and they don't know what they'd do without it."

## The Basics

As described by the National Institute of Neurological Disorders and Stroke (NINDS), "myasthenia gravis is a chronic autoimmune neuromuscular disease, characterized by varying degrees of weakness of the voluntary muscles of the body. Myasthenia gravis, from the Latin and Greek languages, means 'grave muscle weakness.' MG is characterized by muscle weakness that increases during activity but improves after resting. Certain muscles, like those that control eye and eyelid movement, facial expression,

chewing, talking and swallowing are often, but not always, involved in the disorder. The muscles that control breathing and neck and limb movements may also be affected."

Not surprisingly, it is often misdiagnosed or not diagnosed at all. Waltz went 16 years before her doctors figured it out, while another patient, Heywood Auerbach, was at one point diagnosed with sleep apnea. Langhals, meanwhile, was told her symptoms weren't MG, but depression. Waltz says this problem with diagnosis is one reason that she volunteers with the Maryland/D.C./Delaware chapter of the Myasthenia Gravis Foundation of America.

According to NINDS, "myasthenia gravis is caused by a defect in the transmission of nerve impulses to muscles. It occurs when normal communication between the nerve and muscle is interrupted at the neuromuscular junction—the place where nerve cells connect with the muscles they control. Normally, when impulses travel down the nerve, the nerve endings release a neurotransmitter substance called acetylcholine. Acetylcholine travels through the neuromuscular junction and binds to receptors, which generates a muscle contraction."

In MG, antibodies prevent the receptors from working, and can even destroy them. Somehow, these antibodies are produced by the immune system, which means the body is attacking itself. Typically, drugs used to treat MG include neostigmine and pyridostigmine, which help improve neuromuscular transmission and increase muscle strength. Immunosuppressive drugs such as prednisone, cyclosporine and azathioprine may also be used. These medications improve muscle strength by suppressing the production of the abnormal antibodies. In addition, some patients benefit from removing the thymus, which may help put the immune system back in balance. The other important therapy is plasmapheresis, a procedure that removes abnormal antibodies from the blood.

## Patient Testimonials

IVIG is not the first treatment of choice, despite some success in patients such as Langhals and Waltz, who

# IVIIG Therapy



receive IVIG about every four to six weeks. Interestingly, the treatment is more common in inpatient settings than outpatient, mostly because it's much less expensive than plasmapheresis.

"It's not clear that there is direct evidence to base support for its use in treatment of chronic MG," says Henry J. Kaminski, MD, who will become the chairman of the department of neurology and psychiatry at Saint Louis University in June. "Having said that, and I think all myasthenia gravis experts would agree, it's highly valuable to administer for chronic treatment."

And many MG patients who are treated with IVIG agree. "I really don't know why it works for me, but it works very well for me," says Auerbach, who has run the gamut of MG treatment—with most of the side effects, including weight gain and anemia—since being diagnosed in 1999. Currently, he receives IVIG about every three months. "I know some people aren't as successful, but I have confidence in it. The first time I had it, it was like magic. I didn't even know it was off-label, since it has worked very, very well for me."

So why isn't IVIG used more often? Physicians cite a variety of reasons:

- **Not approved first treatment of choice.** James M. Auberle, MD, of the Toledo Clinic in Toledo, Ohio, says, "Yes, it might be a reasonable thing to do with a lot of patients, and for a lot of autoimmune diseases that affect the neuromuscular system. But it's not the first thing many people think of."
- **Cost of IVIG.** IVIG is much more expensive than traditional treatments, although there is anecdotal evidence that IVIG can maintain some patients' health and functionality, which, in the long-term, represents a cost savings.
- **Side effects of IVIG.** Some patients, says Dr. Auberle, experience fever and chills that can be as bad as some of the side effects of other treatments. Even those who get significant relief, such as Langhals, report some side effects during and immediately after treatment, and there are some risks to long-term use of IVIG, such as neurodegeneration and renal failure.
- **Difficulty in acquiring IVIG.** "I think we would be using it more often if not for that," says Dr. Auberle. "When you can't get it, you have to use an alternative treatment."
- **Lack of awareness of IVIG.** Dr. Kaminski explains it is very rare that patients come to him and ask for IVIG for MG. Rather, he discusses it with them as one of the options for treatment. They're familiar with most of the traditional choices, but not IVIG.

## Prognosis for MG

According to NINDS, the prognosis for MG patients is good, with treatment. "They can expect to lead normal or nearly normal lives. Some cases of myasthenia gravis may go into remission temporarily, and muscle weakness may disappear so that medications can be discontinued."

"The most important thing in choosing a treatment is what's most effective for the patient," says Dr. Auberle. "Should Lisa have asked for [IVIIG]? Of course. What you have success with will lead you in a direction. The ultimate thing is that she has had success with it."

IVIIG therapy makes sense to many people with myasthenia gravis. "More and more, you're seeing MG patients getting it," says Waltz. "I think it's also part of getting MG diagnosed more quickly. The sooner they're diagnosed, the more it can help. IVIG gives me a needed boost, stops the slide I get. In that, it has been a lifesaver."

And that has been as welcome as it was unexpected. ■

## For Myasthenia Gravis Information

Visit the Myasthenia Gravis Foundation of America at [www.myasthenia.org](http://www.myasthenia.org).

Lynn Waltz can be contacted through the Maryland/D.C./Delaware chapter office at 410-432-6193 or 866-437-3761.

# Functional Foods and Health Claims: How to Make Good Choices

By Jessica Schulman, PhD, MPH, RD

For thousands of years, humans have used food and botanical sources as therapeutic remedies. Today, we know that diet plays a role in about half of the leading causes of death, including heart disease, certain types of cancer, stroke and diabetes, and research suggests that consumption of certain foods is linked to health more generally. As consumers take greater responsibility for their own well-being, and as the population ages, it makes sense that people look to their diet to improve their quality of life.

As the health claims associated with functional foods proliferate, the support for these claims has become a topic of increasing concern. How can the consumer figure out if a claim is realistic or not? The goals of this article are to explain how food manufacturers make health claims for their products and to provide some guidelines for making good choices with respect to functional foods.

## What Are Functional Foods?

Foods that provide health benefits beyond basic nutrition (e.g., vitamins and minerals) are referred to as “functional foods.” Some functional foods are whole foods that carry added health benefits. In other cases, manufacturers may add extra ingredients or modify a food in some way that allows for a “health claim” on the label or in advertising materials. As interest in the healing properties of diet has grown, selling foods that claim curative properties has become a profitable business, with sales currently in excess of \$25 billion per year and growing. As manufacturers chase these dollars, the public has been inundated with advertisements for foods that have been modified or supplemented in some way intended to reduce disease risk and improve health. Some of these additions are now commonplace: Iodine is added to salt to prevent goiter; and

vitamin D is added to milk to improve bone mineralization.

The first functional food claim recognized by the Food and Drug Administration (FDA) was the link between oat bran and reduced risk of heart disease, but many other functional food claims have never been tested systematically.

## What Is a Health Claim?

A “health claim” is any claim on a label (package or advertisement) that suggests a relationship between an ingredient in a food and a disease-related condition. Health claims can be explicit and clearly stated or implicit and unspoken. For example, an implied claim can appear as a third-party endorsement, such as “The American Heart Association recommends eating a variety of grain products to reduce heart disease.”

## Why Is There Concern About Certain Functional Food Claims?

Although functional foods have great potential to improve the health of the public, the recent history of functional foods has been marked by difficulties with regulatory compliance. When you use a medication, the product insert provides you with information about the drug and the prescription tells you how much to take and for what duration. In contrast, when you use a functional food, there is rarely a product insert or prescription and consumers are left to navigate their own way. This is frequently the case when herbal ingredients are added to functional foods, rather than being sold separately as dietary supplements. For example, the Center for Science in the Public Interest (CSPI) notes that labels on echinacea, when it is sold as a food supplement, warn users against taking it for an extended period of time. Other botanical



advertisements also caution consumers who live with weakened immune systems, autoimmune disorders or take medication. However, these warnings are hardly ever found on functional foods that contain these potentially harmful ingredients. (For this reason, an astute anesthesiologist or surgeon will always ask their patient if he or she uses any herbal ingredients—supplements or functional foods—before doing any procedures.) *Table 1* presents some examples of historic functional food health claims, which came under scrutiny in 2001. None of these products remain on the market.

To protect the public from potentially misleading health claims, the FDA, until the 1990s, prohibited manufacturers from claiming that a food could reduce risk of disease. If a label mentioned a disease, the food was considered “unregulated” and treated as an unapproved drug. A few years later, the FDA got into the business of regulating specific health claims. This worked well until food makers developed creative ways to sidestep regulations. For example, instead

of making a health claim about the benefits of a specific product, companies began associating their products with general claims about the structure or function of the body. So consumers started to see products reminding them that “calcium can build strong bones” or “fiber maintains bowel regularity.” The manufacturer was responsible for ensuring the accuracy and truthfulness of such statements.

About five years ago, consumer interest groups began to raise concerns about whether unsubstantiated claims about the health benefits of functional foods risked harming the public. For example, in 2001, CSPI and the Council of the Better Business Bureau urged the FDA to prohibit misleading health claims on herbal teas because these claims were not authorized for the botanical ingredients the teas contained. At that time, Arizona Rx Memory Mind Elixir contained ginkgo biloba, an ingredient not recognized as safe and one that has not been proven to improve memory. Yet, despite these concerns, the FDA was unable to enforce compliance and the tea claims continued for more than ➤

**Table 1: Historic Functional Food Health Claims**

Manufacturer	Product	Claim
Snapple	Gravity Echinacea	Provides "strength" to "maintain endurance and support the immune system."
Apple & Eve	"Tribal Tonics," Immune Boon, Echinacea	"This herbal tonic enhances immunity and bolsters the body's defense system."
Hansen's Beverage Co.	"Healthy Start": Immune•ox Juice echinacea purpurea	"[M]ay help stimulate the body's production of interferon, a cell protecting protein."

**Table 2: Selected Examples of Functional Foods With Potential Health Benefits**

Functional Food	Ingredient/Component	Health Benefit
Whole oat products, barley, certain yeasts and breakfast cereals	Beta glucan	May reduce risk of coronary heart disease (CHD) and some types of cancer; may contribute to maintenance of healthy blood glucose levels
Certain fortified margarines	Plant sterol and stanol esters	Reduces total and LDL cholesterol
Cold-water or fatty fish and their oils	Polyunsaturated fatty acids: Omega 3s, *Eicosapentaenoic Acid [EPA]/Docosahexaenoic Acid [DHA]	May reduce risk of CHD; may contribute to maintenance of mental and visual function

\*EPA and DHA are also found in breast milk.

one year. While that was happening, CSPI conducted a review that identified more than 100 functional foods making misleading or unsafe health claims. The General Accounting Office (GAO) also criticized the FDA for failing to protect consumers from the possible dangers of unproven ingredients.

Today, concerns about unsubstantiated health claims continue. New products that have raised concerns include, but are not limited to: (1) Enviga™, a carbonated drink from a Coca-Cola/Nestle partnership that claims that the ingredients (caffeine and EGCG from green tea) burn more calories than the drink provides; (2) Mars/Masterfoods CocoaVia®, a candy bar that claims to be heart healthy; and (3) DanActive™ Immunity, a dairy drink that Dannon claims will “strengthen your body’s defenses.”

Attorney Illene Ringel Heller, for the CSPI, points out that enforcement of existing regulations is challenging: “When the FDA has warned companies, such as the makers of Mars candy and Arizona Iced Tea, that they were violating the law, the firms largely ignored the agency and have continued to market their products,” (cited in CSPI Newsroom, 2006). Recognizing that some manufacturers have short-circuited the regulatory process, some worry that functional foods will become “about as dependable as 19th century snake oil.”

