Medical Research
The Who, What, When, Where and Why of It

Sites of Care
Maybe You Prefer Hospital Outpatient?

Not Another Sinus Infection!
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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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A patient recently shared with me some of the myths surrounding immune globulin and why so many providers and patients have trouble obtaining it. Like most myths, these have a touch of the fantastic about them:

1. *Rich people are buying it all up for their elderly parents with Alzheimer’s.* Impossible—there isn’t enough of the stuff.
2. *Doctors are prescribing the IVIG for themselves.* Huh? For what?!
3. *It’s all going to the wounded military in Iraq.* Wrong: The wounded are looking for rehab and jobs, not IVIG.

Although myths could explain the mysteriously elusive IVIG, we do not live in ancient times; we no longer require myths to explain the sun’s rising. Neither do we need them to explain the IVIG crisis; facts will do:

1. Reimbursement for IVIG therapy is inadequate.
2. Prices are inevitably increasing.
3. The current IVIG supply does not meet the growing demand.

They lack a fantastic flair, but they are a lot more sensible—and common sense may be what we need: Since January 2005, when the Medicare IVIG reimbursement methodology took a sharp turn for the worse, a few committed souls have been trying to resolve the crisis. But it persists, now for more than two years, while patients plead for help, while physicians tilt futilely at the marketplace, while the government studies the problem, while bureaucrats wait for directives, while patients’ pleas are unanswered.

What to do? Again, perhaps some common sense will help:

1. IG Living will support the IG community through education, communication and advocacy, striving to be a voice for patients and providers.
2. We will support a new national alliance of patients, physicians, decision makers and industry, dedicated to improving affordable access to immune globulin products.
3. We will help create a national registry of all patients who rely on immune globulin, to document outcomes and demand.

Will you join us? If so, please email editor@igliving.com.

Despite the adversity, we are grateful to see growing recognition of the crisis. For example, with input from Congressman Jim McCrery, one of our long-term committed souls, Congressman William M. Thomas was able to include the following statement in the Tax Relief and Healthcare Act of 2006:

“The House is also concerned by reports that some Medicare beneficiaries have trouble accessing IVIG therapies from providers. It is our hope that the Office of the Inspector General (OIG) and the Office of the Assistant Secretary for Planning and Evaluation (ASPE) studies focused on IVIG are promptly completed. The House hopes the Secretary would promptly review such studies, and report to the House regarding the adequacy of supply and Medicare reimbursement related to the cost of acquiring IVIG and the complexity of IVIG infusions. The House strongly urges the Secretary to continue the IVIG pre-administration fee until the Secretary either assures the House that Medicare reimbursement is adequate or a new payment methodology is implemented to address concerns regarding access to IVIG.”

Unfortunately, we’ve also seen setbacks. In many states across the country, Medicare insurance carriers are establishing local coverage determinations that are reducing patient access to IVIG. We will report in-depth on local determinations in our April-May issue. If you have fallen victim to this trend, please contact us by email at editor@igliving.com or call Kris McFalls at 800-843-7477.

Obviously, this will be a critical year for patients, their families and providers, and we hope for significant progress. In the meantime, we’d like to encourage you to consider the small joys that sustain us when faced with adversity, the perfect joke your infusion nurse shared during your last treatment, the image of your child trying to infuse the cat, the moment of inspiration that resolved that nagging problem of how to conveniently store your infusion stuff at home—and share these moments with us. We all need a good laugh!

Kit-Bacon Gressitt, Editor

Please send your letters to editor@igliving.com.
What IVIG Crisis?
By IG Living Readers

I have myasthenia gravis and Guill Barré syndrome. Medicare has denied payment for five IVIG [infusions] received at home and prescribed by my neurologist. Medicare claims new studies show IVIG has no effect on these autoimmune diseases. Is this true?

I was just cut off from my weekly IVIG treatments (yes, I realize how lucky I am to have received them), to monthly—a 75 percent reduction. I am scared to death I will die from this dramatic reduction.

I have received IVIG infusions since 1994, two healthcare coverages, Medicare and a private insurance carrier. I never had a problem reimbursing until the Medicare Modern 2003. This legislation has made rein IVIG a financial and administrative nightmare for me and my private insurance carrier. I have tried to fight and appeal for life-saving coverage going into my third year. This creates tremendous emotional stress which is a catalyst for my syndrome symptoms—tragically unnecessary.

An investigative article on reduced Medicare reimbursement for IVIG therapy and the consequent withdrawal of hospitals from providing IVIG infusions for patients on Medicare is needed.

I have common variable immune deficiency. I was diagnosed in 1986 and have received IVIG regularly in the medical center outpatient suites until last May, when the medical center moved us to the cancer center. So far, it has not been a good experience. The center threatens to stop giving IVIG all together. Last week they had a shortage of the brand I use.

Please help us raise awareness in the government of the importance of IVIG to the life of patients with immune mediated neuropathy.
Inspiration is something I’ve been thinking about since I first met Laurel Falconer. Age 72, from Alexandria, Minn., with a sparkle in her eye, Laurel caught my attention from the other side of the room at a local chapter meeting of the Immune Deficiency Foundation in the Twin Cities. The sparkle said, “Hey, look at me, I’m a survivor.” I had to get to know her.

Laurel has come to think of living with primary immune deficiency disease (PIDD) as a blessing. “You have an opportunity to be on the cutting edge of new treatments and research,” she says, “and we live in an era when there are help and treatment options.”

Laurel was not diagnosed with PIDD until she was 60. She had a long history of illnesses that included multiple pneumonias, polio, scarlet fever, diphtheria, rheumatic fever, all of the childhood diseases, multiple strep throats, kidney and urinary tract infections, chronic sinusitis and bronchitis, diabetes type II, diverticulitis, rheumatoid arthritis, and now some pulmonary problems.

Why get out of bed when you have all of these health problems? According to Laurel, it’s to swim with her friends! Every morning, Laurel swims with nine others, ranging in age from 45 to 84. She admits that quite a bit of the time they are socializing, but they are in the water and moving. Laurel also spends hours every week making quilts and other items she donates to charities. She grew up in poverty and was taught at a tender age the importance of always helping others. According to her husband, David, she firmly believes that God expects her to use her talents this way.

Even when Laurel is feeling rotten, sewing is her diversion. She shares her warmth, both in personality and in the form of blankets she donates to anyone in need—hundreds of them per year. David says that Laurel seldom lets her illness get her down.

Laurel raised two boys. Their father is now deceased, but at one time, they had 13 kids who needed a place to stay under their roof. One of her sons had constant infections and heart problems, but he refused to be tested for PIDD. Sadly, he died last year, and Laurel has to live with the grief of losing her much-loved son. She claims that she is the oldest parent at grief counseling. On the other hand, she very proudly admits to being a great grandmother to four kids.

Laurel has learned and shared many lessons of how best to live with PIDD. She advises staying away from salad bars, airplanes, large groups and sick kids. She recommends wearing plastic disposable gloves, long-sleeved shirts and a mask while gardening.

Laurel has been particularly fortunate to find doctors she trusts. Her immunologist is in the Twin Cities, and she has a special relationship with her internal medicine doctor, whom she visits every six weeks to stay on top of her long list of ailments. And she finds time to serve on the board of directors of her hospital.

Laurel is happy to declare she has a good life. She has David, her family, and BuddyBoy, her cat. “You do what you have to do,” she says. “If you do what you want along with it, life is tolerable—sometimes downright fun!”

now, sharing and giving, her love of life, Laurel is an inspiration for me. “When I go to bed at night, someone should be better off today because I lived,” Laurel says. I want to grow up to be just like her.
Listen to That Little Voice

I have learned you must always listen to the “little voice” in your head.

From age 43 to 49 the little voice had me seeking help from MD after MD. No one could explain why I was always sick – except that I had “severe allergies.” Many MDs (five allergists, three primary care physicians and a pulmonary specialist) all told me I had allergies. All these MDs tried allergy shots, which made me more congested.

The little voice told me they were all wrong. I did not have “just allergies.”

By the time I had to leave teaching at age 49, I had had pneumonia six times in a 180-day school year. Still, all the MDs insisted I had severe allergies. No one suggested an immune disease until an ENT advised my primary care physician, rheumatologist and allergist that I needed immune system testing. All disagreed.

By now, the little voice had found the Internet. Armed with information, I gave my allergist one last chance to test me. It involved three vials of blood and a two-day response time. In two days the little voice had finally been heard: At age 56, I was diagnosed with a primary immune deficiency. My allergist was stunned.

Always listen to your little voice.

— Betty, Rhode Island

Thank You

I just received my first issue of IG Living and love it. As a CVID patient, I was so impressed that every article and all the information provided was so necessary and useful! Thank you so much!

— Elaine, Georgia

Are There Families Like Mine?

I have thoroughly enjoyed reading IG Living whenever it arrives. I also share it with friends and family. I gave a copy to two of the schools in my district. It really helped me adjust my kids’ 504 plans this year. Many people have commented about the “Lifestyle” section and how interesting it is to read about other families. But, I have yet to read an article that is similar to my family. I am 38 and my daughters are 8 and 9. All three of us have PIDD. We have all been through extremely tough times, just like most families with this, but one of the biggest struggles, when I am not feeling well, is that I still have to be the brave, positive one for my kids. Thank God for my husband, because the only time I can vent my frustration and anxiety is behind our closed door when the kids aren’t listening.

I know there are other families who have gone through many of the same situations, but to date, I haven’t met a family where the parent is just as sick as the kids. Are there many? It is really hard when this happens. I was just in the hospital, and my girls really struggled.

I guess I am writing to say thank you for the magazine and to see if you would try to find some families who are in a similar situation. There is no support group in my area, but hopefully, by the time my children are older, there will be. If not, they certainly have one at home!

Thank you, again!

— Kelliann, Arizona

Editor’s note: Please contact K-B at editor@igliving.com or 800-843-7477 x1143 if you have a situation similar to Kelliann’s and would like to touch base with her.
Medical Research: The Who, What, When, Where and Why of It

In companion articles, two writers share their perspectives on medical research participation. Lauren Gerstmann reports on the history of medical research and the evolution of its safety practices, and she describes the various kinds of studies and their benefits. Terri Cerda focuses on the future of medical research, outlines practicalities to consider before you volunteer and describes current studies you may want to investigate.