### Is There a Place for Functional Foods?

Despite these concerns, there is a place for functional foods in health promotion. Functional foods have been used for the prevention of osteoporosis, cardiovascular disease, anemia and neural tube defects, among other conditions, and there is growing interest in how they might be used to improve immune function. Scientists have begun to study the role of diet and nutrition in hyper-

sensitivity, atopic disease and food allergy. For those with health concerns, there may be real benefits of using functional foods (see *Table 2*).

At the same time, it is important to be selective and cautious. In a recent study, post-operative surgical patients were given a formula that contained arginine (an amino acid). Researchers found improvements in cellular immunity and recovery for those who were given the enhanced formula. However, the same amino acid that helped the surgical patients caused harm in other patients, leading the researchers to caution that the enhanced formula may increase risk of an inflammatory response and mortality among patients with underlying problems. Thus, the benefits for those who live with a primary immune deficiency, systemic inflammatory response syndromes or sepsis remain uncertain.

Even if food ingredients such as amino acids, probiotics, selenium, antioxidants, vitamins, etc. are shown to play a role in enhancing immune function and resistance to infection, the optimum intake level and recommended servings of functional foods have not been established for many disease-specific groups. For example, a few years ago, when studies suggested that diets high in soy decrease risk of heart disease, manufacturers of functional foods began in earnest to fortify food with soy isoflavones. Soon after, scientists reported that isoflavones actually increased risk among a unique disease-specific group—those with estrogen-dependent breast cancer.

In other words, when it comes to functional foods, one serving size does not fit all. The American Dietetic Association suggests that certain foods that have health benefits for some groups may be contraindicated for others. Increasing intake of whole-grain foods, for example, may reduce heart disease in adults but excess fiber may cause

malnutrition in growing infants. Clearly, there is more work to be done before scientists identify functional foods that can be recommended to the public at large. Unless the benefits and record of safety are known, nutrition goals should focus on preventing or treating nutritional deficiencies rather than on attempting to modulate disease.

### How Can One Make Informed Choices?

An important preliminary question that savvy consumers should always ask themselves about any health claim for functional foods is: "Is it too good to be true?" As with most things, if a claim sounds too good to be true, it is most likely misleading.

A second important question is whether a specific health claim is supported by any research. Addressing this question is actually a lot more complicated than it sounds, because some manufacturers conduct their own poorly designed studies as a way of justifying health claims for their products. *Table 3* contrasts some of the distinguishing features of quality scientific research and pseudoscientific research, both of which may be used to test functional food claims.

Use caution when deciphering food claims and always consult with your healthcare providers about any herb, dietary supplement or questionable ingredients that you are using, including herbal beverages.

### Summary

Technological advances in the food industry, in combination with clinical trials and governmental control, will eventually improve the credibility of nutrient content claims and the public's confidence in functional foods. Until more is known, however, consumers must understand that, although functional foods show promise to improve health, many ➤

**Table 3: Distinguishing Between Scientific Research and Pseudoscience**

"Gold Standard" Scientific Research	"Pseudo" Scientific Research
<b>Rigorous testing</b> Specific, conservative, replicated claims. Usually: (1) double blind, (2) random assignment, (3) control group with clinical trial.	<b>Misleading</b> Vague, exaggerated or unreplicated claims. Does not follow high standards of scientific research. No clinical trial in humans. Not objective.
<b>Theory driven</b> Claims are supported by an explicit process, or accepted theory, to explain the effect.	<b>No Theory</b> Lack of an explicit mechanism, or half-baked theory, to explain the effect or claim.
<b>Strong links</b> Statistically strong correlation coefficients or cause and effect study designs.	<b>Weak link</b> Provides indirect links and assumptions. Relies on weak associations.
<b>Universally accepted language</b> Definitions and operational terms are described.	<b>Tries to impress</b> Misuse of technical jargon to give claims with superficial trappings of science.
<b>Study is limited</b> Provides context for the results and conditions under which the claim applies.	<b>A perfect study</b> Does not address the limitations, such as under which conditions the claim applies.
<b>Balanced findings</b> Reviews current literature and considers other data that conflict with findings.	<b>Only positive findings</b> Suppresses or refuses to consider data that conflict with the claim.
<b>Freethinking is accepted</b> Burden of proof is on the researcher making the claim, not on the consumer.	<b>Skeptic is wrong</b> Burden of proof is on the consumer rather than the scientist or manufacturer.
<b>Science by peer review</b> Blinded review and revision prior to publication. Rigorous standards.	<b>Science by press conference</b> Evasion of peer review before publishing results. No standards.
<b>Disclosure</b> The methodology is provided in detail so that others can reproduce the claimed results.	<b>Secrecy</b> Details of the study design and methodology are hidden, forgotten, omitted.
<b>Reproducible</b> Experiments are repeated by scientists internationally with increasing precision.	<b>Casey</b> First study is often the last or fails to check procedures and correct errors over time.
<b>Problem solving</b> Conflicting evidence is discussed. Self-questioning. Logical reasoning.	<b>Ego driven</b> Attacks the character of those who question the study methods and the claims.

# Everyone Has a Story, and This Is Gini Lea's

By Carol K. Milette

**G**ini Lea Ennis, 32, bubbles over with enthusiasm because of her new lease on life. She spreads this good cheer everywhere she goes and to everyone she meets. The cause of this unmitigated joy? She has regained her health.

"I have CVID [common variable immune deficiency], but it doesn't have me!" Gini declares.

Life has not always been this good for Gini, though. She cannot remember being well as a child. Her younger sister, Jessica, was born with a rare liver disease, and for the 18 months that she lived, most of the family focus was on Jessica.

As a child, Gini spent years with severe, chronic upper respiratory infections. She was taken to see a specialist when she was 12. The doctor referred to her "anemia" without fully explaining the problem to her parents. He put her on intramuscular immune globulin shots for a few years, but Gini's reaction was not good, so after a few years they were stopped. Gini continued to be sick after quitting the shots, but quietly endured the viruses and illnesses during her high school years so she would not be considered a hypochondriac.

Later, during her first year teaching high school, Gini was unable to stay well, so she revisited her doctor to discuss her health. He was shocked that Gini had chosen teaching as a profession "given her disease."

"What disease?" Gini asked. Then 24 years old, Gini and her family were shocked to learn that she had lived with untreated primary immune deficiency disease (PIDD) all this time.

Gini began receiving monthly treatments of intravenous immune globulin (IVIG). The infusions often took up to eight hours, and she had to miss work the day of infusion and the following day, due to severe head and muscle aches associated with her IVIG. She tried to put on a brave face, but with the terrible side effects she was experiencing, it



*Gini Lea with her brother, Jeremy.*

was often very difficult to do. Eventually, after a year of IVIG, the medical staff had difficulty accessing her veins, and they suggested she use a port.

Recalling the moment, Gini said, "Honey, I wasn't about to explain a port to my dates!" Instead, she begged her way into a subcutaneous immune globulin (SCIG) clinical trial, and hasn't looked back since. "Woohoo!" she hooted regarding this method of administration.

Today, Gini works hard to stay healthy because she believes that her quality of life is directly impacted by her energy levels: They are one and the same to Gini.

She is very thankful that her SCIG treatment gives her enough energy to pursue her master's degree, teach high school biology, coach the tennis team and advise the Marine Science Club. She also scuba dives and plays recreational tennis.

And, she has become an advocate for herself and others regarding health. Her students refer to her as the "Clean Queen" because of her obsession with handwashing—hers and her students. They also call her "The Last Comic Standing," referring to the way in which she teaches. Gini's mantra is, "I don't love to teach, I teach to love."

Gini is not without the additional health issues many PIDD patients experience, but she said she comes from a family of Steel Magnolias. She has learned many lessons from the women in her family—especially the lesson of living gracefully with a chronic illness.

When asked how she views her future, Gini Lea said she is more herself than she has ever been, and she is both grateful and hopeful. ■

# ASK KRIS



IG Living is pleased to introduce Kris McFalls, the newest addition to our IG Living team. Kris will function in a role that's a bit of a spin on the traditional ombudsperson: Instead of responding to complaints (thankfully, we don't receive many!), she will respond to questions from our readers. And these we do receive—about all kinds of things!

Kris has two adult sons with chronic diseases treated with IG, and she is currently on the challenging quest for her own diagnosis. Formerly a physical therapist assistant, Kris is an avid patient advocate and now works with NuFACTOR, a sponsor of IG Living. Kris is eager to find answers to your questions. Email them to [editor@igliving.com](mailto:editor@igliving.com).

**Barbara:** Are there other diagnoses that go hand-in-hand with primary immune deficiency diseases (PIDD)?

**Kris:** I was at a conference where this very question was addressed. The doctors at the conference reported that indeed there are certain diseases associated with PIDD. However, the causes of the links are unknown. The doctors said the following diseases are more frequent and commonly seen in people with PIDD, especially common variable immune deficiency, and in family members of PIDD patients:

- hypothyroidism or Hashimoto's disease
- inflammatory bowel disease
- ulcerative colitis
- certain types of lymphoma
- arthritis
- other autoimmune diseases

Now, keep in mind that while they are more common in certain PIDDs, the doctors at the conference said there is currently no way to predict which patients will contract them.

**Lisa:** I currently use intravenous immune globulin (IVIG) for myasthenia gravis. I was wondering if subcutaneous IG (SCIG) would be an option for me.

**Kris:** There is no clinical data on the use of SCIG for autoimmune or neurological diseases. However, if you are serious about trying SCIG, gather as much information as you can, and share it with your doctor. Send him or her the information before your appointment, so you are both prepared to discuss it. Listen to what your doctor has to say, and make sure you are making decisions based on all the information. But don't be afraid to push a little to make your points.

**Betty:** Thank you for your recent article that mentioned intravenous immune globulin (IVIG) infusion rates. My only problem with the article is I could not translate the rate equation, as it is not in plain English. Is there a way to say what my rate should be in lay terms?

**Kris:** IVIG infusion rates can be very confusing, I agree! The rate referenced in the article is only one manufacturer's guidelines. Actual infusion rates vary—by product and by individual. Your rate should depend on your weight and on how you feel and what your reactions are to infusions.

All manufacturers recommend infusion rates in their product package inserts as guides, and these guides can certainly be used to start new patients. However, IVIG is often administered in a "ramping" fashion, meaning the rate of infusion starts out slowly and then is increased in increments until a maximum rate is obtained. This allows the patient to slowly become accustomed to the infusion. Each patient's infusion rate must be individualized to each brand of IVIG. Bottom line: Ask your doctor to explain your rate to you, and talk about any uncomfortable reactions you have to your infusions. ■

Have questions about living comfortably with your IG therapy?  
Send them to Ask Kris at [editor@igliving.com](mailto:editor@igliving.com).

# A Single Sugar Found Responsible for an Antibody's Ability to Treat Inflammation

For years, researchers have struggled to understand how IVIG worked. Its ability to treat autoimmune diseases seemed to be an apparent contradiction.

Intravenous immunoglobulin (IVIG) is a complex mixture of IgG antibodies made from human plasma that contains the pooled antibodies from thousands of people, and is only FDA-approved to treat a few assorted conditions; nonetheless, practitioners have used it off-label with varied success in patients with lupus, arthritis and asthma, among other autoimmune disorders. In the body, the antibodies in plasma act as part of the immune response to identify and deactivate foreign invaders. When they begin attacking the body's own cells, the same protective immunoglobulins (known as IgG antibodies) can cause autoimmune disorders like lupus, arthritis and asthma. And yet, when IVIG is infused into people with those exact autoimmune conditions, it calms inflammation rather than causes it.

Jeffrey Ravetch, Theresa and Eugene M. Lang professor and head of Rockefeller's Laboratory of Molecular Genetics and Immunology, was struck by this inconsistency. "If IgG triggers autoimmune disease, how could it be pathogenic and therapeutic?" he asked. "We call it the IgG paradox." Six years ago he started an investigation into exactly how IVIG worked, and what he's discovered could one day lead to a whole new class of therapeutics. In a paper published in the journal *Science*, Ravetch and his colleagues, Falk Nimmerjahn and Yoshi Kaneko explain what makes IVIG effective: A small fraction of the IgG antibodies in the IVIG solution

carry a sugar called sialic acid that is required for its protective ability.

IgG antibodies bind to and activate specific immune cells, with different forms or "subclasses" binding to specific receptors (called Fc receptors) on the immune cell's surface. Antibody subclasses have different abilities to induce inflammation in the body by virtue of their selective ability to engage either activating or inhibitory Fc receptors. Earlier work had shown that IVIG infusion changed this ratio of activating and inhibitory receptors

*"If IgG triggers autoimmune disease, how could it be pathogenic and therapeutic? We call it the IgG paradox."*

on the cells that trigger inflammation, rendering the pro-inflammatory autoantibodies in autoimmune diseases, like lupus and arthritis, less inflammatory. The next logical step then, Ravetch says, was determining how the IgG molecules in IVIG preparation could have an anti-inflammatory effect.

Because a therapeutic, anti-inflammatory response to IVIG requires a concentration of IgG antibodies that's hundreds of times greater than is normally used for antibody therapy for cancer or infection, for example, Ravetch and his colleagues began to look for something that was only

present in IVIG in small amounts. That's how they discovered that just the very terminal sialic acid on the Fc portions of the IgG molecule were the root of the anti-inflammatory activity. When the researchers removed the sialic acid, the molecule retained its structure and its half-life, but it lost its protective abilities. "This is a very interesting condition that's set up," Ravetch says. "IgG can shift from a state that is quite inflammatory to a state that is actively anti-inflammatory by just changing a sugar." This switch occurs during a normal immune response to a foreign substance, shifting the IgG antibodies from an anti-inflammatory state to one that is pro-inflammatory and able to efficiently dispose of the foreign challenge.

To test the theory, Ravetch and his colleagues tried enriching IVIG for the IgG molecules that contained sialic acid. They found that just enriching for this IgG species increased IVIG activity by a factor of ten, while removing it wiped out the therapeutic activity altogether. This discovery, Ravetch says, has potentially huge implications, and his lab is now working to generate a recombinant form of IgG that, by virtue of a sialic acid molecule attached in the right place, will be anti-inflammatory and could act as a novel treatment for autoimmune disorders. "We have the opportunity to make a much better form of IVIG that will work 100 times better and be a pure molecule—to build a much better class of therapeutics based on a property that already exists in nature." ■

*Note: This article is provided by The Rockefeller University.*

# Stiff-person Syndrome

By Dan Bennett

It's an illness that can cause those who suffer from it to fear crossing the street, entering a department store or even standing in the middle of a large space—due to a sudden onset of body rigidity that renders the afflicted person almost immobile.

Yet even with such dramatic symptoms, the rare condition stiff-person syndrome (SPS—formerly known as stiff-man syndrome) is practically unknown, although people with SPS, and their doctors, are seeking to change that. And the name of the condition helps by getting right to the point, at least superficially: Stiff-person syndrome causes—among other difficult symptoms—stiffness or rigidity in the body that renders the legs and arms unable to bend.

"My symptoms started in 1993, when I was 22 and had just graduated from college," said Anisah Hassan. "I got to the point where I couldn't bend my right leg. Two weeks later I couldn't bend either leg, like I was walking on stilts."

Hassan, a St. Louis resident, is not alone with such early symptoms, but there are very few people with whom she shares the affliction, as only a few hundred people are diagnosed in the U.S.

The National Association for Rare Disorders describes stiff-person syndrome as an "extremely rare neurological disorder, characterized by progressive muscle stiffness and spasms. It usually begins in young adults, first involving muscles of the trunk and progressing to affect muscles of the legs. ... Those affected have a characteristic stiff-legged way of walking and increase in the curvature of the spine—lordotic posture."

The syndrome might begin as recurring episodes of stiffness and spasms, often triggered by being startled or minor physical contact.

While symptoms are bad enough, perhaps just as painful

to those with SPS is the fact that the exact cause of the syndrome is not known. And, because the disease is so rare, it is often misdiagnosed.

"I went to the emergency room, and eventually, after a few consultations, was diagnosed with multiple sclerosis," Hassan said. "The six years of injections and steroids that followed caused me problems. Then I saw a number of neurologists who said maybe it was not MS, maybe it was Parkinson's. Then I received a diagnosis of rigid-spine syndrome, and we stuck with that one until February 2006, when a doctor finally said to me 'stiff-person syndrome.'"

Although Hassan was finally diagnosed accurately, the previous misery and confusion had taken its toll on Hassan, who at one point, before being correctly diagnosed, attempted to check into a psychiatric ward. At the time, a doctor encouraged Hassan to practice crossing the street, but she refused. Like many sufferers, anxiety attacks about such activities are one of the SPS symptoms, leading medical researchers to link the illness with the brain. Peculiar to the condition is that it can be set off when the patient is startled by something, such as a car horn or the loud clapping of hands.

"My IVIG helps calm the startle reflex," Hassan said.

Debra Kemery, a resident of Colorado, was 36 when she was diagnosed with SPS.

"When I was diagnosed in 1994, there wasn't much Internet access and people couldn't connect," Kemery said. "So I eventually started a website where those who suffered or those who wanted to know more could go. I wanted to give a personal spin to the illness so that people could crawl inside the mind of somebody with a chronic illness. I wanted to give hope."

Kemery's beautifully designed site does just that, detailing

her early struggles with the illness, then how she eventually coped with medication and immune globulin (IG) infusions. The site is the place where a support group of those who have the illness tell their stories and learn from one another.

"The support group has been helpful," Kemery said. "It's a good base to share anecdotal experiences. You don't feel like such a pariah. When an illness is as rare as this, and the name kind of frivolous, the name doesn't let people know how serious it is. The site and support group help get the word out."

The site encourages people with SPS to learn as much as they can about their illness, and to share what they hear from others with their doctors.

"We talk a lot about being proactive," Kemery said. "It's easy to play the victim, but you can't do that."

The same goes for battling health-insurance issues, Kemery said. She's waged an ongoing battle with insurers since the Medical Modernization Act of 2003 stripped some benefits of those trying to get intravenous IG treatments and other medications.

"It's a tragedy when beneficial treatments may be unavailable to those who can benefit from them the most," Kemery said.

John Crawford, a retired business owner in Florida, suffers from SPS and chronic inflammatory demyelinating polyneuropathy (CIDP). Crawford says sharp pain in his ankles led to tests for everything from diabetes to multiple sclerosis. Eventually, he was diagnosed with SPS, in addition to his previously diagnosed CIDP.

"The course of action for me was IVIG," Crawford said. "We were asking for the IVIG to combat two illnesses, but there was significant improvement for a long time. I started therapy in 2000 and continued until 2003, when I started to decline a little."

At the same time, Crawford's insurance ran out, and he was forced to stop IG treatment.

"I figured this was going to be the end," he said. "My wife and I started to make my funeral arrangements."

Then, at his wife's suggestion, Crawford began acupuncture treatments, which Crawford said helped him tremendously, and continue to do so.

"A month after I started, though, my wife died, a tremendous blow," he said. "But two years later, I'm walking without a cane much of the time."

Crawford says his experience talking with doctors and conducting research tells him that if a cure for SPS is found, it will happen as a result of a breakthrough in curing other illnesses, such as multiple sclerosis or diabetes.

"I'm not optimistic there is going to be a lot of expensive research on an illness that affects only 300 people," he said. "I am optimistic, though, that there is research going on in other areas that will somehow help us."

Meanwhile, patients who receive support from others with the same disease appear to be hopeful about the future.

"I try to keep a positive attitude," said Windelle Smith, a 51-year-old Florida resident. "My daughter helps me through things, and I keep trying to push myself to stay strong."

Perseverance is a common theme among those who suffer from stiff-person syndrome.

"And humor," says Debra Kemery. "Before I was diagnosed, I took the future for granted, the future was a guarantee. Once I became ill, I became aware that I'm mortal. Nobody has a guarantee. But that's not a bad thing. Being mortal means you're alive today, so live the best you can today." ■

## Resources

For more information about  
stiff-person syndrome

National Association for Rare Disorders (NORD)

[www.rarediseases.org](http://www.rarediseases.org)

800-999-6673

[orphan@rarediseases.org](mailto:orphan@rarediseases.org)

Debra Kemery's website

[www.stiffman.org](http://www.stiffman.org)

American Autoimmune Related Diseases Association Inc.

[www.aarda.org](http://www.aarda.org)

800-598-4668

[aarda@aarda.org](mailto:aarda@aarda.org)

Autoimmune Information Network Inc.

[www.aininc.org](http://www.aininc.org)

732-262-0450

[autoimmunehelp@aol.com](mailto:autoimmunehelp@aol.com)

National Institute of Neurological  
Disorders and Stroke (NINDS)

[www.ninds.nih.gov](http://www.ninds.nih.gov)

800-352-9424

[braininfo@ninds.nih.gov](mailto:braininfo@ninds.nih.gov)

# ZLB Behring Becomes CSL Behring—and Retains Its Vision

By Kit-Bacon Gressitt

**A**s ZLB Behring transitions its name to CSL Behring around the world, the company, maker of immune globulin products Carimune NF and Vivaglobin, is maintaining its focus on patients and safety, according to Robert D. Lefebvre, vice president and general manager of CSL's U.S. Commercial Operations, headquartered in Pennsylvania.

"CSL stands by its passion," Lefebvre said in a recent interview, "our passion to serve the patient communities, to bring them what they tell us they need. ... And we've been a leader in bringing safe immunoglobulin products to the world. Our record on safety is beyond reproach. We've continued to invest in improvement, not just of the product itself, but of the manufacturing process as well."

CSL Behring, one of several companies owned by Australia-based CSL Limited, has a record of more than 90 years experience in healthcare, developing and manufacturing vaccine and plasma protein therapies, including immune globulin products. The company's commitment to research and development—and to its customers—resulted in the U.S. introduction of Vivaglobin last year, the first U.S.-licensed subcutaneous immune globulin (see IG Living April-May 2006).

"Vivaglobin has had a tremendous effect on patients," Lefebvre said. "The more people who go on Vivaglobin, the more IVIG is available for other patients. Well over 1,000 patients are on treatment." He estimates that figure will grow to 3,000 within six to 12 months.

"We've also acquired a new product from MedImmune," Lefebvre said. "CytoGam is a hyperimmune IVIG used for solid organ transplantation. CSL's expertise in sourcing and manufacturing, coupled with our knowledge of marketing, we can make a significant difference there. It's a totally new therapeutic area for us."

CSL is also launching a flu vaccine, yet another new arena for the company, which appears to be on a bit of a business development roll. But, what does this all mean for the patient?

Lefebvre indicated it means a lot of good things to come, that the company is committed to getting patients the products they need through its integrated supply channel.

"How do you ensure that the protein gets from the plasma of a human volunteer into the vein of the patient in need?" Lefebvre mused. "We take all of the steps that need to happen to ensure that that's done so the right patient receives the right protein and receives it in a way that will be safe and therapeutic. How do you collect it, how do you turn the plasma into an effective plasma protein product, how do you get it from the manufacturer to the care provider, and how do you get it into the vein of the patient? That is the channel.