The Evolution of Medical Research

By Lauren Gerstmann, MPH

You’re already being poked and prodded by the doctor on a regular basis, so why would you choose to be poked and prodded more as part of a medical research study? In addition to increasing medical knowledge and helping to improve clinical practice, participation in medical research can offer you personal benefit. Medical research, particularly clinical trials, can offer you access to a medical device or drug years before it is commercially available. And, as your health will be closely monitored during your participation, it can offer you a way to obtain medical care for little or no charge without the involvement of your insurance company. But, there are risks involved. Historically, inadequate care was taken to minimize these risks. Government and local oversight has greatly improved the ethics of medical research, but it is still up to you to decide if the potential benefits of your participation justify your involvement.

Early History of Medical Research

Experimental research first began toward the end of the 18th century. Methods were crude but effective, although there was no oversight of the ethical implications of research. For example, Edward Jenner (1749–1823) is known for developing the first smallpox vaccination. His work eventually led to almost total eradication of smallpox in the developed world, but his methods were incredibly risky as he first tested unproven smallpox vaccines on his son and neighborhood children.

At the beginning of the 20th century, the average age at death was 45 years. People were dying at young ages in large part from infectious diseases such as measles, mumps, rubella (illnesses we vaccinate against today). By the beginning of the 21st century, the average age at death had increased to 78 years.1 Vaccinations were discovered through experimental research and, along with increased sanitation, are widely considered responsible for increasing the average age at death by more than 30 years.

Unfortunately, ethical practices lagged way behind the increasing sophistication of research protocols.

Developing Ethical Practices

In the mid-1900s, the public became aware of the darker side of unregulated experimentation when 23 physicians under the Nazi regime went on trial at Nuremberg for experimenting on prisoners. The legal judgment resulted in, among other things, 10 mandated standards to which physicians must conform for the ethical conduct of research. These points are known as the Nuremberg Code.

Although the code was internationally respected, it was not adopted in the United States because it was widely assumed that U.S. research was conducted with a higher standard. Although this was generally true, there were exceptions, including the Tuskegee Public Health Service Syphilis Study, conducted between 1932 and 1972. The study was designed to study the treatment and natural history of syphilis in African-American men. The study subjects did not give informed consent, and, when other researchers confirmed that penicillin could effectively cure syphilis and it became a standard

treatment in 1947, information about the treatment and the drug itself were withheld from subjects.

In 1964, the World Medical Association reinterpreted the Nuremberg Code as the Declaration of Helsinki. Research journals worldwide required that any research published must be in accordance with the ethical precepts set forward in the declaration. As American researchers became more involved with the ethics of research, Congress authorized a National Commission for the Protection of Human Subjects in Biomedical and Behavioral Research to identify the basic ethical principles that underlie the conduct of human research. The commission developed the Belmont Report, setting forth the idea that all medical research should follow three principles:

- Respect for persons, which specifies that subjects must give informed consent to participate
- Beneficence, which specifies that research should have social and/or scientific value
- Justice, which specifies that research subjects should be chosen fairly

The Belmont code remains the standard that researchers follow today. At a national level, it is interpreted by the National Institutes of Health (through the Office for Human Research Protections) and by the Federal Drug Administration. At the local level, all research must be conducted under the guidance of an Institutional Review Board (IRB), a multidisciplinary, objective group that reviews all studies to make sure that they are conducted ethically, legally and within the spirit of the Belmont Report.

When IRBs evaluate a research study, they spend a significant amount of time evaluating the risk-to-benefit ratio. In other words, they determine whether the potential benefit to subjects is worth any risk that the subjects might incur to their health or privacy. Scientists use several different study methods, and some are inherently riskier than others. But generally, the riskier a study is, the more potential for immediate, measurable benefits. If a study is being conducted at multiple locations, a Safety and Data Monitoring Committee will oversee the study at all of its locations, and will make reports to each local IRB, allowing each to evaluate the safety of the overall study.

**Observational Studies**

Some studies are simply observational. In other words, scientists observe behavior or collect questionnaire data, but do not change their subjects’ behavior in any way. These types of studies were used to investigate whether fluoridating the drinking supply caused excess cancers. Observational studies are the least risky studies, involving some risk to privacy but no risk to health. But, while they describe behaviors, it is very difficult to draw any conclusions from an observational study. For instance, there is a well-known study that concludes there is a relationship between fluoridation and cancer. But, when the National Health Service examined this along with 50 other observational studies in 1991, they concluded that other factors (such as smoking patterns and changes in occupational exposures) may have caused these cancers. Despite the fact that cancers increased when fluoride was introduced into the drinking water, observational studies alone can suggest, but cannot prove, that the fluoride caused the increase in cancer. If there are birds flying in the sky every time a pedestrian is hit by a car, we cannot conclude that the birds are causing the accidents. Correlation does not always equal causation.

**Randomized Studies**

The most effective research will cause a change in treatment or behavior and will measure the effect of that change. For us to be sure that the effect we are measuring is due to the change we have implemented, we need to factor out any confounding information. For example, if we are measuring the effect of a nicotine patch on smoking cessation, we need to be sure that any decreases in smoking are due to the patch that subjects are wearing, rather than to a confounding doctor visit where subjects receive smoking cessation counseling.

Scientists do this by designing randomized studies. In other words, subjects are assigned to a treatment or a non-treatment (placebo or control) group by chance. People who are being treated are compared to people who are not being treated. The study should be large enough that the two groups are similar in almost every way, including age, gender and race/ethnicity. At the end of the study, the two groups are compared, and any new changes between them can be explained by the study.

For example, a group of researchers set out to evaluate whether aspirin could help prevent the recurrence of

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colon polyps (benign growths with the potential to progress to cancer). They studied more than 1,000 people who had polyps and randomly assigned them to a low dose of aspirin, a high dose of aspirin or a placebo. Statisticians evaluated all three groups to make sure that there were no other significant differences between them. At the conclusion of the study, the researchers were able to determine that low-dose aspirin can play a role in preventing colon polyps from recurring.4

Kinds of Prevention
Research studies are defined in terms of primary, secondary and tertiary prevention. Primary prevention is anything done to prevent negative health outcomes in the general population. For example, diabetes screening and counseling at a public health fair is primary prevention (although it is not a research study). Secondary prevention is targeted at high-risk groups. Providing colonoscopies to people with a family history of colon cancer is secondary prevention. If you take it a step further, and design a randomized clinical trial to determine whether aspirin will prevent colon cancer in this high-risk group, you are working on a secondary prevention clinical trial.

Tertiary prevention is not what we typically think of as prevention at all. It is the clinical term for treating a condition someone already has. Chemotherapy is a tertiary prevention for cancer. The preferred method for research in tertiary prevention is a randomized clinical trial, but these trials need to be conducted very carefully in order to minimize risks to the participants. There are several ways to do this. A trial of a new medication to treat heart disease might use an established medication as the control (rather than using a placebo control) so that no subjects have to take the risk of going completely off medication. The medication in a new AIDS trial, if proven very effective, might be offered to all enrolled subjects (including those who were taking the placebo) after the study is over.

Weighing the Decision
Because of the ethical guidelines now in place for medical research, every patient who participates in medical research is a volunteer, and should be treated fairly and with respect. When you make the decision to participate, you need to determine your own comfortable risk-to-benefit ratio. Generally, primary prevention studies are the least risky but they will offer the least personal gain. Secondary prevention studies generally carry low-to-moderate risk, and may prevent some individuals from getting a disease. But, the main benefit of a secondary prevention trial is to help at-risk groups learn more about disease and/or disability prevention. As with participation in any medical research trial, be sure to discuss your participation with your doctor in order to determine that it is reasonably safe for you. It is also important to keep in mind that you always have the right to withdraw your participation in the research study without jeopardizing your care by your physician or at your physician’s institution.

Tertiary studies are the riskiest, but can also offer you the most immediate or significant results. For example, tertiary studies have been used to determine the safest and most effective doses of immune globulin therapy, and also to determine the least painful methods of immune globulin administration. But, these types of studies expose you to the most risk: risk of side effects and risk that the drug or device will not work. You need to carefully evaluate whether the potential benefits are worth it.

Read on to learn more about the decision to participate in medical research.

Medical Research: The Decision to Participate
By Terri Cerda

Choosing to take part in medical research is a very important and very personal decision. Before deciding to participate, it is crucial that you learn as much as possible about the study you are considering. Ask questions and become well-informed. Discuss it with family, physicians and research staff. You should have enough information to feel confident when you make your decision—whether or not you enroll.

The following is a list of questions to consider as part of your decision-making process.
1. What is the purpose of the study?
2. Who is conducting the study?
3. Why do researchers believe the experimental drug or treatment will be helpful or effective?
4. Has the medication or treatment been tested before? If so, what was the outcome?
5. What kinds of tests, medications or experimental treatments will be used to gain information?
6. What are the possible risks, side effects and benefits and how do they compare with the possible risks, side effects and benefits of my current treatment?

Primary Immune Deficiency Studies

According to Katherine Groden, research coordinator at the National Institutes of Health (NIH) in Bethesda, Md., researchers face many challenges in finding volunteer participants for clinical research. Because primary immune deficiency diseases (PIDDs), particularly common variable immune disease (CVID), are considered rare, with a limited patient population, it is difficult to identify individuals to take part in PIDD studies. Additionally, the criteria for participation are often narrow and specific, further complicating the recruitment of patients who qualify for the study. In fact, the NIH and other organizations recruiting for research purposes are limited in their ability to contact patients and must rely heavily on referrals from physicians. Nonetheless, patients are recruited, although sometimes slowly, and many quickly recognize the benefits of participating.

Cyn and Drew Olivera of Southern California have learned firsthand the importance of medical research participation. At age 2, their son, Drew, was diagnosed with X-linked agammaglobulinemia (XLA). After taking part in an XLA research study at St. Jude’s Children’s Research Hospital this past year, they learned that Drew has an extremely rare PIDD called mu heavy chain defect.

The Oliveras cited their ability to access specialists not otherwise available to them and their wish to help others as their reasons to enroll Drew, now 6, in the research program. Despite all the blood tests, a bone marrow aspiration and X-rays Drew underwent during the course of the study, the Oliveras have no second thoughts about their decision. They report no adverse or negative experiences. As a result of their participation, Drew will be closely monitored, returning to St. Jude’s every six months for follow-up. Drew sees the entire experience as an exciting opportunity to travel.

“We need to be a resource for each other. There may be just one unique thing about Drew that gives insight for many others and their fight to gain information [about primary immune deficiencies],” states Cyn Olivera. The Oliveras have participated in one other research program, administered by Dr. Robert Roberts at UCLA Children’s Hospital, Department of Allergy and Immunology. In the future, they plan to enroll in other studies that relate to Drew’s disorder and satisfy their desire to help others, but only after weighing the risks versus the benefits.