"Our mission is to see where we can bring our expertise—in research and development—into rare and medically important disease areas to solve problems in these areas," Lefebvre continued. "We'll work closely with the physician and the patient to continue bringing that passion to serving our patients' needs well."

And that's exactly what patients need from the companies making the products that keep them alive. ■

## CSL's Vision

CSL Behring is passionate about safety, quality and the consumers we serve. Our extensive research and development and range of support services demonstrate our ongoing commitment to all who count on our products. We are deeply committed to providing the best possible therapies and services.

## Great Resources at the CSL Behring Website

### About Plasma and Plasma Therapies

<http://www.zlbbehing-us.com/s1/cs/enus/1153606276924/content/1151517255178/content.htm>

### Reimbursement Resource Center

<http://www.zlbbehing-us.com/s1/cs/enus/1151517254336/content/1151517254307/content.htm>

### Immune Mediated Disorders

[http://www.zlbbehing-us.com/s1/cs?childpagename=US%2FWeb\\_Content\\_C%2FContentDetail&c=Web\\_Content\\_C&pagename=Web%2FWrapper&packedargs=lang%3DEN&cid=1151517256807](http://www.zlbbehing-us.com/s1/cs?childpagename=US%2FWeb_Content_C%2FContentDetail&c=Web_Content_C&pagename=Web%2FWrapper&packedargs=lang%3DEN&cid=1151517256807)



# Let's Talk!

By Shirley German Vulpe, EdD

For this column, I interviewed Joe Wise, a young man of 13 years, with mitochondrial myopathy muscular dystrophy, which causes his immune system to be compromised. Joe swims for health—and happiness.

**Shirley:** Can you tell me a little bit about your illness?

**Joe:** I was diagnosed with muscular dystrophy [MD] when I was in fifth grade. I had a pain in my hip that wouldn't go away. At first, we thought it was a football injury. But the blood work told us it was MD.

**Shirley:** Why do you receive immune globulin (IG)?

**Joe:** My immune system is very low. I was getting very sick, lots of colds and the flu. My disease makes me very tired.

**Shirley:** When do you get your IG?

**Joe:** I get it every Sunday.

**Shirley:** Do you have to do anything else to treat your illness?

**Joe:** I also use a ventilator to help with my breathing. I sleep with it every night. I use it two hours in the day. Fifteen minutes at lunch time, one and three-quarters hours when I get home.

**Shirley:** What grade are you in, Joe, and what activities are you involved in?

**Joe:** I'm in eighth grade in public school. Mostly swimming. I like that a lot. I swim every day for two hours. It keeps me strong, to fight through my disease. I also ride bikes and do weight training with my dad.

**Shirley:** Do you swim with a team?

**Joe:** Yes, the Palo Alto Stanford Aquatics.

**Shirley:** What strokes do you swim?

**Joe:** Long distance freestyle and backstroke.

**Shirley:** Have you been in any meets?

**Joe:** Yes, four [in 2006]. I did pretty good: sixth out of 144 kids.

**Shirley:** What is your best race?

**Joe:** The 500 freestyle and 400 freestyle, long course.

**Shirley:** When did you start swimming?

**Joe:** Seven years ago, when my mom encouraged me to try it. Then, after I was diagnosed, my doctor said swimming was the only sport I could do. When I started, I didn't really like it much. That's because I was the slowest swimmer on the team. Then I started training harder and began to improve. Now that I'm one of the fastest kids, I really like it, and I swim all year-round. I have a lot of swim team friends.

**Shirley:** Do you swim in any other meets?

**Joe:** Yes. The Paralympics for the disabled.

**Shirley:** How did you learn about them?

**Joe:** I saw a kid on TV, and I have a friend who is also a paralympic athlete.

**Shirley:** What happened next?

**Joe:** I joined. I won a gold medal in the 400 freestyle in August, at the U.S. Paralympics Nationals in San Antonio. I found out I was the second fastest in the S10 classification in America. I'm ranked the 17th fastest 400 freestyle S10 swimmer in the world.

**Shirley:** What does S10 mean?

**Joe:** Somewhat disabled, not a lot.

**Shirley:** What do you want to do next?

**Joe:** I hope to get picked for the 2007 Para-Pan America Team. The qualifying meet's in Canada, and the Para-Pan American Games will be held in Rio de Janeiro, Brazil, in August. Then I hope to qualify for the U.S. Paralympics Beijing Team for September 2008.

**Shirley:** That is great! Sure sounds to me like you have a good chance.

**Joe:** I'm going to work very hard for it! My motto is: You can do anything you want if you put your mind to it. ■

## Resources

### Muscular Dystrophy

Muscular dystrophy is a genetic disorder that weakens the muscles of the body. For more information, visit the Muscular Dystrophy Association website at [www.mda.org](http://www.mda.org).

**U.S. Paralympics** [www.usoc.org/paralympics](http://www.usoc.org/paralympics)

U.S. Paralympics is a division of the United States Olympic Committee, formed in 2001 to increase support for paralympic sport in the United States. U.S. Paralympics coordinates the training and selection of athletes for the U.S. paralympic teams. U.S. Paralympics also works with national and locally focused sports organizations that offer paralympic programs to children and other developing athletes. Through these efforts, U.S. Paralympics promotes Olympic ideals throughout the American population, especially among those Americans who have physical disabilities.

# Looking for Labbie Love

By Cheryl Haggard

**H**ave you ever found yourself in a conversation with a non-immune compromised person offering suggestions on how to clear up your PIDD kid's 3-month-old sinus infection? Maybe it's the bags under my eyes or the bald spots from pulling my hair out as a result of my kids' chronic crud that encourages the healthy to offer advice such as:

*Try a warm compress on her forehead. It really works for our perfectly healthy 18-month-old, who has never been up all night long screaming from an ear infection.*

Or:

*Breast milk is nature's antibiotic. Try rubbing a bit into his pus-filled eye, and I guarantee you he won't need that eye-duct surgery.*

How about:

*Have you ever tried gummy-vitamins? I sell them, but I will let you have a month's supply at my wholesale price, as I see the IV line hanging out of your son's arm probably costs a pretty penny.*

And my all-time favorite:

*Gosh, I am so sorry to hear your daughter was in the hospital with diarrhea. Purified clay does the trick when I get the runs. Mix it in with a little Jell-o, and she'll slurp it down like it's liquefied chocolate."*

I know people mean well, but they sometimes don't know how off the mark they are when giving advice on keeping my PIDD kids healthy. If all it took was a chewy vitamin, some well-placed breast milk or even chocolate-covered dirt to solve our illnesses, you would find me in the backyard mining the best clay possible. It's just not that simple.

But after a long wrestling match, trying to get an IV placed in our son Caleb, the Child Life Specialist offered us a suggestion that changed our lives forever.

"Why don't you get a dog?" she offered, wiping sweat from her brow.

A dog? I pondered. That's all I need, something else in the house that eats and poops.

"How is a dog going to help?" I asked the woman who



Illustration by Kim Kimman

gave the suggestion, a woman who would not be cleaning up after this aforementioned four-legged creature.

Her arguments for a dog's calming effects led me to an Internet search and an application to the local Labrador retriever rescue center. A two-week wait and pounds of paperwork resulted in an official approval: We were worthy to adopt a living lump of hair.

"We've found the perfect Lab for your family," the nice woman announced from the other end of the phone. "There are, however, a couple of medical issues with George that I want to make you aware of."

Great, more problems. That's all I needed. George was supposed to ease my family's problems, not add to them.

After a heavy sigh, I asked her to "give it to me straight."

"He's a bit of a food hog, so he's about 20 pounds overweight, and he's got a problem with ear infections, so he requires antibiotics regularly," she sheepishly confided.

That's when I lost it. I'm sure my peels of laughter echoed in Timbuktu.

"We'll take him!" I shrieked. "We're all about 10 pounds overweight, and ear infections are as normal to us as turkey at Thanksgiving!"

George, the 110-pound, ear-infected chocolate Lab, fit right into the family. Even antibiotic distribution has become something we celebrate instead of dread. Nightly, I line everybody up for their prophylaxis and we all cheer: "One for you, one for you and one for George! Yeah!"

Mary Poppins sang "a spoonful of sugar makes the medicine go down." For us, it is a hunk of burning Labbie love.

So when someone offers me fish oil for flatulence, yellow squash for yeast or crushed cranberries for the crud, I smile sweetly and respond, "Thanks for the advice." Then I thank my lucky stars for the 110 pounds of chocolate I have waiting for me at home. ■

# The Juggling Act

By Lauren Gerstmann, MPH

Having primary responsibility for the health of another person can be gratifying and rewarding; it can also be a terrifying juggling act. If you are a parent earning an income and the caregiver of a chronically ill child, how do you balance the needs of your child, your job, your mate—and still take care of yourself?

In theory, there are U.S. government policies in place to protect parents in this position.<sup>1</sup> Under the Family and Medical Leave Act of 1993 (FMLA), covered employees are entitled to a total of 12 administrative workweeks of unpaid leave during any 12-month period for:

- the birth of a son or daughter and care of the newborn;
- placement of a son or daughter for adoption or foster care; care of your spouse, son, daughter or parent with a serious health condition; and
- your own serious health condition that makes you unable to perform the duties of your position.

You are covered under FMLA if you work for an employer with 50 or more employees or if you work for a public agency or school. Additionally, if you are a federal employee with a medical emergency, and you have exhausted your own leave, the leave transfer program allows other federal employees to donate annual leave to you. Some private employers also

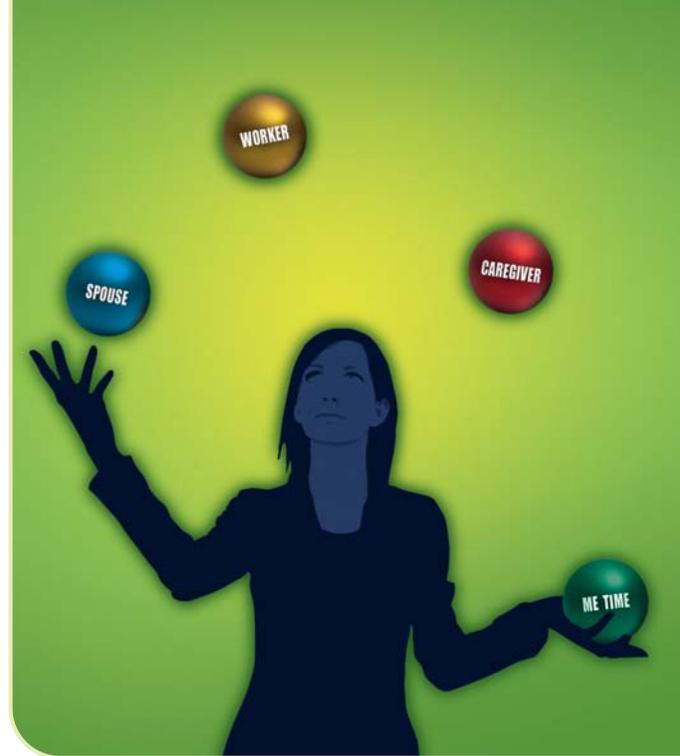
have leave-banking programs. These programs allow members (those who contribute a specific amount) to apply for leave from the leave bank in the event of a medical emergency.