“We look at the bigger picture; it’s not just us dealing with this disorder.”

Over the years, advances made in the knowledge base regarding PIDD have been greatly enhanced through research since the identification of “hypogammaglobulinemia” by O.C. Bruton in 1952.1 Many researchers and immunologists readily admit there is still a great deal to learn about the more than 120 disorders classified as primary immune deficiency diseases. The number of identified disorders is sure to grow. There remain unanswered questions concerning the role of genetics in these disorders and adequate and appropriate ways to treat or prevent them in the future. Finding answers depends on the ability of researchers to gain the participation of individuals seeking change and improvement in the ways PIDD is diagnosed, treated and prevented.

If you ultimately decide to participate in medical research, you have likely determined that the potential benefits outweigh the potential risks. Your decision also represents, whether conscious or not, a commitment to help other patients who will benefit from the knowledge derived from the research. ◼

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Studies Recruiting Patients

The following list is a small sample of the current studies that are recruiting volunteer participants for PIDD protocols. For information about any of these studies, contact the National Institutes of Health, Patient Recruitment and Public Liaison Office at 800-411-1222 or email prpl@mail.cc.nih.gov and include the study identifier in your inquiry.

**Developing Newborn Screening for Infants With Primary Immune Disorders**
- **Identifier:** NCT00113464
- **Sponsor:** National Human Genome Research Institute (NHGRI)
- **Purpose:** There is currently no screening process for infants that identifies immunodeficiency disorders at birth. The intent of this study is to develop a screening test based on the current dried blood spot tests used for screening other disorders at birth.

**Genetic Analysis of Immune Disorders**
- **Identifier:** NCT00001467
- **Sponsor:** National Human Genome Research Institute (NHGRI)
- **Purpose:** This study is designed to identify the genes responsible for certain immune disorders, learn about the medical problems they cause, and predict the likelihood of developing the disorders or passing them to offspring.

**Immune Regulation in Patients With Common Variable Immunodeficiency and Related Syndromes**
- **Identifier:** NCT00001244
- **Sponsor:** National Institute of Allergy and Infectious Diseases (NIAID)
- **Purpose:** This study will explore the cause of CVID and other related PIDD syndromes, including IgA deficiency, Hyper-IgM syndrome, thyma, agammaglobulinemia, hypogammaglobulinemia and others to find ways to correct the underlying defect.

**Genomic Analysis of Immune Abnormalities in Patients With Common Variable Immunodeficiency With and Without Gastrointestinal Symptoms**
- **Identifier:** CT00015431
- **Sponsor:** National Institutes of Health (NIAID)
- **Purpose:** This study will determine whether people with CVID, with and without GI symptoms, have inflammation or loss of function of the gut and changes in immune cells and chemicals in the blood and gut. Many patients with CVID have stomach and intestinal problems that may include chronic diarrhea, absorption issues and intestinal infections caused by bacteria.

**Studies of Disorders in Antibody Production and Related Primary Immunodeficiency States**
- **Identifier:** NCT00266513
- **Sponsor:** National Institute of Allergy and Infectious Diseases (NIAID)
- **Purpose:** Research will investigate the gene abnormalities in PIDD, including X-linked Hyper-IgM syndrome and NEMO-associated immune deficiency.

**STA-5326 Mesylate to Treat Gut Inflammation Associated With Common Variable Immunodeficiency**
- **Identifier:** NCT00263237
- **Sponsor:** National Institute of Allergy and Infectious Diseases (NIAID)
- **Purpose:** This clinical trial will determine if the medication is safe to use in CVID patients who have gut inflammation. It will determine if patients taking this medication show improvement.

In addition to the studies being conducted through NIH, there are numerous studies at academic institutions across the country. Ask your immunologist for additional information about these types of research opportunities.

**Two PIDD Studies at UCLA Medical Center will soon be enrolling patients**

1. **Eligibility:** PIDD patients 13 to 75 years old who already receive intravenous immune globulin (IVIG) or subcutaneous immune globulin (SCIG). Participants will receive intravenously an IG product, already approved for IV use in the United States, for three to five months. Then they will be treated with the same product administered subcutaneously (adjusted to a weekly dose) for another six months.
2. **Eligibility:** PIDD patients 3 to 75 years old already receiving immune globulin. Participants will receive an IG product intravenously every three to four weeks for a year. The product is already approved in Europe.

The IG products being tested and medical visits for both studies will be free to the participants who will receive some monetary compensation for their participation. For more information, contact Dr. Robert Roberts at roberts@mednet.ucla.edu or leave a message at 310-825-6777.

Online Search for Studies

The following websites list clinical research protocols that are currently recruiting in addition to detailed information regarding the purpose, criteria and requirements of specific studies. These websites provide visitors with the tools to search for studies that involve specific disorders.

http://clinicalstudies.info.nih.gov/

Check out the official website for the National Institutes of Health patient recruitment program. This site provides summaries and criteria for studies as well as the ability to search for studies being conducted for a specific disease or disorder.

www.clinicaltrials.com

This site has a registration form to request that you be notified about recruitment for future studies.

www.centerwatch.com

This website provides a wealth of information about clinical trials and volunteer participation. It gives you the ability to specify the disorder you are interested in, the location of the study, and the medication names or research protocols.

www.webmd.com

WebMD has a service that matches volunteers with trials. There is an online questionnaire to complete and you will be notified via email of upcoming studies that match the criteria of your questionnaire. You can also search for specific studies.
One of the many uses of immune globulin is the treatment of idiopathic thrombocytic purpura, commonly known as ITP. ITP is an autoimmune disease. Unlike a primary immune deficiency disease, in which the body lacks an immune function, an autoimmune disease causes the immune system to attack itself, or part of itself, believing it to be foreign. ITP is a diagnosis of exclusion, meaning other explanations for low platelet count must first be ruled out, such as side effects from medication, leukemia, metastatic cancer and lupus. The name alone explains much of the disease:

   Idiopathic—cause unknown
   Thrombocytic—a decrease in blood platelets
   Purpura—excessive bruising

**ITP Symptoms**

ITP occurs when antibodies attack platelets, the cells that control bleeding. The most obvious signs are excessive bruising and petechiae, tiny red dots that look similar to bruises or a rash under the skin. These symptoms indicate a low platelet count. However, they are not the only possible symptoms. Sometimes bleeding, internal or external, may occur, especially if the platelet count has fallen below 50,000 (the normal range is 150,000 to 450,000 platelets per microliter of blood). With children, the onset is usually sudden; conversely, adults often have a gradual onset of the disease.

**ITP Population**

Adults account for about 60 percent of all patients and tend toward a chronic ITP disease state. Adult females are affected more than adult males by almost 3 to 1. In children, males and females are affected in even numbers, and 80 percent to 85 percent of the cases resolve within one year and never return.

**ITP Effects**

The effects of ITP on patients range from mild to severe, the most common effect being dependence on one’s platelet counts to determine life activities because of the risk of hemorrhage. When the platelet count falls below 50,000, the risk of internal bleeding is high, so the patient must use extreme caution to avoid injuries, especially head injuries. Some patients at this point are required by their doctors to wear protective gear to protect their heads, elbows or knees. Additionally, patients must avoid contact sports or any activity that may put them at increased risk of injury. Many patients experience depression or anxiety as a result of their condition, and they also report fatigue.

**ITP Treatment**

The severity of the disease varies from person to person and often dictates the treatment course. Many patients are able to sustain themselves with close medical attention and no medical intervention. For cases that require medical intervention, possible treatments include steroids, which in most patients is the first line of defense; IVIG, which increases the platelet count for about a month, and may need to be repeated (IVIG treatment for ITP is an on-label usage, meaning it is approved by the FDA); and anti-D treatment. IVIG and anti-D treatments are often used when steroids fail to raise platelets or when the case is more severe. Other drug therapies include antirejection drugs; however, these are often avoided except in severe ITP cases because of the severity of the side effects. Splenectomies may also be considered, because the platelets are most often destroyed in the spleen. Chemotherapy is a last resort, and is almost exclusively used for adult patients with life-threatening cases.

If you have been diagnosed with ITP, you may feel isolated and frightened, but there is a lot of information available and opportunities for networking with other patients (see ITP Information and Resources). Consider visiting the ITP support group at www.ITPpeople.com, and get informed.

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**ITP Information and Resources**

Platelet Disorder Support Association: www.ITPpeople.com


Infusion Network Systems Article: The Expanding Use of IVIG provided by ZLB Bioplasma Inc http://www.infusionsystems.net/article-ExpandingUseofIVIG.html

ITP: Idiopathic Thrombocytic Purpura http://familydoctor.org/113.xml


Insurance Definitions Gone Wild

By Cheryl Haggard

ew Year’s resolutions tend to be just another way to self-inflict pain, and this year is no different. However, my resolution had nothing to do with eating less chocolate or adding one more lap to my mall walks. No, in fact, my resolution was to wrap my brain around the true meaning of our insurance claims—that’s real pain.

So, in order to soften the blow to my insurance-impaired cerebrum, this is what I was able to come up with.

EOB: Estrogen On Board: The uncanny ability my insurance company has to send a denial letter at the height of my monthly hormonal schedule.

COBRA: Cheryl OverBoard, Requesting Attention: This is most probably reflective of EOB (see above)—and the only source of attention I’ll acknowledge must be chocolate-covered.

Appeals Process: One method your insurance company employs to delay payment for your claim while they consider a good reason to deny your claim.

Network Savings: This is the reduction in boredom achieved by the Cartoon Network while your PIDD kid is being infused and you are worried about who is paying for it.

Amount Paid: The junk food tab parents of PIDD kids must shell out on infusion day. If the PIDD kid needs prednisone, triple the bill.

Copay: The physical and emotional energy spent by a parent of a PIDD kid while trying to force pureed antibiotics down their throats or when coming at the poor kid with an IV needle.

Deductible: A potentially creative place for a PIDD kid to hide upon discovering that he or she is heading to the part of the hospital that does lab draws.

Deductible Status: The successful attempt by the PIDD kid to duct tape him- or herself to the examination table upon finding out that, indeed, someone is going to draw blood to test his or her IgG level. (Not to worry—this gets easier with age!)

Charges: The actual amount on your credit card statement, reflecting the bribery gifts purchased in the hospital gift shop, while your PIDD kid recovered in the hospital from a bout with rotavirus.

Service Provider and Claim Information: A bill you received with the name of a pathologist or lab you have never heard of or remember ever seeing. Then it dawns on you at 3 o’clock in the morning that it is probably the name of the lab and/or pathologist to which your last blood draw was sent. Then you are irritated and cranky the rest of the night because you can’t fall back to sleep, worried over a regrettable voice mail you left with your insurance’s customer service department.

Provider Charges: Amount billed by the unselfish people in our PIDD community, for use of their own blood, time, expertise and energy on behalf of the immune compromised. The provider usually writes the charges off, as they are some of the most generous and kind-hearted people you will ever meet.

So there you have it, folks. While I attempt to find the delicate balance between the insanity of our difficult-to-treat diseases and the insurance industry that is desperately trying to keep things simple, sometimes all I can do is laugh.

And, if laughter is the best medicine, I am willing to do everything I can to meet my family deductible and make my copayments.
My daughter, Kate, spent 15 days in the hospital a few months back, and an additional week at home with a PICC (peripherally inserted central catheter) line, and I learned things in the hospital—a lot of things.

I learned I never want to be in the room when my child goes under anesthesia. Take her away, and then put her under. It’s scary! Frankly, I’m not a big fan of watching her come out of anesthesia either.

I learned my daughter is going to be a pleasant drunk person. No kidding! When she was coming out of surgery, my little girl could not tell me enough times how much she loves me and how great I am.

I learned that kids regress after spending that long in the hospital. Suddenly, Kate’s afraid of the dark. (I would be too if I got poked and prodded in the middle of the night as much as she did.) She wants me to sleep right next to her every night. (Well, I did, for 15 days, in a twin bed – why do hospitals do that to parents?)

I learned if they say it will be three days inpatient, I should pack 10 days worth of clothes and underwear. And, if they tell us that we’ll be going home the next day, ask, “Unless what?” Chances are, whatever can go wrong, will.

I learned that being the world’s most compliant 3-year-old during medical exams gets you a ton of attention, including the entire medical school class coming by to watch your examination; the attending medical students will find it funny when a 3-year-old tells the chief resident he can’t touch her yet because he hasn’t washed his hands; and the chief resident will not find this amusing at all.

I learned too much fun time in the hospital leads to a kid who doesn’t want to leave. Yes, my child cried because she didn’t want to go home. She much preferred playing with the volunteers in the playroom to sharing toys with her brothers.

I learned it is entirely possible to put on weight while eating hospital food, despite the popular belief that it’s inedible.

I’ve learned 3-year-olds, no matter how much medical knowledge they have, still think they’re sick because they did something wrong. It breaks my heart the first time I hear it, and every subsequent time, too.

I learned my children cannot handle being separated for weeks on end, and any desire I have to protect my boys from the germs in the hospital will be outweighed by their need to see their sister and know she is OK.

I learned I trust our doctors: They know a ton, and they love and want the best for my kiddo. But, I also learned that we would never survive without our clinic nurse and our child life specialist.

I learned when one child has a medical need, and I do everything I can to meet it, it doesn’t matter how much I know I’ve done the right thing, I feel guilty for not being there for my other children.

I learned nothing makes me appreciate my child’s disease more than a child with a terminal illness.

And finally, I learned that time gives me perspective. When things seem really low, and I wonder how I’ll make it through, a little time will shed light on the darkness.
My son loves all things military. Give him a stick; it becomes not a gun but an M-16 rifle. Give him paper; he folds it into not an airplane but an F-18 Hornet. Give him Legos; he will build not a tank but an M1A1 tank. His favorite television station is the Military Channel. His favorite DVD is anything Blue Angels.

“Dad,” my son bellowed from his bedroom early on the morning of his latest immune globulin infusion. “Yes, Caleb,” I muttered trudging up the stairs.

“Where is my camouflage shirt?”

That was an odd question from Caleb: Every shirt he owns is camouflage. I guess he just couldn’t see them in his drawer. “Here, son,” I said, pulling one out from his drawer. “Cool!” he said, with an additional, “Thanks, Dad.”

I took a closer look at him. After slipping his shirt over his head, he donned a camouflage hat. He pulled the hem of his shirt over the waistline of a pair of camouflage shorts. When he turned around, I noticed the coup de grâce of his outfit: His face was painted in numerous shades of my wife’s makeup to match his garments.

I’m glad I mowed the lawn yesterday, I thought, or we’d never find him.

Nurse Nancy arrived a few minutes later, and started the process of getting my son hooked up. When it was his time, he stood at attention before her, giving her a sharp salute, then marched to the overstuffed chair in our living room. With snap and precision, Caleb pulled his shirt up and exposed his port.

“Oh no,” I muttered when I saw that the foundation, eye shadow and rouge that was applied to his face was also applied to his chest and belly. “God bless Mary Kay.”

My soldier sat at attention, stoic, as Nancy cleaned cosmetics away from the site of his port, then stuck a needle into his frail, but confident body. Once accessed, Caleb made one last request: “The Military Channel.”

“You bet,” I replied with a broad smile, “whatever you want.”

I left the room to the cheers of, “Cool—Air Force Thunderbirds!”

What a far cry from previous years. My son started IVIG when he was 2 years young. Terrified, he fought with every ounce of his toddler strength to avoid being stuck with a needle, no matter how cleansing the medication was to his body. I was usually chosen to get him into a submission hold so the nurse could access him. When the ordeal was finished, I was often damped with sweat and my muscles ached; it’s amazing how strong a 2-year-old can be when trying to avoid torture.

In more recent days, Caleb has donned the war motif in his fight against his primary immune deficiency disease. Usually that means dressing in blue and holding his miniature Blue Angels, but today he is an Army Ranger.

Caleb realized something long before I did: He is in the fight for his life, for the rest of his life; a fight against germs, viruses and other pathogens intent on killing him. Originally I thought his sicknesses were a mere nuisance; they are much more than that. We are in a war; a war that will require every available resource to win. It has taken a great deal of the resources of my family. Fortunately, we have allies in the immune deficient community, and immunologists at numerous hospitals to help defeat this shadowy, unseen enemy.

Later on in the day, I was in an upstairs bedroom when I saw a flash of camouflage cross the front lawn. My young soldier, armed with a bright green squirt gun, rolled from one unseen rock to another. I opened the window, raised my fist to my mouth and called, “Ksht, come in Lieutenant Caleb, what is your position? Over.”

“I’m at war, Dad.”

I nodded and quietly whispered, “Yes, you are.”
Once every three weeks, Clydean McCann gets in the car and drives half an hour to Northwest Hospital in Seattle. McCann, 76, a retired newspaper woman, must receive four to six hours worth of infusions to deal with cancer and a primary immune deficiency disease (PIDD).

And, frankly, she says, she wouldn’t want to do it anywhere else.

“I prefer to stay in the hospital,” says McCann, who hasn’t let her treatment interfere with a hectic schedule that keeps her busy well past dinner time. “They can monitor my temperature and blood pressure more conveniently, and all the equipment is there so I don’t have to have any of it at home. And, there aren’t a lot of disturbances, which can happen if I do it at home. What happens if the phone rings or the dog wants to play?”

McCann is part of a patient group—perhaps the largest group of infusion patients in the country—who receives infusions in a hospital outpatient setting. For, while doctor’s offices, infusion suites and homecare gain in popularity, there is still a large demand for hospital outpatient infusions. Some of the demand is motivated by financial considerations, as some hospitals and health systems see infusions as a welcome revenue source in an era of tighter budgets. Some of it is insurance-related, with many carriers preferring that higher-risk patients receive infusions at the safest possible location, while others prefer the least expensive setting. And some of it, as with McCann, is personal preference.

“Every person is different,” says Curtis Pease, 23, a Seattle-area shipyard worker who has been receiving infusions for his PIDD for some 20 years—first at the hospital, and today at home. “In some ways, the hospital is easier, especially with insurance paperwork. And in some ways, doing it at home is easier. It all depends on what the person feels most comfortable with.”

Hospital Overview

Some patients, of course, must have their infusions in a hospital, says Michelle Kromelis, RPh, the director of pharmacy services for Children’s Medical Center in Dallas. These are patients who require critical care or who may be getting an infusion for the first time and no one—patient or physician—is quite sure what the reaction will be.

In addition, says Kromelis, whose hospital’s outpatient unit does as many as a half-a-dozen infusions a day, some doctors prefer the hospital setting. For example, it helps with compliance, since, if it’s done in the hospital, the doctor knows it has been done. Also, she says, it’s often easier for new patients, who have to go to the hospital for an appointment anyway, to combine the two trips.

“For some people, it’s just more convenient,” says Theresa Gettman, BSN, who works with infusion patients at Children’s Hospital & Regional Medical Center in Seattle. “It’s one thing to do subcutaneous at home, but it’s very hard to get IV access if you’ve never done it before.”

Although many insurers are trying to move patients away from hospital infusions to cut costs, some are not. In fact, says Kromelis, it’s not unusual for some insurers to ask physicians at Children’s to handle the infusion at the hospital. In one recent case, she says, “It was about the controlled environment. The insurer was looking at the risk perspective versus reimbursement, and wanted to reduce risk.” Plus, it’s much easier for patients who dread paperwork to let the hospital handle it, which is usually the case in an outpatient setting.

In addition, as infusions become more common and as new therapies are developed and new drugs discovered, hospitals see an opportunity to serve their communities and improve their financial situation. In McAllen, Texas, Texas Children’s Cancer Center in Houston and Baylor Medical School worked with local authorities to expand the local infusion center this fall. The upgraded center means most of the children who need hospital infusions don’t have to travel to San Antonio, 240 miles away.

Meanwhile, the largest public hospital in Alameda County, Calif., Highland General, opened an outpatient infusion center in October as part of its $23 million effort to boost revenue, reduce costs and improve patient access to therapies. Hospital officials say the center reflects a growing national trend of treating cancer patients on an outpatient...
basis, as breakthrough drug therapies turn once-terminal diseases into chronic conditions. The hospital could save thousands of dollars in drug costs—and free up hospital beds—by setting up the outpatient center.

The Personal Touch

All of this, of course, doesn’t matter much if the patients aren’t comfortable in their surroundings. And by and large, those who give and receive hospital infusions say they are. The hospital centers are not sterile, off-putting places. The McAllen, Texas, facility has brightly colored walls and what officials describe as an art nook. Pediatric patients leave with a toy, so they’ll have one less reason to be scared. At John T. Mather Memorial Hospital in Port Jefferson, N.Y., patients can watch television or listen to music. At all times, a staff member is nearby in case something happens that shouldn’t.