These are helpful policies but may be more practical to allow you to care for someone with an acute illness rather than a chronic illness. Also, since these policies legislate unpaid leave, you may still have trouble if your income is a key family support. Additionally, there are subtle pressures not to use the leave to which you are legally entitled. Jane's son, Stuart,<sup>2</sup> needed surgery on his airways a few weeks after he was born. Post-surgically, he was incredibly vulnerable to infection and anything he ate could get into his airways and cause an infection. His doctors gave Jane strict instructions that anything other than breast milk could be very dangerous for him. In theory, Jane had access to a very generous leave policy in her office, but the reality was that other people were financially dependent upon her being in the workplace. Jane's funding and her staff's salaries were dependent upon quotas that they could not meet without Jane's direct supervision. Jane went back to work.

It is very stressful when the work that allows you to pay for health

insurance and medical care also can take you away from your child. The situation becomes additionally stressful if your child has a chronic illness that prevents his or her functioning in a group care or group educational situation. Individual Education Programs allow children with special needs to be mainstreamed into public school and give their parents the freedom to return to the workplace. But, if your child is too ill to come into contact with many other children, you need to find a solution that will allow your child to stay at home.

The stress and overwork may end up compromising your own health. Research has shown that women even more than men may suffer health consequences. But, this may be a result of how caregiving is structured rather than due to a true gender difference in health outcomes.<sup>3</sup> Traditionally, men have cared for their families by offering financial support, while women have borne the brunt of hands-on care. But more and more families can no longer afford to have one parent stay at home. As a ➤



<sup>1</sup> U.S. federal policy can be found at: <http://www.opm.gov/oca/leave/html/levbro.htm>

<sup>2</sup> All situations presented in this article are true, but names have been changed to protect privacy.

<sup>3</sup> Burton P, Lethbridge L, & Phipps S, "Children With Disabilities and Chronic Conditions and Longer-term Parental Health" <http://atlanticresearchdatacentre.dal.ca/> 2006

result, fathers are increasingly sharing in major caregiving duties. Soon, we are likely to see them suffering the same burden of stress and health consequences. Fathers also face some unique pressures as the emphasis in the literature and in the workplace is on improving the work and family balance for mothers. Men may be less comfortable taking lengthy leave to bond with a newborn child, and may be less likely to discuss their caregiving responsibilities with colleagues.<sup>4</sup> As a result, the discussion may shut men out, making men feel more alienated and less able to share in childcare. Ironically, this leads to more of a burden on mothers.

Janet Gornick and Marcia Meyers have extensively studied the balance between work and family life in the United States. In their 2003 book "Families That Work,"<sup>5</sup> they compare the quality of the balance that families are able to achieve here with the balance that European and Canadian families are able to achieve. Gornick and Meyers conclude U.S. federal policies are less supportive of families than are the policies of any of the 12 other countries that they studied. Based on what they have learned from other Westernized nations, their goal is to create a "dual earner-dual career society." They define policies that will support entire families, allowing women more choices in returning to the workplace, making caregiving support more inclusive of men, maximizing children's time with their parents, and making sure that all children have access to high-quality care. Specifically, they advocate more generous family leave policies (for both parents), reducing the number of hours in the work week, increasing the quality and amount of publicly provided early childhood care (for example, public preschool), and matching the time that children are in school more closely to the time that parents are at work. While their work is targeted toward families in general, some of their policies (particularly their suggestions for leave policies) are especially relevant for families caring for chronically ill children.

Unfortunately, our government is a long way away from adopting the ideals of Gornick and Meyers. But some of their suggestions can be adapted to personal use. For example, (when possible) involve both parents in childcare—especially if both parents work. Take turns using your sick and vacation time to care for a child at home. Perform a cost benefit analysis for your job versus

your healthcare responsibilities. If both parents are working, but specialized schooling, increased sick time, and increased payments to professional caregivers total up to more than the income of one parent, perhaps that parent should stay home. Don't automatically assume that this means Mom needs to stay home—Dad may prefer to and should feel equally supported if he makes this decision. Fathers who wish to stay home with their children should know that they have plenty of company. In 2003, roughly 160,000 fathers made the choice to stay at home to care for their children.<sup>6</sup>

Another option, instead of taking the financial hit of having one parent stay at home, is that some families share caregiving by making the decision to "tag-team" parent. Parents stagger their work schedules so that one parent is always home with the children. This approach has a number of advantages. It is very economical because it allows for two incomes without the cost of paid childcare. And, if your children have complicated health needs, it means that they will always be with the people who know best how to care for them. Tag-team parenting may also allow both parents to pursue their careers. But, for all of the benefits, consider this option cautiously as all that time apart can place serious stress on your marriage.<sup>7</sup>

If you are a single parent or if both parents need to work, you might want to think outside the box to consider unusual solutions. Jane, mentioned above, felt obligated to return to her office even though her son was chronically ill and dependent upon her. However, Jane was unwilling to jeopardize the health of her child, so she spoke with her boss and explained that she could come back to work only if she could bring her nursing infant with her. Her boss allowed the baby in Jane's office, and Jane was able to take care of her child and follow through on her responsibilities to her staff.

Another way to spend more time at home is to reduce your hours in the office, either through a reduced schedule or by telecommuting. Part-time work is a great option but often pays less than full-time work and may not provide you with benefits. Sometimes, though, you may be able to reduce your hours without officially working part time. For example, at the University of Southern California, you are eligible for benefits as long as you work more than 50 percent of a full-time schedule. So you could work 21 hours per week and still receive the benefits. There are

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<sup>4</sup> Cooper, M, "Being the Go-to guy: Fatherhood, masculinity, and the organization of work in Silicon Valley" in "Families at Work: Expanding the Bounds." Vanderbilt University Press, 2002.

<sup>5</sup> Gornick JC & Meyers MK "Families That Work: Policies for Reconciling Parenthood and Employment" Russell Sage Foundation, 2003.

<sup>6</sup> Census Bureau; 2003 March CPS : America's Families and Living Arrangements <http://www.census.gov/prod/2004pubs/p20-553.pdf>.

<sup>7</sup> Boushey, H, Tag-Team Parenting: Center for Economic and Policy Research August 2006 [http://www.law.georgetown.edu/workplaceflexibility2010/News\\_Roundup/2006/September/1/documents/tagteamparenting.pdf](http://www.law.georgetown.edu/workplaceflexibility2010/News_Roundup/2006/September/1/documents/tagteamparenting.pdf).

also options to increase your flexibility while working full time. If your work can be done over the computer or telephone, you may be able to telecommute part of your hours. Telecommuting might provide benefits for your employer as well, especially if office space is hard to come by or if your employer is active in preserving the environment. In 1992, AT&T implemented a corporate policy to encourage telecommuting. The plan has been both popular with employees and good for the environment (which is, of course, good for AT&T's reputation!). "Reducing the number of AT&T people in single-occupancy vehicles all day or during rush hour can result in enormous savings in fuel consumption, pollution and traffic congestion," says Kathie Fink, of AT&T's Environmental Health and Safety team.<sup>8</sup>

If you are interested in some of these options but are unsure how to make it work, check out WorkOptions.com.<sup>9</sup> For a fee, you will find templates and tips for writing a

proposal to telecommute, work part time, job share or switch to a compressed workweek schedule.

So to sum it up, because our government does not provide a comprehensive program to assist you in supporting the health and financial needs of your children, you need to be creative. But there are solutions out there, and which solution is best depends upon your individual situation. If you are in a position to forgo some income, Mom or Dad can stay at home, work part time or make more efficient use of employee leave policies. If you cannot forgo income, try an alternative work schedule, telecommuting, staggered work schedules or working a compressed week. Maximizing your ability to care for your child while earning an income is daunting, and American families do not get a lot of help. But, with creative time management and research into the resources available to you, you can do both of your jobs well. ■

<sup>8</sup> AT&T website: [http://www.att.com/ehs/annual\\_reports/ehs\\_report/report96/page11.html](http://www.att.com/ehs/annual_reports/ehs_report/report96/page11.html).

<sup>9</sup> <http://www.workoptions.com>.

## A Resource Toolbox for Working Caregivers

### Alternative work options

#### Home Based Careers

[www.edirectoryofhomebasedcareers.com/dir2.htm](http://www.edirectoryofhomebasedcareers.com/dir2.htm)

Download an eDirectory of home based careers.

#### Work Options

[www.workoptions.com](http://www.workoptions.com)

Templates and tips for writing a proposal to telecommute, work part time, job share, and/or switch to a compressed workweek schedule.

### Caregiver support services

#### Caregivers USA

[www.caregivers-usa.org/db/index.html](http://www.caregivers-usa.org/db/index.html)

Provides an index of local and state caregiver support services.

#### AARP

[www.aarp.org](http://www.aarp.org)

If you are caring for a spouse or parent, AARP has valuable online resources.

Please see their research report on "Caregiving in the United States" at [www.aarp.org/research/housing-mobility/caregiving/fs111\\_caregiving.html](http://www.aarp.org/research/housing-mobility/caregiving/fs111_caregiving.html). Links to caregiving resources are also provided at this site.

### Financial resources

To help you obtain insurance, medical care or drug benefits:

#### Patient Advocate Foundation

[www.patientadvocate.org](http://www.patientadvocate.org)

800-532-5274

A national nonprofit organization that serves as an active liaison between the patient and his insurer, employer, and/or creditors to resolve insurance, job discrimination and/or debt crisis matters relative to his diagnosis through case managers, doctors and attorneys. They offer a pharmaceutical co-pay assistance program to patients who qualify medically and financially.

#### Families USA

[www.familiesusa.org](http://www.familiesusa.org)

202-628-3030

A nonprofit organization dedicated to the achievement of high-quality, affordable health and long-term care for all Americans. At [www.familiesusa.org/resources/program-locator](http://www.familiesusa.org/resources/program-locator), they offer a program locator that will direct you to a local program to answer questions and assist you in obtaining health insurance. Programs may also be able to refer you to low-cost or free health care, including prescription drug assistance.

#### Health Insurance Resource Center

[www.healthinsurance.org](http://www.healthinsurance.org)

A resource for families, individuals and the self-employed, this website provides the tools to become a better-informed health insurance consumer. It is also possible to obtain quotes from reputable, financially stable insurance companies. This website will also help you find insurance if you have a pre-existing medical condition.

#### The Medicine Program

[www.themedicineprogram.com](http://www.themedicineprogram.com)

Volunteers who are dedicated to assisting the growing number of patients who cannot afford their prescription medication established this organization. Their function is to help patients who may qualify to enroll in one or more of the many patient assistance programs that provide prescription medication free of charge to individuals in need.

#### SKIP: Sick Kids Need Involved People

212-268-5999

This is an advocacy group that helps families receive financial aid, nursing services and government medical services that they may be entitled to for their chronically ill child.

# Munchausen Syndrome

By Jeff Siegel

It's one of those things that's only supposed to happen in a bad movie. A mother, whose child has been sick for much of his young life, keeps taking the child to doctors who have trouble coming up with a diagnosis; when they do, it seems to be the wrong one.

The mother takes her child from doctor to doctor trying to find one who can help, and that's when the story really takes off. The child, says the next doctor, is fine, if a little depressed. It's the mother who has the problem—who imagines these diseases for her son because she enjoys the attention it brings her. There's even a medical name for it—Munchausen syndrome by proxy (MBP), and it's not pretty by any name.

Or at least that's what Connie Worthen discovered. Her son, Branson, was eventually diagnosed with common variable immune deficiency, but not before Connie was told she was committing Munchausen by proxy. "If you're a parent, it's about the worst thing that you can imagine happening," she says.