For many children, the idea of comfort and reassurance is an even more important consideration. At Seattle’s Children’s, says Gettman, one young girl would make a day of her infusion session. She and her grandfather would go to lunch, have fun and spend time together that they might not have otherwise. The experience not only made her brothers and sisters envious, an interesting development given the reason for the visit, but it gave her and her grandfather something that Gettman describes as special.

Also important in a hospital setting: The security given to patients who might be afraid of their infusions or who may not quite understand what is going on. One 4-year-old patient, says Gettman, had moved to home infusions, but the process didn’t go well. He would run around the house, and his mother and the home healthcare nurse literally had to chase him to get him to sit still. On the other hand, when his family brought him back to the hospital, he sat quietly through the procedure. “It’s like they have a sixth sense,” Gettman says. “It’s not so much about the environment as it is they know there are people there they can trust.”

Yet, having said this, hospital infusions are not for everyone. For every patient who finds a visit pleasant and reassuring, there are others who don’t want to make the trip or who find their time is better spent at home, where they are in a familiar setting and around their own things.

“If you live close by or you don’t mind the commute, or if you have a lot of checkups or you’re in a study, then the hospital is OK,” says Pease. “But if it’s not, then you want to do it at home. When I heard about subcutaneous [infusions], and that I could do it at home, I said, ‘That’s for me.’” In fact, Pease—and brothers, Jeff, 20, and Mitchell, 16, who also require infusions for immune deficiencies—have been infusing at home for several years. It was, not surprisingly, a question of logistics. It became too time-consuming, says Curtis, to drive everyone the 35-mile round trip.

But, as Pease emphasizes, that was his experience. It might be entirely different for someone else. And, as McCann demonstrates, there are plenty of examples of that someone else. “I just don’t like to do it at home,” she says. “The time just seems to drag and drag. And, if I’m at home, there are things I’d rather do than get an infusion. I have the same four nurses I’ve had for years. It’s comfortable and comforting, a very nice situation.” And, a situation that doesn’t seem to be going away, despite all of the other changes in the way healthcare is administered in this country.

Says Kromelis: “If someone is that sick and it can’t be done anywhere else, … then this is the environment that everyone wants it to be done in. It’s clean, it’s safe.”

Which is why hospital outpatient infusions are still a viable choice.

For some objective criteria to consider when choosing your site of care, consult the professional care guidelines provided as part of the IVIG toolkit on the American Academy of Allergy, Asthma and Immunology website at www.aaaai.org.
**Let’s Talk!**

By Shirley German Vulpe, EdD

*Shirley:* Thank you for agreeing to this interview. Please tell me about your illness.

*Vicki:* My greatest goal in telling my story is to inspire other PIDD patients to never give up on their hopes and dreams. They can accomplish their goals, live productive lives and have some fun along the way.

Because of my health problem, I know that I really appreciate the healthy times and treasure how precious life is each day. I was sick from the time I was born. My older sister remembers me constantly being on penicillin. In high school, one year, I was out sick over 30 days, and still managed to graduate a year early.

My father died when I was 16, and his last words to my mother were, “Find out what is wrong with Vicki.”

In college, I changed my major from music to education due to constant battles with bronchitis and sinusitis. I married and started teaching. I caught every illness the students had at school. I was well, after my first sinus surgery (one of five), and became pregnant. My two pregnancies were great. I felt better than ever!

Then, after both children were born, I was too sick to return to work. I lived on antibiotics.

*Shirley:* Have you had help?

*Vicki:* Yes, I’ve been so blessed with a supportive husband and children. My daughter studies nursing, so she can help me.

My doctors thought allergies caused my illnesses, and I always “looked too healthy.”

Eight years ago, a nurse friend told me to go to a hematologist, who said the magic words: “I think I can help you.” He told me I had a primary immune deficiency, and started IgG infusions (IVIG). I improved enough to go back to work. However, I still continued to battle illnesses.

*Shirley:* What did you do then?

*Vicki:* We started researching my illness. My husband found the Immune Deficiency Foundation (IDF) website. We received the newsletter and attended a family retreat. I was impressed with the speakers and so happy to finally meet people with the same health issues. I also met representatives from NuFACTOR, who sincerely want to help people with PIDD. Next, I attended a national IDF conference and learned about subcutaneous (SubQ) infusions of IgG. I found a local immunologist, Dr. Kathy Liddle, who said I was a perfect candidate for SubQ. She worked with NuFACTOR to organize training. My principal, accommodating my disability, allowed me to leave work early for the training.

*Shirley:* Great! How has it been going?

*Vicki:* Vivaglobin has been an answer to a prayer. I have gone six months without a sinus infection. A record for me! The quality of life I have now means so much to me and my family. My father’s wish has come true for his “little girl.” I am greatly blessed!

In this sixth column of Let’s Talk, I interviewed Victoria Bowen McCallum, an elementary school teacher from South Carolina. She has a primary immune deficiency disease (PIDD) and is thankful, after 49 years, to have finally conquered her chronic health problems. Persistence, a supportive family, a friend who is a nurse, her school principal, the Immune Deficiency Foundation (IDF) and a home healthcare company were the keys.

**Resources**

Immune Deficiency Foundation can be reached at www.primaryimmune.org and 800-296-4433.

The Americans with Disabilities Act of 1990 protects 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.
Three-year-old Jonathon is a beautiful child. He has had some medical problems, and was conceived through in vitro fertilization.

When he was disobedient, his mother meekly and ineffectually corrected him. In the course of an exam, she said to me, “He is our miracle child.”

I acknowledged that he was a wonderful boy, but asked if she felt reluctant to discipline him or maintain firm limits.

“He gets everything he wants,” she said, with a big smile on her face.

Grandma, who was in the room, looked at me and rolled her eyes.

A child who has had a significant medical episode or condition is at risk of being treated as a “vulnerable child.” Described in 1964 by a pediatrician at Brandeis University, the term refers to an exaggerated perception of a child’s illness, fear that the disease will come back or fear that the child has an unusual susceptibility to disease or death. Vulnerable child syndrome can lead to various problems, including parents and children being unable to separate from each other, overreaction and overprotectiveness on the parents’ part, an inability to set age-appropriate limits, and parents tolerating misbehavior and even abuse from a child. Much of this is based on a parent’s understandable anxiety.

An infant who was jaundiced or premature at birth, or a child who was previously hospitalized, may be brought into a doctor’s office or an emergency room with inappropriate frequency because the parent is frightened by the previous experience. The parent may insist that the child be seen immediately for the slightest ailment and request unnecessary tests or may not allow anyone else to baby-sit for the child. One study a number of years ago found that 40 percent of parents whose children were diagnosed with an innocent heart murmur continued to restrict the child’s athletic participation into early adolescence.

A child who is treated this way may have sleep problems or hyperactive behavior. The child may become an under-achiever, reinforcing the parent’s fear about the child’s ability to cope with life. Additionally, perceiving a child as more vulnerable can prevent effective control of a child’s behavior.

If you are reading this, your child most likely has a significant medical problem that requires immune globulin (IG). Whatever it is, it is not trivial, and you have
every right to be concerned about your child’s health.

Nonetheless, the purpose of and wonderful property of IG is that it helps your child live a normal or near-normal life. A child with thrombocytopenia (low platelets) whose count is normal should be allowed to climb a tree or jungle gym and, yes, risk a broken arm. A child with an immune deficiency that is well controlled on IG should be kept away from children who are obviously ill, but cannot and should not be kept from socializing with other children, germ-laden as they might be.

Certain common-sense precautions need to be taken, of course. A small daycare with a handful of children is a better choice for children with primary immune deficiencies than a large center. Children with thrombocytopenia should probably not play football. (I don’t think any child should but that’s another story.)

But, when raising a chronically ill child, common sense can sometimes be difficult to define. Are you overprotecting your child and creating a vulnerable child situation?

Ask yourself if you have trouble with separation issues. Are you overly concerned with your child’s bodily functions or unable to set firm disciplinary limits because she has “gone through so much”? Is your child’s behavior out of control? Are there sleep problems? Is your child clingy and afraid of the world? If you think you may be falling into this trap, your doctor or sometimes a therapist or other mental health professional can help you deal with it.

As parents, our goal is to help our children become effective and successful adults. Depriving them of independence and the chance to learn the rules through discipline can prevent or delay that. We want them to develop the courage and self-esteem to face the world confidently, not cower in fear.

If you think you might be overprotective, talk with your child’s medical team. Your child’s doctor should be aware of your level of confidence, how much support you need, and help you and your child lead as normal a life as possible. A referral to a therapist might be helpful if you are having trouble coping. Perhaps you need some clarification about what is appropriate care and what is too much. Most important, you need to examine your own anxieties and do all you can to avoid imposing them on your child.

Life is what we make of it. So, when it comes to having a chronic disease, what sort of life does that make? I mean, are we our diseases? I want to say no, absolutely not. I am much more than a disease. On the other hand, my disease does dictate what I do and don’t do.

Before I was diagnosed with any of my illnesses, I could be in a crowded room without panicking; I’d put my hands all over any railing; and I didn’t think twice before eating without washing my hands or hugging a sick person. I ate anything I wanted, maybe to an unhealthy degree at times, but I was just having fun. I suppose I was carefree or careless, depending on how you look at it.

I can’t imagine living that lifestyle now. It wouldn’t be any life to live. I would feel terrible, constantly fighting germs. So I guess that is where I took control. I control my choices—and isn’t control something we all strive for?

As a young adult, control is the one thing that has sometimes seemed the farthest out of reach. There are so many things that I want for my future. I have them all planned: where I will live and go to school, who I will marry, what my job will be, how many kids I will have, even what my dog’s name will be. Although having my life planned out gives me some relief, I know most things don’t go as planned: I don’t have control over my common variable immune deficiency or my lung disease. But in some ways, I have all the control I need. Nothing is out of reach. I can live where I want. I can go to school where I want. I will marry the man who is right for me. My job will be whatever I want it to be, whatever makes me happy. I will have kids when I am ready. And when I get that cute dog of mine, her name will be Moxie—no ifs, ands or buts!

No, I don’t have control over my diseases, but I believe that having my life threatened by them, potentially taken from me, makes me experience life with more intensity, more appreciation. This great life may be taken from me at any time, or at least the quality may be jeopardized, so it behooves me to live it to its fullest every moment, to be a risk taker, to survive and enjoy life. None of us knows when our last day will be.

While I was working on this article, I asked my dad to read it and tell me what he thought. He was just about done and he turned to me and asked, “Are you trying to break my heart?”

This got me thinking: If someone I am so close to could get so sad just by reading a few words, then why am I not sad? He is affected by my illness in a totally different way than I. Not to say that he doesn’t have a positive vision for my future, because I get the impression from him that I can do anything with my life. But he worries I will always have to worry about my health. While I agree with my dad that I will always have a little bit of worry stashed away, I don’t see my life as lacking. I don’t even see my life as challenged.

I see my life as this great energy pushing me to do something outstanding, and my situation will just make it that much more memorable. It will by no means hold me back, because, although I am absolutely affected by my disease, I am not my disease.
When talking with patients and nurses familiar with plasmapheresis, they sometimes good-naturedly compare the procedure to an oil change. And, as everybody knows, oil changes are a necessary component to a smooth-running and long-lasting engine. With plasmapheresis, the materials are different but the philosophy is the same: Out with the old and in with the new is a positive. Or, in other words, change can do you good.

Since the 1970s, patients with autoimmune disorders and their healthcare providers have supplemented medication therapies with plasmapheresis, wherein the autoantibodies responsible for the disorder are removed from the bloodstream through a mechanical process.

Patients continue taking other prescribed medications, or sometimes intravenous immune globulin (IVIG), between plasmapheresis sessions. But, a main benefit of the procedure is it allows reduction in the amount of medications needed, thereby reducing the side effects of those medications patients often experience when taking high doses.

The procedure is usually performed on an outpatient basis, and most patients report discomfort but no notable pain. Patients have reported such side effects as faintness, dizziness and cramping, although these can often be treated by repositioning the patient, who is usually reclining in a chair. Patients must be monitored for allergic and other reactions, either to the solution used to replace the plasma or the sterilizing agents used for the tubing. With proper dosage, improvements are noticed at different times, depending on the patient, from a few days to several weeks.

Plasmapheresis procedures are performed at many hospitals and outpatient medical centers. As always, consultation with a qualified healthcare practitioner is necessary before beginning the procedure.

The University of Rochester Medical Clinic in New York performs some 1,300 plasmapheresis sessions each year.

"Patients with a variety of medical conditions visit us for these treatments," said Donna Cimbalo, nurse director of the apheresis unit at the medical center. "We see people who have had organ rejection, immunology problems, Guillain-Barré syndrome, multiple sclerosis and many other conditions. They might be referred by a specialist, a hematologist or an immunologist."

For most of these patients, the procedure is relatively simple.

"As long as there is adequate IV access, it can take less than two hours," Cimbalo said. "It's fairly well tolerated by most patients. The most common side effect is that people are usually tired afterward."

Cimbalo tells of patients who have enjoyed dramatic changes following the treatment.

"We had one gentleman come in who had open wounds on his feet and very painful fingertips," Cimbalo said. "We got him on a regular course, and now he plays golf and walks three miles a week. Better strength and quality of life are the positive results most often reported."

For some patients, that quality comes with a couple of visits a year, others may require multiple treatments each month.

Kathy Lynne Cook, 50, has Guillain-Barré syndrome and receives plasmapheresis treatments twice a year, along with regular IVIG therapy.

"I didn't know why I couldn't swallow or breathe right," Cook said. "The plasmapheresis treatments helped me regain function. It can sometimes get uncomfortable. I might feel nausea or headaches during the treatment, but the nurse slows down the procedure and then I'm OK. By the third day following the treatment, I really feel the results."

Cook says she now sees the plasmapheresis treatments as the perfect accompaniment to her IVIG infusions. She's a regular contributor to a chat room concerning the treatment.

"So many people who could be helped by this treatment have not heard about it," Cook said. "Within the community of people who are affected by Guillain-Barré, we try to get the word out."

For Cook, the treatment is the opportunity to regroup, and cites that now-familiar automotive reference.

"This treatment is my oil and lube job," Cook said, laughing. "It gets my motor running again."

In fact, in many cases, plasmapheresis is an acceptable alternative to IVIG therapy for autoimmune diseases.

Annie Tu, a nurse manager at the University of Washington Medical Center, has been performing plasmapheresis treatments for some 30 years.

"The procedure has changed dramatically, mostly in the sense that the measurements are much more precise now,"
Plasmapheresis

Tu said. “When we first began giving plasmapheresis, we could only make a best guess on how much product was needed. Now computers can do much of that work for us.”

Tu and other plasmapheresis nurses use various factors, such as the nature of the illness, the weight of the patient and previous treatments to input information into the plasmapheresis computing system, which in turn indicates the proper dosage and length of treatment.

“Every patient is different,” Tu said, “and arriving at the proper treatment is not the same for everyone. In general, you will find that patients who weigh more require longer sessions and a more ambitious course of treatment than patients who do not weigh as much.”

Tu says transplant patients can be blood-tested for the proper treatment, while with neuropathy patients, it’s often a case of having the treatment and then monitoring the results.

“We see what the reaction is, then move forward from there,” Tu said. “Sometimes it will take a couple of treatments to get to the right place, and the patient helps in that decision-making process by reporting their reaction. We ask the patients to help us come up with the best treatment approach based on how long it takes for them to feel better and what sort of reactions they are experiencing, either positive or adverse.”

Tu says patients can usually feel the treatment, but most handle it fine.

“For those who soon are feeling much better, the discomfort doesn’t seem to bother them as much,” she said.

As for the future of plasmapheresis, Tu and others say the procedure will continue to get fine-tuned as technology develops.

“There will probably come a time when we can arrive at the exact treatment a patient needs based on technology,” Tu said. “Computerized medical techniques such as this are becoming more and more capable of registering exact needs, and I expect that as those needs are met, even more patients will seek out and benefit from plasmapheresis.”

What Is Plasmapheresis?
From the Muscular Dystrophy Association and Myasthenia Gravis Foundation

Plasmapheresis is a process in which the fluid part of the blood, called plasma, is removed from blood cells by a device known as a cell separator. The separator works either by spinning the blood at high speed to separate the cells from the fluid or by passing the blood through a membrane with pores so small that only the fluid part of the blood can pass through. The cells are returned to the person undergoing treatment, while the plasma, which contains the antibodies, is discarded and replaced with other fluids. Medication to keep the blood from clotting (an anticoagulant) is given through a vein during the procedure.

What’s involved in a plasmapheresis treatment?

A plasmapheresis treatment takes several hours and can be done on an outpatient basis. It can be uncomfortable but is normally not painful. The number of treatments needed varies greatly depending on the particular disease and the person’s general condition. An average course of plasma exchanges is six to 10 treatments over two to 10 weeks. In some centers, treatments are performed once a week, while in others, more than one weekly treatment is done.

According to the Myasthenia Gravis Foundation of America Inc., plasmapheresis may be recommended for a few reasons:

• To stabilize a rapid decrease in muscle strength
• To reduce moderate to severe muscle weakness before surgery
• To add to present treatment if current forms of therapy are providing insufficient control of the disease

References
Muscular Dystrophy Association: “Facts About Plasmapheresis”
www.mdausa.org
Myasthenia Gravis Foundation of America Inc.: “Plasmapheresis”
www.myasthenia.org
Many readers have asked if intravenous immune globulin (IVIG) is covered in the homecare setting under Medicare Part B or Part D.

**Answer:** This question is extremely important, and the answer depends on your diagnosis. If you have a primary immune deficiency disease (PIDD), you are covered under Medicare Part B, and your coverage includes reimbursement for the drug only; it does not cover nursing services to administer the IVIG or durable medical equipment, such as a pump.

The reimbursement for IVIG in the homecare setting under Medicare Part B is at the same rate as in the physician’s office (although physicians can also bill separately for administration of the IVIG), so it will be difficult to find a homecare company that will provide IVIG for PIDD patients as long as the reimbursement rate continues to be lower than the cost.

If you are told that you are or would be covered under Medicare Part D for IVIG therapy for PIDD, this is not the case and you should not sign up for Medicare Part D for such coverage. However, Part D will cover your other prescriptions, so if you do not have a secondary insurance, you should consider Part D for your other prescriptions.

If you have a diagnosis other than PIDD, such as chronic inflammatory demyelinating polyneuropathy (CIDP), Guillain-Barré syndrome, idiopathic thrombocytopenic purpura (ITP), myositis, myasthenia gravis or other disorders normally covered by Medicare in the physician’s office or hospital outpatient setting, Medicare Part D should cover your IVIG in the homecare setting.

Although, like Part B, Part D does not cover nursing services or durable medical equipment, the reimbursement rate for IVIG is higher in Part D than in Part B, which increases the likelihood that homecare companies will be able to serve you.

Please remember that you will have to pay out-of-pocket expenses until you reach $3,850 (including the deductible, copay and the “doughnut hole”), and then you should be covered at 95 percent to 100 percent for the rest of the year, depending upon your policy.

Please review all of the Medicare Part D policies: Some of the policies may offer supplemental coverage during the “doughnut hole” period or may assist with the $3,850, if this is a financial burden.

A newly diagnosed CIDP Medicare patient asked how she can afford to receive her IVIG. Her physician does not provide IVIG in his office, her local hospital is not taking new patients, and the homecare company to which she was referred said her treatment would cost $11,000 per month.

**Answer:** If you have Medicare and have a diagnosis other than PIDD, you should consider signing up for a Medicare Part D policy. The patient in this case does have...
Medicare Part D, but the homecare company did not understand how the coverage is implemented: The patient must pay out-of-pocket expenses up to $3,850, including the deductible, copay and doughnut hole, at which point Medicare Part D will cover 95 percent to 100 percent, depending on the policy. Additionally, if you have a secondary insurance other than Medicare, your nursing services for administering the IVIG will also be covered.

This patient was able to afford to pay $3,850 out of pocket, and the homecare company had the IVIG brand that she needed, so, after a little education, the problem was solved, and the patient is currently receiving her IVIG in her home.

A patient with PIDD asked how to resolve her insurance dilemma. She was applying for disability and her case was scheduled to be seen by an administrative judge. She decided to stop the process of trying to obtain disability because of the reimbursement problems for IVIG under Medicare and because she has another healthcare policy. She thought that she would be forced to have Medicare become her primary insurance coverage in the physician’s office. She also thought that, if she turned down Medicare, she would not be able to receive her Social Security check when she turns 65.

Answer: When you apply for Medicare either under disability or when you turn 65, Medicare Part A is mandatory. Medicare Part A covers your hospital inpatient services. Currently, the IVIG reimbursement crisis has not affected Medicare Part A, because it is not necessary for most IVIG patients to receive their infusions in the inpatient setting.