She has since discovered that other parents of children with primary immune deficiency diseases (PIDD) have experienced the same thing. Their children's problems were so difficult to diagnose and so baffling to even experienced physicians that the parents were suspected of Munchausen by proxy. Not surprising, the number of such misdiagnosed cases are unknown. There aren't even any good statistics on Munchausen syndrome by proxy or its more common sibling, Munchausen syndrome, which involves fabricated illness for oneself. But, Gary L. Malone, MD, a psychiatrist who is the medical director of behavioral services at Baylor All Saints Medical Center in Fort Worth, Texas, says the misdiagnosis is not an unusual occurrence.

"Some doctors want a simple, take-home message," says Dr. Malone, whose practice includes patients who have been misdiagnosed and have suffered mentally as well as physically. "But in cases like these, there's not a quick, easy diagnosis. And some doctors don't like to not know what's wrong with patients."



## Defining Terms

Munchausen syndrome is mental illness in which someone acts as if they have a physical or mental disorder, when they have actually caused the symptoms themselves, according to information from the Cleveland Clinic. This is not a con or a scam; people with Munchausen syndrome are genuinely ill, not physically but mentally. They have a need to be seen as ill or injured and are even willing to undergo painful or risky tests and operations in order to get the sympathy and special attention given to people who are really sick.

Munchausen syndrome by proxy describes a pattern of behavior in which care providers deliberately exaggerate, make up or even induce physical and psychological problems in others, usually (but not always) their children. It's a form of child abuse, say physicians, that often includes more common behavior like physical or sexual abuse, but where the parent also commits the acts for the attention he or she receives.

The disease is named for Baron von Munchausen, an 18th-century German soldier who exaggerated stories about his life. The exact causes of Munchausen and Munchausen by proxy are not known, but researchers believe biological and psychological factors play a role. Some theories suggest that a history of abuse or neglect as a child, or a history of frequent illnesses requiring hospitalization, might be factors associated with Munchausen syndrome. Even less is known about Munchausen syndrome by proxy. Sometimes, but not always, a care provider

performing Munchausen syndrome by proxy is a well-educated and overly attentive mother who is solicitous of medical staff and able to speak medical terminology. She is always willing to change doctors and hospitals, asks for specific procedures and invasive and painful treatments for the child, is resistant to new diagnosis, and gives incomplete medical histories.

Doctors think both syndromes are extremely rare (although they do seem to make regular appearances on TV shows). Despite the lack of reliable statistics, Vali Hawkins Mitchell, PhD, the executive director of the Kirsha Foundation in Richland, Wash., says that psychiatrists and psychologists who see Munchausen and Munchausen by proxy in a clinical setting are beginning to think both may be more prevalent than previously thought, due to what she calls their inherent covert nature. "Just as immune deficiency is now more clearly recognized, as are other disorders that were once not diagnosable like tuberculosis, consumption, diabetes, and cystic fibrosis, Munchausen by proxy is now on the radar," she says.

### Looking for a Diagnosis

So what's a mother to do? Because, as Worthen and Kelliann Connor, who has two children with PIDD, learned, they each fit the Munchausen by proxy profile.

"It got to the point where I was kind of leery to even tell the whole story," says Connor, a pharmacy technician whose older daughter took several years to be diagnosed and went through the gamut of physicians and tests. "I got so used to not being believed that I started to think the new doctors wouldn't believe me either."

In fact, says Dr. Malone, some parents (as well as immune deficiency patients themselves) even give up going to a doctor, once they're convinced no one will believe them.

What causes this misdiagnosis? Doctors Malone and Mitchell say there are several reasons, one of which stems from the best of intentions. Most physicians genuinely want to help the child, and their assertion of Munchausen syndrome by proxy is done to protect the child when they can't find anything else wrong. "There are always two sides to a story," says Dr. Malone, "and the last thing the doctor wants to do is to feed into a hoax."

Yet, that being said, physicians do sometimes mistake primary immune deficiency, which is relatively rare, for Munchausen syndrome or Munchausen syndrome by proxy, which are rarer still. Sometimes this is attributable to inexperience, since most physicians will see few, if any, cases of either during their careers. Some of it, meanwhile, is just part of the way medicine is practiced, says Dr. Mitchell. "Most caregivers, who have less than a few

moments with the patient, will eliminate the obvious, categorize the simple, and avoid the time it takes to listen with a critical ear for the most subtle of messages about immune deficiency or Munchausen by proxy," she says. "This is not necessarily the fault of the caregiver as much as it is simply economically predictable."

And sometimes, says Dr. Malone, it's a mistake, whether from arrogance or overwork or the too common need for doctors to have an answer for every case they see. "Some doctors," he says, "always have to be right."

All of which means it's up to the parents to persevere. "You are your only advocate for your child," says Worthen, who has two other children.

This is much the same advice that Doctors Mitchell and Malone offer. Parents must:

- Educate themselves about what's wrong with their children. It's not enough to tell the doctor the child is sick. Know the symptoms and know what they mean, such as the relationship between sinus infections and immune deficiency.
- Understand that doctors are human, and that they make mistakes just like anyone else. "Doctors are your consultants," says Dr. Malone. "Use them as consultants, and use them for what they are capable of doing."
- Realize that they—and not the doctors—are the people who must make the decisions about treatment and care. Be patient, persistent and document everything.

According to Dr. Mitchell, in the end, perseverance pays off. In one case, a child seemed to suffer from recurring pancreatitis. The child was brought to a clinic 14 times and received 14 diagnoses. Eventually, the mother was assessed for Munchausen syndrome by proxy. Her child would have severe symptoms at home, but seemed fine when the child saw a doctor. It wasn't until the 2-year-old vomited on several visiting physicians and remained unconscious that the doctors decided the child was truly sick.

"My general rule of thumb," says Dr. Mitchell, "is that mothers do know what is going on with their children better than the caregiver's four-minute intake interview. I think everyone needs to know that MBP is real and heinous, and so is [immune deficiency]. Both need time, attention, and subtle listening and communication skills that many care providers and patients may not have developed, especially under duress."

It's a rule of thumb that parents like Worthen and Connor understand. ■

# Victoria's Miracle Horse

By Tom Russo

This is a story of King Hill Patty Murphy, a spunky miniature horse raised by my friend, Tom Newkirk. Tom is the owner of King Hill Miniatures in Freehold, N.Y. Patty, a 6-year-old silver chestnut, is the lead mare of Tom's herd, but right from the beginning, Patty was a wild child.

Tom was often overheard saying, "I can't do anything with this horse. She never listens to me, raises Cain on the farm and runs around like a demon!"

Then came my 5-year-old daughter, Victoria—Tori. Like many little girls, she is crazy about horses. Tori is also a bright, independent girl with a primary immune deficiency disease called common variable immune deficiency or CVID.

Newkirk, as we call Tom, was concerned about Tori's meeting Patty, but Tori insisted, so let me set the scene: It was a beautiful spring day, and Tori had bugged me all morning about going to see the horses. On this particular day, she was feeling pretty good for a kid who had had 10 severe upper respiratory infections already that year and had just finished what seemed like her millionth round of antibiotics. I couldn't let her down.

When we arrived at Newkirk's farm, he warned, "Watch Patty, she's a ramrod today!" as Tori went into the field and yelled Patty's name. In the distance, we saw Patty raise her head from grazing and bolt toward us like the maniac she was. My wife, Lisa, took Tori in her arms to protect her from the wildly running horse. Directly in front of them, Patty stopped dead in her tracks, gave Tori a sniff, and whinnied softly as if to say, "Hi."

Newkirk looked at me and said, "That horse has never done that for me! I call her and she runs the other way like a rocket shot."

With some hesitation, Lisa put Tori down, and Tori grabbed and tugged on Patty's mane and tail as Patty

just stood there without moving or acting like the nut she usually was. Tori ran around like any 5-year-old and Patty followed her as though she was her own. When the day came to an end, we knew Patty and Tori had bonded in a special sort of way, immediate best friends.

Newkirk was amazed and kept shaking his head. "She never does that for me."

As we were leaving, Tori asked Newkirk, "How much do you want for Patty?"



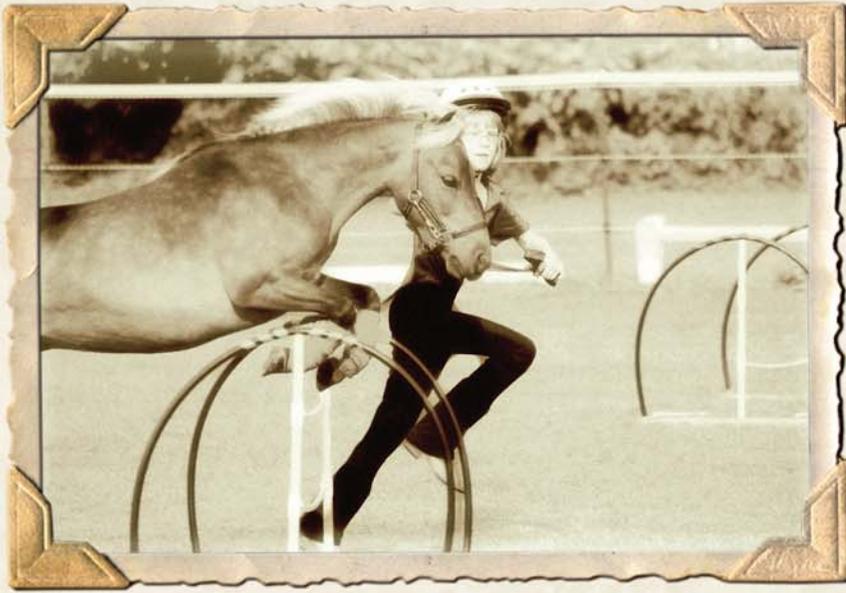
"If Patty follows you down the driveway, she's yours," Newkirk replied.

"Dad, please, can we keep her?" Tori pleaded.

"We have to check where we can keep her, and see if your doctors say it's OK."

As we drove down the driveway, we heard a loud commotion and stopped to see what it was. Patty was trying to break down the gate from inside the pasture and neighing like crazy, as if to say, "Don't leave me."

Again, Newkirk shook his head in disbelief, while Tori beamed with the biggest smile ever. After watching my



brave little girl fight potentially life-threatening infections day after day, that smile was so wonderful to see. It brought us all to tears.

Let's go a little further into time. The day was Tori's sixth birthday. As she was opening her presents, Newkirk showed up and said, "Your parents have another surprise for you out front."

The kids, with Tori in the lead, took off running. Newkirk went to the back of his trailer, and down the ramp came Patty with a huge purple ribbon saying, "Happy Birthday!"

Newkirk put Tori on Patty's back. Patty just stood there as if she had been ridden all her life. Again, Newkirk just shook his head, "These two amaze me every time they are together."

The following year, Tori's illness became worse. It was one infection after another, doctor visit after doctor visit, specialist after specialist. But Tori was always asking to go see Patty. This once-energetic child, now too sick to run around and play, would sit in the field for hours as Patty stood over her listening to her talk and sing. It truly was amazing how these two had bonded.

Let's fast forward again, to when Tori was 7. She heard about a 10-mile trail ride called "Saddle Up for St. Jude's."

Tori said to Lisa, "I want to take Patty on that ride and raise money to help research my disease. Maybe they will find a cure someday." Lisa agreed but reminded Tori that Patty had never really been ridden before and had never been off the farm, either. Tori was convinced Patty would do it for her. In fact, they did a 5-mile ride, raising almost \$2,000 for St. Jude's, while we walked beside them.

When we finished, Newkirk was waiting for us, as usual shaking his head in astonishment at what these two could do together.

Unfortunately, the next few years were very rough for Tori. She was still getting infections every few weeks and was on at least 12 different medications. The side effects left her totally wiped out and too tired to go see her beloved Patty. I would go over to Newkirk's, and I swear Patty would look at me as if to say, "Where's my little friend, Tori?"

We took Tori to Johns Hopkins University Hospital in Baltimore, and went to numerous conferences on her disease until we finally found the right treatment to help

her: time-consuming infusions of intravenous immune globulins. Over the next two or three years, she started to feel better and get much stronger, strong enough to go see her adored Patty again. Boy, did they miss each other!

By the time Tori was 10, she decided she wanted to show Patty in our local Miniature Horse Club events. Of course we told her she could, but not to expect too much as they were both new to showing. Tori replied, "It's OK, Dad and Mom, I just want to show people how great my Patty Pat is."

They were both up to the challenge, and their first year showing they did very well together. At a benefit horse show, Patty was named Grand Champion Show Pony. Then at the end-of-the-year banquet, Tori came away with High Point Youth. Wow! Guess what Newkirk was doing? Yep, shaking his head, smiling, and telling everyone, "I could never do anything with that horse, but she and Tori together are a team."

I have to say that little horse is the best investment I have ever made for both my daughter and my family. Without Patty, I do not know if Tori would have continued to put up such a valiant fight to control her illness, which will require infusions the rest of her life. We are truly blessed to have this miracle horse.

Thank you, Tom Newkirk, for all you have done for my family and daughter. Your farm may not be the biggest, it is probably one of the littlest, but what it lacks in size it sure makes up for in heart and love. Your horses are the best and Patty is the Queen. ■

# The Fight in the Dog

By Mark T. Haggard

Caleb came back with tears falling behind his 7-year-old hands.

"What's wrong?" I asked.

"The football hit me in the face," he whimpered.

"Well, get your hands up," I barked. "If you catch the ball, it won't hit you in the face." That is a basic tenant of football: Catch the ball with your hands, because, if you let the ball hit your body, you are less likely to hold on to it. "It's pretty easy, son; just catch the ball. Now get back into the game."

"I wanna stay out for a few plays. It hurts."

I peered into my son's eyes and proclaimed, "You've been through worse."

Dads are cold. If you want sympathy, go see Mom.

It's not that dads don't love their children; we would do anything to protect them. There is a

double-edged sword to raising PIDD kids, however; we must teach them to be tough, but at the same time, protect their youth. Sometimes PIDD kids learn toughness despite us.

Unrelenting sinus infections plagued Caleb through the first years of his life. After a second sinus surgery, doctors decided to fight his

infections with daily infusions of IV antibiotics via a PICC (peripherally inserted central catheter) line.

Two male nurses took Caleb into a procedure room to place his line. He was given a sedative to make him loopy and forgetful.

It did neither. The medication only made him angry, hostile and out of control. When one of the nurses crawled on to the table to restrain him, my instinct was to body check the man that was trying to help. But intervention was out of the question. This was Caleb's battle.

The nurses ordered me out of the procedure room and continued threading the PICC line through the chest of my combative 3-year-old. I left to screams of "I want my daddy! I want my daddy!" Caleb's pleading echoed through the hallway and melted my heart as I sank to the cold floor. Seconds turned to minutes and minutes turned into the longest hour of my life.

I put my head in my hands and stared at the floor. My ultimate purpose as a dad is to protect my son, and I was slumped over in defeat. I was prohibited from doing what was the nature of a father: coaching my son to conquer the disease that was trying to take over his body.

When the procedure was finished, I was escorted to Caleb's side. Faint from the fight, I kneeled next to the bed, caressed his IV'd arm and asked him to forgive me.

"Hey, Dad," Caleb whispered, "can we get a Happy Meal on our way home?" He had forgiven me.

I have discovered that PIDD kids in general are remarkably resilient. No matter how many times they get knocked down, they still get up off of the mat. They can have their wind knocked out, recover and quickly get back into the game whether we're with them or not. PIDD kids are the fortunate ones: Inner strength comes with their unique challenges, and a disease that insists on systematically weakening their bodies only makes them stronger.

And we are all the better for it.

"Hey, Caleb," I called as he turned toward the field. "Remember: It's not the size of the dog in the fight."

My son stopped, turned around, rubbed his nose and answered, "Yeah, I know. It's the size of the fight in the dog."

Woof! ■



Illustration by Caroline Carlson

# Camp Shangri-La

By Dayna Fladhammer

I like to travel. If my kids' primary immune deficiency diseases (PID) were suddenly taken away, and I regained all the time and money required to manage it, my kids would be experienced travelers by now. I would love for them to have already visited all 50 states, and seen every museum across the country.

Instead, they can tell you what a B cell is, and what it does. Still, I'd rather they were out traveling the world wide, rather than the halls of healthcare facilities.

The truth is, part of the reason we don't travel too far from home is because I'm a worrier. A big worrier. I think about things like what hospital we can go to if they get sick, where the nearest pharmacy is. I worry about how we'll deal with illness on the road, how soon to take them in to the doctor when they start to show symptoms or complain of pain or discomfort. I worry.

Actually, though, the kids probably travel better than I do.

Recently, our family spent a weekend as guests at The Painted Turtle, a Hole in the Wall Gang Camp, in Lake Elizabeth, Calif. The Hole in the Wall Gang Camps were begun in Connecticut by Paul Newman, for children with chronic illnesses. The first camp has expanded to an association of similar camps located across the country.

These camps are for kids who should be out experiencing life at a time when exploring the world is a wonderful and natural thing. Instead, the campers, kids like my own, spend many of their days in clinics or at home sick or worse yet, worrying about being sick. This has been a major concern for our family for the last three years. My husband and I want so much to give our children a normal life. We don't want them to be able to explain the importance of B cells to their peers. We don't want them to be able to instruct their doctors on the best types of needles to use for their veins. We want them to run and play and live!

So, that weekend at camp was a revelation: It was the first time all my worries about traveling with the



kids never entered my mind. Instead, we boated, fished, sang, danced and hiked, did arts and crafts, made new friends, and renewed old friendships. Most of all, we watched what my husband and I value most in life, our children experiencing what we have wanted for them since they were first diagnosed: the freedom to be children.

Any concerns we normally have about medications or healthcare were set aside: Doctors and nurses sat feet away, disguised as normal people, ready to help if we needed it. And we did need it. We forgot the antibiotics in all our excitement to depart for camp.

Instead of calling pediatricians, waiting for a return call, looking for a pharmacy and waiting for medication, we simply walked to the Well Shell, the camp's on-site clinic, got what we needed and walked away.

When Charlie complained for the third time in an hour that his knees and legs hurt, we didn't have to worry all weekend, we didn't have to leave to seek medical care. Twenty minutes in the Well Shell, a little Motrin and a lot of TLC, and we were back on the lake, fishing.

The kids had the time of their lives at camp—they still talk about it—and they frequently ask when we can return. We hope the camp will be held again next year, and we know other camps across the country provide similar programs for kids.

So, until the next time, we'll hang with the friends we made, look at the pictures, and know there is a place out there where the kids can be free to be kids—and we can travel worry-free, even if just for a weekend. ■

## ...Guillain-Barré Syndrome (GBS)

### Websites and Chat Rooms

1. The GBS/CIDP Foundation International, [www.gbsfi.com](http://www.gbsfi.com), has 23,000 members in 160 chapters on five continents. 610-667-0131
2. The GBS Foundation Discussion Forums provide the opportunity to talk to other GBS patients and learn more about ways to manage the illness: [www.guillain-barre.com/forums/](http://www.guillain-barre.com/forums/).

### Online Pamphlets

1. A GBS Fact Sheet is found on the National Institute of Neurological Disorders and Stroke website: [www.ninds.nih.gov/disorders/gbs/detail\\_gbs.htm](http://www.ninds.nih.gov/disorders/gbs/detail_gbs.htm).
2. The Mayo Clinic has an overview of Guillain-Barré syndrome at [www.mayoclinic.com/health/guillain-barre-syndrome/DS00413](http://www.mayoclinic.com/health/guillain-barre-syndrome/DS00413).
3. The National Institute of Neurological Disorders and Stroke has an information page about CIDP: <http://www.ninds.nih.gov/disorders/cidp/cidp.htm>

### Online Peer Support Links

1. GBS & CIDP Discussion Forum—UK Bulletin Board—For Ireland and England: <http://www.gbs.org.uk/cgi-bin/ikonboard3/ikonboard.cgi>
2. GBS Support Group—UK chat room—requires registration: <http://www.jsmarcussen.com/gbs/uk/chat.htm>
3. GBS Foundation Discussion Forums [www.guillain-barre.com/forums](http://www.guillain-barre.com/forums)
4. Yahoo Support Group Discussion Board: [http://health.groups.yahoo.com/group/GBS\\_CIDP/](http://health.groups.yahoo.com/group/GBS_CIDP/)

### Books and Articles

1. "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](http://www.gbsfi.com).
2. "Bed Number Ten," by Sue Baier, provides a view of long-term care through the eyes of a patient totally paralyzed with GBS.
3. "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](http://www.gbsfi.com).
4. "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

1. ITP Support Association, UK: <http://www.itpsupport.org.uk/>
2. Platelet Disorder Support Association: [www.ITPpeople.com](http://www.ITPpeople.com) 87-PLATELET (877-528-3538) or 301-770-6636

### Online References

1. Idiopathic thrombocytopenic purpura [http://en.wikipedia.org/wiki/Idiopathic\\_thrombocytopenic\\_purpura](http://en.wikipedia.org/wiki/Idiopathic_thrombocytopenic_purpura)
2. Idiopathic Thrombocytopenic Purpura: Michael A. Silverman, MD <http://www.emedicine.com/EMERG/topic282.htm>
3. Immune Thrombocytopenia: Current Understanding and Future Research, Robert S. Mocharnuk, MD: <http://www.medscape.com/viewarticle/459249>

4. Infusion Network Systems Article: The Expanding Use of IVIG provided by ZLB Bioplasma, Inc.: <http://www.infusionsystems.net/article-ExpandingUseofIVIG.html>
5. ITP: Idiopathic Thrombocytopenic Purpura: <http://familydoctor.org/113.xml>

## ...Kawasaki Disease

### Websites

1. Kawasaki Disease Foundation: <http://www.kdfoundation.org/>  
PO Box 45, Boxford, MA 01921  
Tel: 978-356-2070 · Fax: 978-356-2079  
Email: [info@kdfoundation.org](mailto:info@kdfoundation.org)
2. Kids Health discusses Kawasaki disease at <http://www.kidshealth.org/parent/medical/heart/kawasaki.html>
3. Overview from the American Heart Association focuses on how the disease affects the heart. <http://www.americanheart.org/presenter.jhtml?identifier=4634>

## ...Multiple Sclerosis (MS)

### Websites and Chat Rooms

1. The mission of the National Multiple Sclerosis Society is to end the devastating effects of MS. <http://www.nationalmssociety.org/>.
2. 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: <http://www.mult-sclerosis.org/index.html>.
3. Multiple Sclerosis Foundation works for a brighter tomorrow for those affected by MS: <http://www.msfacts.org/>.
4. Multiple Sclerosis Association of America seeks to enrich the quality of life for individuals with multiple sclerosis: <http://www.msaa.com/>.
5. MSWorld's Chat & Message Board features patients helping patients: <http://www.msworld.org/>.
6. National Multiple Sclerosis Society aims to end the devastating effects of MS: <http://www.nationalmssociety.org/>.

### Online Peer Support Groups

1. Friends with MS website: <http://friendswithms.com/>  
forum: <http://health.groups.yahoo.com/group/FriendsWithMS/>.
2. My MSViews website: <http://www.mysviews.org>  
forum: [http://health.groups.yahoo.com/group/MSViews\\_Multiple\\_Sclerosis/](http://health.groups.yahoo.com/group/MSViews_Multiple_Sclerosis/).
3. MS Support Group: <http://health.groups.yahoo.com/group/mscured/>.
4. The MS Carousel—A Place to Meet With People Who Understand MS! <http://health.groups.yahoo.com/group/themscarousel/>.