Medicare Part B is where the majority of the reimbursement problems for the IVIG community are occurring. Medicare Part B is optional when you apply for Medicare. If you are covered by a health insurance plan other than Medicare, and want to keep your current insurance policy as your primary, discuss that option with your health insurance benefits manager and Medicare, and determine if you should obtain Medicare Part B benefits.

Medicare Part D is an optional prescription drug plan that should be considered by all patients, regardless of diagnosis, for their prescription drugs—just remember that Part D will not cover IVIG for patients with PIDD, so, again, such patients need Part B to cover their IVIG.

Linda of Rhode Island asked how she can get Medicare to pay for her treatment when the guidelines are neither clear nor realistic. She said she is required to have her blood tested every three months, and, based on the test results, Medicare is reducing the number of grams of IVIG she receives, even though she is having severe infections.

Answer: In Linda’s case, the local Medicare carriers for Rhode Island, Blue Cross and Blue Shield and Mutual of Omaha, have implemented a local coverage determination (this is a very important term for patients to learn) for PIDD patients who require serum trough level testing every three months. The serum trough levels must be no higher than 400 to 600 mg/dl in order for patients to continue to receive their infusions at their current dosage and frequency. If the levels are higher, the hospital can reduce the number of grams per infusion or increase the number of days between infusions until the trough level drops to 600 mg/dl or lower. For many PIDD patients, this level is too low and results in increased and/or more severe infections. Additionally, the local coverage determinations for dosing are inadequate. The initial IVIG dose is 200 to 400 mg/kg body weight and maintenance doses are 100 to 200 mg/kg body weight, administered approximately once per month. According to the American Academy of Allergy, Asthma and Immunology, the usual IVIG dose for antibody replacement is 300 to 600 mg/kg per month, delivered every two to four weeks through intravenous route; and an acceptable starting point for maintenance dosing is 400 mg/kg every three to four weeks. In fact, because Linda tested outside of Medicare’s target range and the new dosing requirement, they reduced her monthly IVIG dosage, which has resulted in her having continuous, severe infections.

Rhode Island is one of 47 states that have implemented local coverage determinations for PIDDs that can include at least this restriction, and some states are imposing additional restrictions that can also lead to inadequate IVIG treatment. Such local coverage determinations do not appear to be based on sound medical science, and they are affecting PIDD patients and patients who rely on IVIG for other indications.

Please contact IG Living if your treatment has been detrimentally impacted by a local coverage determination, so we can continue to compile information about the effects of such determinations on patients’ quality of care.

editor@igliving.com 800-843-7477 x1143

Editor’s note: Please check our next issue of IG Living (April-May 2007) for a follow-up story on Medicare local coverage determinations. We will include all of the current restrictions in place, by diagnosis and state.
Proper diagnosis is one of the first challenges for families dealing with primary immune deficiency diseases (PIDDs). Consequently, more and more organizations are addressing the need to increase awareness of the diseases. The latest effort is a collaborative one by American Academy of Pediatrics (AAP), the Jeffrey Modell Foundation (JMF), and Talecris Biotherapeutics.

Last October, the three organizations announced a new continuing medical education (CME) series, the goal of which is to increase awareness and speed diagnosis of primary immune deficiencies in children. The program, “Immunodeficiency in Pediatrics,” is supported by grants from JMF and Talecris, and will be mailed to the 35,000 subscribers of Pediatrics in Review. It can also be downloaded for free from www.prepaudio.org.

The CME program features a panel discussion about immune deficiencies in pediatric patients. The panel consists of Rebecca H. Buckley, MD, AAP, the J. Buren Sidbury Professor of Pediatrics, and Professor of Immunology at Duke University Medical Center; Melvin Berger, MD, Professor of Pediatrics and Pathology at Case Western Reserve University; and Thomas A. Fleisher, MD, Chief of the Department of Laboratory Medicine at the National Institutes of Health.

“This program provides some very useful information for pediatricians who may be encountering challenging patient cases that could be a primary immune deficiency,” said Dr. Buckley. “Given that the time for accurate diagnosis averages longer than nine years from the time of a first infection, this program can help provide an earlier alert for pediatricians to consider the possibility of a primary immune deficiency, and make a referral that will lead to faster diagnosis.”

The discussion also addresses the concern that primary immune deficiencies may be more common than previously thought, and that pediatricians need to have a high degree of suspicion when evaluating certain key signs and symptoms, such as recurrent or resistant infection in their patients. This increased vigilance can then result in steps leading to more rapid diagnosis.

Fred Modell and his wife, Vicki, co-founded the Jeffrey Modell Foundation after their son died as a result of his PIDD. Modell commented on the importance of collaborative efforts to improve disease awareness and diagnosis. “From our perspective, this is an outstanding example of an effective collaboration that benefits patients. Bringing together the passion and unique contributions of leaders from academia, industry and the patient community creates the best opportunity to advance care and improve outcomes. If this collaboration can speed diagnosis and get patients to effective treatment more quickly, then it has accomplished a very important goal for children and their families.”

Alberto Martinez, MD, President and CEO of Talecris Biotherapeutics, reinforced Modell’s comments. “As a leading provider of therapies that treat primary immune deficiencies, Talecris recognizes and acts on its corporate responsibility to support programs complementary to the therapeutic outcomes we strive to achieve through our product offerings. As a pediatrician, I am particularly pleased that this project has the potential to improve awareness and recognition of primary immune deficiencies in children, leading to earlier institution of therapy.”

For more information on the “Immunodeficiency in Pediatrics” program or the PREP® Audio series, contact the American Academy of Pediatrics at 866-843-2271 or visit www.prepaudio.org.
"And I think to myself, what a wonderful world."
— Bob Thiele and George David Weiss, songwriters

Over the past year, I’ve written about primary immune deficiency disease (PIDD), worked as an advocate for PIDD patients, and struggled with the disease myself. In that time, I’ve come in contact with the most incredible people, learned numerous life lessons, and witnessed successful struggles I thought were impossible to surmount.

On this journey, one of the most important lessons I’ve learned is that it is possible to be healed while not cured. I do not look in the mirror and see a sick person, but a whole person in body and soul. Living with a chronic illness does not make me less, it actually makes me more.

I’m able to spend more time reflecting on life, my eyes have opened to the wonders and beauty of the world I never had time to see before, my heart has grown to allow many more souls and smiles in, my arms have opened wide enough to hold all the loving and caring people I’ve met, and my mind has opened to the daily acts of bravery I see PIDD patients and caregivers perform. We handle our illnesses differently, but one thing is common to us all: We are as brave as any warrior there is.

I’ve had the good fortune to meet many such fighters and their families. I’ve seen how we hold each other together in difficult times, and I am humbled by their strength and courage. I’ve seen young patients fighting for a normal childhood, sometimes winning, sometimes not. I’ve met older patients who have learned so many lessons along the way. Their knowledge is invaluable, and they become our teachers.

The patients in the middle years of their lives might be holding down a job while raising a family and struggling with PIDD, and to them I tip my hat. There are also those who live in the state of denial, and I do remember what a glorious state that was, so enjoy it while you can.

I’ve learned the importance of putting myself first. It was a difficult lesson to learn, but if I don’t take care of me, there is no room for you. I spend many of my days covered in books and blankets knowing it will then lead to many days playing and impersonating a healthy person. An example of that would be the day I was on a bike ride when suddenly the gear locked, then the brake locked, and finally the back tire pushed forward and I went flying like Peter Pan. My right hip hit the street and my left leg flew over the bar and was cut and bruised. I walked my bike home in so much pain that every step was pure hell. I was sore everywhere. I then called everyone I knew to tell them that I was hurt and it felt so good! It was normal person pain, not sick person pain. I was so excited to feel normal.

While we are sick, most of us have been very fortunate to find caring healthcare professionals. They appear as a nurse, doctor, administrator, specialty pharmacy rep, drug manufacturer or support group. When we find someone who actually partners with us to find the best method of treatment, really cares about our outcome, and treats us like humans instead of a chart number, we know we are fortunate. They make our journey so much easier.

Thank you to all—patients, friends, family, healthcare professionals—for everything you’ve taught me this year and for everything you’ve been to me this year.

"And I think to myself, what a wonderful world."
Like so many people, Dominick Spatafora’s illness took him by surprise. “I was a 29-year-old, healthy guy,” Spatafora said. “I was a regular Generation X kind of a guy who thought of himself as a consumer, not a patient. I was often looking at life and asking, ‘What’s in it for me?’”

Spatafora’s perspective changed when he began experiencing a serious tremor in his right hand. By the time he decided to visit a doctor a year later, atrophy had set in. “I was misdiagnosed by two different neurologists,” he said. “One diagnosed me with Lou Gehrig’s disease, and gave me three to five years to live. The next diagnosed me as needing an ulnar nerve transplant.”

Finally, Spatafora was diagnosed correctly—with multifocal motor neuropathy, a condition caused by deterioration of the peripheral nerves, disrupting the body’s ability to communicate with its muscles, organs and tissues, according to the definition provided by The Neuropathy Association (www.neuropathy.org).

“I was fortunate in that my employer paid for a second opinion, and the diagnosis was reconfirmed,” Spatafora said. “I went on monthly IVIG treatments, and my condition improved dramatically.”

Then one day, Spatafora received a phone call that would change his life a second time. It was his physician, telling him there was a national shortage of IVIG, and that he would no longer receive the product. Going months without treatment, Spatafora lost the use of his right hand. “I was a 30-year-old guy who had completely lost the use of his right hand, and I needed that IVIG to live my life,” he said.

It was then that Spatafora’s career background in the public-policy industry paid off. Spatafora had long been involved in public healthcare efforts. While working for former House Speaker J. Dennis Hastert, Spatafora was part of sweeping healthcare reform efforts, and later became one of the leading healthcare policy advocates in Arizona, as an employee of the state’s medical association.

“Being involved in public-health policy, I knew where to go,” he said. “I appealed the decision by my health insurance company, and won that appeal.”

It was the letter that followed that prompted an already angry Spatafora to realize that patients with neuropathy needed an advocate. “The letter informed me that I had won my appeal, but then asked me not to tell anyone I was getting the IVIG, because other patients would not receive it,” he said. “That made me irate. I knew that other patients without my background and experience would not be able to navigate through this complex system.”

Spatafora began planning and developing the San Francisco-based Neuropathy Action Foundation. “It’s about awareness and empowerment,” Spatafora said. “We want to educate the general public on what neuropathy is. Most people don’t realize that it is one of the most debilitating kinds of pain, and that 20 million Americans suffer from this.”