## ...Myasthenia Gravis

### Websites and Chat Rooms

1. The Myasthenia Gravis Foundation of America (MGFA) is the only national volunteer health agency dedicated solely to the fight against myasthenia gravis: <http://www.myasthenia.org/>.
2. Myasthenia Gravis Fact Sheet prepared by National Institute of Neurological Disorders and Strokes: [http://www.ninds.nih.gov/disorders/myasthenia\\_gravis/myasthenia\\_gravis.htm](http://www.ninds.nih.gov/disorders/myasthenia_gravis/myasthenia_gravis.htm).
3. Mayo Clinic's overview of myasthenia gravis: <http://www.mayoclinic.com/health/myasthenia-gravis/DS00375>.
4. The blog at <http://www.ivig.blogspot.com.ca> covers Information related to IVIG treatment of MG in Canada.

# Wanted to Know About...

## Online Peer Support Groups

1. MGFA's Forum: <http://health.groups.yahoo.com/group/MGnet/>.
2. Bette's Myasthenia Gravis Support: <http://health.groups.yahoo.com/group/bettesmyastheniagravisupport/>.
3. Maddy's MG Support: <http://health.groups.yahoo.com/group/maddysmgsupport/>.
4. Autoimmune Information Network Inc.: [www.aininc.org](http://www.aininc.org)  
PO Box 4121, Brick, NJ 08723, 877-246-4900, Email: [autoimmunehelp@aol.com](mailto:autoimmunehelp@aol.com).

## ...Myositis

### Websites

1. 

 <b>THE MYOSITIS ASSOCIATION</b>	The mission of The Myositis Association, <a href="http://www.myositis.org">www.myositis.org</a> , is to find a cure for inflammatory and other related myopathies, while serving those affected by these diseases. 202-887-0088
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2. 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>.
3. 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>
4. 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: [http://www.hopkinsmedicine.org/rheumatology/clinics/myositis\\_center.html](http://www.hopkinsmedicine.org/rheumatology/clinics/myositis_center.html)

### Online Peer Support Links

1. Juvenile Myositis Family Support Network: [http://www.curejm.com/family\\_support/index.htm](http://www.curejm.com/family_support/index.htm)
2. Myositis Association Community Forum: [www.myositis.org](http://www.myositis.org)
3. Myositis Support Group UK: <http://www.myositis.org.uk/>
4. Yahoo Myositis Support Group Discussion Board: <http://health.groups.yahoo.com/group/OurMyositis/>
5. 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](mailto:rgay@socal.rr.com).

### Books and Articles

1. "Coping With a Myositis Disease," by James R. Kilpatrick, is written by myositis patients telling their personal stories.
2. "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.
3. "Living With Myositis," edited by Jenny Fenton, is an accessible, realistic and sympathetic guide to facts, feelings and future hopes.

4. "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.
5. "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](http://www.myositis.org).
6. "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.

## ...Peripheral Neuropathy (PN)

### Websites

1. 

 <b>The Neuropathy Association</b>	The Neuropathy Association, <a href="http://www.neuropathy.org">www.neuropathy.org</a> , 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
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2. To learn about PN, how it is classified, the symptoms, causes and treatments, see the Peripheral Neuropathy Fact Sheet available at <http://www.ninds.nih.gov/disorders/peripheralneuropathy/peripheralneuropathy.htm>.

### Support Groups

1. Click on the Member Services tab of the website, [www.neuropathy.org](http://www.neuropathy.org), for listings of support groups across the nation.
2. The Neuropathy Action Foundation, at [www.neuropathyactionfoundation.org](http://www.neuropathyactionfoundation.org), educates, empowers and informs patients and physicians about neuropathy.

### Online Peer Support Links

1. Calgary Neuropathy Support Group: <http://www.calgarypnrs.org/index.htm>
2. MSN Support Group: Discussion Board: <http://groups.msn.com/PNPARTNERS>
3. The Neuropathy Association Bulletin Board: [www.neuropathy.org](http://www.neuropathy.org)
4. Yahoo Neuropathy Support Group Discussion Board: <http://health.groups.yahoo.com/group/neuropathy/>
5. Yahoo Support Group—Australia Discussion Board: <http://au.groups.yahoo.com/group/LifeWithPN/>

### Books and Articles

1. "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.
2. "Medifocus Guide to Peripheral Neuropathy" is a guide to current and relevant PN research, organized into categories for easy reading.
3. "Numb Toes and Aching Soles," by John Senneff, discusses the symptoms, causes, tests, treatments and coping strategies for peripheral neuropathy.
4. "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.
5. "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. ➤

## ...Primary Immune Deficiency Disease (PIDD)

### Websites and Chat Rooms

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The Immune Deficiency Foundation (IDF), [www.primaryimmune.org](http://www.primaryimmune.org), is dedicated to improving the diagnosis and treatment of PIDD through research and education. 800-296-4433
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The Jeffrey Modell Foundation, [www.info4pi.org](http://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](http://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](http://www.aaaai.org), has a helpful Q&A section on its website, with resources and tips for those with various immune deficiencies.
- Baxter's website, [www.immunedisease.com](http://www.immunedisease.com), offers in-depth information on immunology, PIDD and treatment with intravenous immune globulin. Click on "Europe" to see SCIG information.
- The Michigan Immunodeficiency Foundation, [www.midf.org](http://www.midf.org), seeks to improve the quality of life for Michigan residents affected by PIDD.
- The International Patient Organization for Primary Immunodeficiencies (IPOPI), [www.ipopi.org](http://www.ipopi.org), promotes the worldwide improvement in the care and treatment of PIDD patients.
- To connect to a PIDD message board, go to [www.info4pi.org](http://www.info4pi.org).
- To chat with peers on IDF's Forum, go to [www.primaryimmune.org](http://www.primaryimmune.org).
- Chat with parents of children affected by primary immune deficiency at <http://health.groups.yahoo.com/group/PedPID/>.
- Chat with peers with PIDD 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: <http://www.nepin.org/>.

### Online Pamphlets and Education

- Go to the National Institute of Allergy and Infectious Diseases site at [www.niaid.nih.gov](http://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 [www.niaid.nih.gov/publications/immune/the\\_immune\\_system.pdf](http://www.niaid.nih.gov/publications/immune/the_immune_system.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 or the 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](http://www.prepaudio.org).

### Online Peer Support Links

- 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](http://www.primaryimmune.org/forums/forum_intro.htm)
- Jeffrey Modell Foundation Message Board  
[www.info4pi.org](http://www.info4pi.org)

### 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.

## ...Stiff-person Syndrome (SPS)

### Websites

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

## ...General Resources

### Product Information

- To learn more about Vivaglobin—the subcutaneous immune globulin (SCIG) go to: [www.vivaglobin.com](http://www.vivaglobin.com).
- For more information about the 10% IVIG solution Gammagard Liquid, go to [www.gammagardliquid.com](http://www.gammagardliquid.com).
- Learn about IVIG treatment with Gamunex at [www.gamunex.com](http://www.gamunex.com).
- For information about influenza and the influenza vaccine, visit [www.cdc.gov/flu](http://www.cdc.gov/flu) or call 800-CDC-INFO (800-232-4636).

### Other Organizations

- 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](http://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](http://www.ncqa.org)

# You Ever Wanted to Know continued...

- The nonprofit Patient Advocate Foundation, [www.patientadvocate.org](http://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](http://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](http://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](http://www.keepkidshealthy.com).
- The National Organization for Rare Diseases, at [www.rarediseases.org](http://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](http://www.aarda.org) brings national focus to autoimmunity through research, education and patient services. 800-598-4668
- Band-Aides and Blackboards: <http://www.lehman.cuny.edu/faculty/jfleitas/bandaides/>, started by a nurse working on her dissertation, focuses on children with chronic illness.

## Education and Disability Resources

- Continuation of Health Coverage—Consolidated Omnibus Budget Reconciliation Act (COBRA): <http://www.dol.gov/dol/topic/health-plans/cobra.htm>
- Social Security: [www.ssa.gov/disability/](http://www.ssa.gov/disability/)
- California State Disability Insurance (SDI): [www.edd.ca.gov](http://www.edd.ca.gov) (Please note that each state has a different disability program.)
- IDEA 2004 Resources: <http://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](http://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 [www.ed.gov](http://www.ed.gov) offers a Parents section that has a subsection titled "My Child's Special Needs."
- U.S. Department of Health and Human Services, Office of Civil Rights: <http://www.hhs.gov/ocr/504.html> spells out your rights under Section 504 of the Rehabilitation Act.
- Wrightslaw: <http://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 <http://www.usdoj.gov/crt/ada/adahom1.htm>.

## IG Manufacturer Websites

Baxter: [www.baxter.com](http://www.baxter.com)  
Grifols: [www.grifolsusa.com](http://www.grifolsusa.com)

Octapharma: [www.octapharma.com](http://www.octapharma.com)  
Talecris: [www.talecris.com](http://www.talecris.com)  
CSL Behring: [www.cslbehring.com](http://www.cslbehring.com)

## Pump and Needle Websites

Intra Pump Infusion Systems: [www.intrapump.com](http://www.intrapump.com)  
Repro Med Systems, Inc: [www.repro-med.com](http://www.repro-med.com)  
Graseby Marcal Medical: [www.marcalmedical.com](http://www.marcalmedical.com)  
Norfolk Medical: [www.norfolkmedical.com](http://www.norfolkmedical.com)

## Medical Research Studies

Check out the official website for the National Institutes of Health patient recruitment program. <http://clinicalstudies.info.nih.gov/>  
The Center Watch website provides a wealth of information about clinical trials and volunteer participation. [www.centerwatch.com](http://www.centerwatch.com)

ClinicTrials.com has a registration form to request that you be notified about recruiting for future studies. [www.clinicaltrials.com](http://www.clinicaltrials.com)

WebMD has a service that matches volunteers with trials. [www.webmd.com](http://www.webmd.com)

## ...Nutrition and Food Safety Information

- American Board of Physician Nutrition Specialists: <http://www.ipnec.org>
- American Dietetic Association: <http://www.eatright.org>
- American Gastroenterological Association: <http://www.gastro.org>
- North American Society for Pediatric Gastroenterology Hepatology and Nutrition: <http://www.naspgn.org/>
- Bovine spongiform encephalopathy (BSE) [http://www.aphis.usda.gov/newsroom/hot\\_issues/bse.shtml](http://www.aphis.usda.gov/newsroom/hot_issues/bse.shtml)
- Childhood choking prevention: <http://www.ific.org/publications/brochures/index.cfm>
- American Academy of Allergy, Asthma & Immunology (AAAAI): [www.aaaai.org](http://www.aaaai.org)  
Patient Information and Physician Referral Line: 800-822-2762
- The Food Allergy & Anaphylaxis Network: [www.foodallergy.org](http://www.foodallergy.org) 800-929-4040
- U. S. Food and Drug Administration, Center for Food Safety and Applied Nutrition [www.cfsan.fda.gov](http://www.cfsan.fda.gov) Food Information Line (24 hours): 888-SAFEFOOD
- USDA Meat and Poultry Hotline: [www.IsItDoneYet.gov](http://www.IsItDoneYet.gov) 888-MPHotline 888-674-6854 · TTY: 800-256-7072

## ...Resources 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.
- "This Is 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 <http://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](http://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](mailto:editor@igliving.com). In this case, more is indeed better!*

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