The foundation also seeks to better inform physicians, as a good number of cases go undetected. “The third goal may be the most unique and powerful, and that is patient empowerment,” Spatafora said. “With the high cost of prescription drugs, insurance companies are often limiting access. We will have a patient hotline where people can seek solutions. We want to establish an appeal process or regulatory body that will help patients. At the top of the website, we have a favorite quote of mine from anthropologist Margaret Mead: ‘Never doubt that a group of small, committed citizens can change the world. Indeed, it is the only thing that ever has.’ Patients need to step up to the plate and share their voice.”

Meanwhile, Spatafora continues his own career as a consultant for DVS Governmental Consulting Solutions, a company he founded. “The most powerful coalition I have seen is that which teams patients and care providers,” Spatafora said. “No health plan or government entity can beat that. And so many patients with chronic illnesses have such passion and energy, but they don’t know where to go. This foundation will help them funnel that energy.”

For more information about the Neuropathy Action Foundation, visit www.neuropathyactionfoundation.org.
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Metchnikoff earned the Nobel Prize in physiology for his research on immunity. He developed a theory of phagocytosis, a process performed by specialized cells to remove foreign bodies and thus fight infection, and promoted consumption of lactic acid bacteria, a microbe in sour milk.

Today's products with live active bacteria cultures are based on Metchnikoff's theory. Called probiotics, they have become a popular commodity and are used to promote gastrointestinal health and immunity. However, nearly a century after Metchnikoff's recommendations, there are still many questions about them. The purpose of this article is to address these questions and enable readers to make informed choices about probiotics.

What Are Probiotics?
Probiotics are "friendly" microorganisms that, when consumed in certain amounts, may confer health benefits. They are listed by: (1) Genus, (2) Species and (3) Strain. For example, *Bifidobacterium infantis* 35624 is (1) Genus: *Bifidobacterium*, or B., (2) Species: *infantis* and (3) Strain: 35624.

On the other hand, pathogenic—unfriendly—microbes can cause disease within and beyond the gastrointestinal tract. Metchnikoff explained this concept with flair: “…Bacteria set up business in your interior departments; overwhelm friendly bacteria, called phagocytes, if you chance to be a little run-down; start gnawing around where they have no right to be; and the first thing you know, you are coming down with stomach trouble.”

In theory, by ingesting probiotics, we may be able to influence the integrity of the gut (a significant part of the immune system) and affect the course of health and disease. Nevertheless, there is still more to learn, and the exact mechanisms by which probiotics reduce disease risk are not entirely clear.

How Do Probiotics Work?
Researchers believe that probiotics use their weaponry (such as natural toxins, fatty acids, hydrogen peroxide, antimicrobial agents, etc.) to create a less hospitable home for pathogens. In certain diseases associated with intestinal permeability (leaky gut), probiotics may serve as a barricade by wedging themselves between other cells and fortifying the gut’s mucosal barrier. In high numbers, they may crowd out or displace pathogens, the unfriendly germs.

Our gut contains hundreds of species of microbes and the composition is fairly stable. However, certain infections, autoimmune states, surgical procedures or drugs (e.g., antacids, antibiotics, chemotherapy) can alter the gut flora. During these vulnerable periods, germs that reside in the hospital, home, countryside, etc. can take up residence in gut flora and affect health in positive or negative ways. When the constitution of the gut flora becomes irregular, an individual is more susceptible to diarrhea, inflammatory, and allergic diseases. The same thing is true for newborns. Because their gut is germ-free, any bacteria—good or
bad—can stumble on vacant terrain and build a colony. So, the aim of probiotic exposure (or colostrum and breast milk, in the case of infants) is to acquire helpful, or nonpathogenic, bacteria that restore the gut and support health.

What Conditions Might be Improved by Probiotics?
Probiotics have potential to minimize gastrointestinal conditions such as diarrhea, irritable bowel syndrome (IBS) and inflammatory bowel disease (IBD), and they may improve health beyond the intestinal milieu. Probiotics also work indirectly to help recovery by enhancing compliance to therapy regimens. In one study, for example, children who were treated for *Helicobacter pylori* (peptic ulcer bacteria) with an antibiotic cocktail and probiotic (*L. reuteri*) had less intestinal distress. As a result, they were more likely to finish their course of treatment than those who used antibiotics alone.

Diarrhea
Antibiotic- and radiation-associated diarrhea is all too familiar for those who live with a weak immune system. A few studies have named probiotics that might minimize this problem: yeast *Saccharomyces boulardii* (Sb), *L. rhamnosus* (strain GG 53103 [LGG]), probiotic mixture VSL#3® ⁴, among others.

In addition, there is solid evidence for the use of LGG in the prevention and treatment of rotavirus-associated diarrhea. LGG shortens the duration of rotaviral diarrhea in children, but it does not help all forms of diarrhea. For example, there are still unanswered questions about the specific types of probiotic bacteria that are most effective for treating the different forms of diarrhea such as *Clostridium difficile* diarrhea. Just as specific brands of antibiotics work for certain types of infections, specific strains of probiotics work for certain types of diarrheal maladies.

Infectious and Inflammatory Bowel Disease
*C. difficile*-associated disease (CDAD) is a recurrent *C. difficile*-induced infection that is now a problem in the hospital and home. Repeated infections contribute to significant diarrhea and injury to the gut (e.g., pseudomembrane colitis). The buildup of *C. difficile* toxin is extremely dangerous in those with slow gut motility and immune dysfunction. In CDAD, the yeast Sb inhibits *C. difficile* toxin A-associated enteritis and may protect against intestinal inflammation. In addition, there are reports that certain probiotic mixtures such as VSL#3® ⁴ may be useful for managing CDAD, bacterial overgrowth, post-operative pouchitis and IBD.

Irritable Bowel Syndrome
IBS has been associated with decreased nonpathogenic (“friendly”) colonies and increased *Clostridium* species. In clinical trials, certain probiotic strains (such as *Bifidobacterium infantis*, 35624), have shown promise in treating IBS when used with gut-directed antibiotics.

Future Areas
Other promising areas of strain-specific probiotic research include asthma, atopic dermatitis, bowel dysmotility, colon cancer and others. Yet, considerable work is still needed to confirm the potential health benefits and develop standard guidelines for patient groups. Likewise, more studies are needed before routine use of probiotics can be recommended for the public or special groups. Even if probiotics are proven to be effective in treating pathogens, such as *C. difficile*, they will not be curative when one has a defective antibody response. In such cases, probiotics should be used judiciously and not as a substitute for antimicrobial drugs or IgG replacement therapy.

What Should the Consumer Know?
Do not interchange different strains. You cannot substitute different strains of the same species and assume that you will get the effect as described in the literature. LGG, for example, may enhance IgA against rotavirus but you will not see this effect with other strains of *lactobacillus*.

Less powerful probiotics colonize the human digestive tract only temporarily. Once you stop taking the probiotic, the bacteria will generally leave the gut. The more powerful probiotics are more likely to take up residence in the gut, but they are not always safe. *Lactobacillus* strains, for example, are resistant to vancomycin (an antibiotic for certain bowel infections).

Do not rely on a health food salesperson for advice. When considering a probiotic, it is essential to review the literature and identify the strain and dose that was used in clinical trials (see “Quick Checklist”). Then, consult with your physician.

Do your homework. Strains are generally not listed on product labels. Contact the manufacturer to find out what compounds are in the product. If the manufacturer cannot answer questions on the “Quick Checklist,” avoid using the product. When the treating physician is not familiar with the probiotic, it becomes the patient’s responsibility to find out the risks and benefits.
Quick Probiotic Checklist

Answer the following questions and then talk with your physician:

1. What is the genus, species and strain of the probiotic?
2. What are the health claims and alleged benefits?
3. What type of study was done? Was it Phase II (randomized, double blind, placebo-controlled (DBPC))? Was it Phase III (DBPC and compared to a standard therapy)?
4. Who was represented in the study? Human subjects? Age? Gender? Animals?
5. What dose or serving was used in the studies or clinical trials?
6. What form was used? Capsule? Powder? Food?
7. What adverse reactions have been reported? Drug- or nutrient-interactions?
8. What are the risks of using this probiotic?
9. How should it be taken? With or without food? Before or after medication?
10. Does it contain any stabilizers or fillers? Is there potential for a food allergy?
11. What are the manufacturing practices? How is quality controlled?
12. What are proper storage conditions?

Are Probiotics Safe?

Reports of adverse effects in generally healthy persons or full-term infants have been limited, and evidence supports the safety of certain probiotics. For example, strains of *Lactobacillus* have been given to premature infants who lack a developed immune system. No significant adverse effects were reported in these studies.

On the other hand, there have been cases of *Lactobacillus* species associated with endocarditis, liver abscess and pneumonia. There are documented cases of *Sb fungemia* (fungus in the bloodstream) in high-risk patients who used...
Schulman: Obviously, patients should involve their doctor but I have found that patients are not getting answers from their healthcare providers. Elmer: Absolutely. They should also talk with their pharmacists. We train our pharmacists about probiotics and there is some information online in databases. Natural Medicines Comprehensive Database [http://naturaldatabase.com] is a good one.

Schulman: There have been some questions about how other countries maintain different standards of quality and purity than we do. How do you extrapolate from one study to another?

Elmer: Many of the probiotics available in other countries, especially in European and Asian countries, are not available here. It is quite a dilemma. Studies may use certain strains and the results look encouraging. Then, you find that the strain is simply not available in this country.

Schulman: For the consumer, do you have any recommendations for finding probiotics that are safe and effective or is it buyer beware at this point?

Elmer: It is not quite that bad. I advise consumers to find probiotics that have been shown to be effective in sound clinical trials for the condition they have, if that is possible. The best-studied probiotics right now are LGG, Sb [S. bouardii], and VSL#3, which is particularly good for IBD. There is another good, well-studied probiotic, L. Reuteri.

Schulman: A further complication for some patients with immune concerns is they have related GI problems, surgical procedures and dependence on medications that alter the flora. It seems that they might benefit from probiotics. However, patients get mixed messages from clinicians—some recommend probiotics and others warn against it. So, for this population, what would you suggest?

Elmer: It's absolutely a dilemma. A few cases of bacteremia or fungemia [blood infections] with probiotics have been [reported] in populations that are immune suppressed or debilitated. The risk of problems with probiotics is very small, but there is a finite risk. The consumer has to weigh the benefits and risks.

Summary

Although probiotics hold promise for improving health outcomes, there is more work to be done. Consult your physician and give careful consideration to probiotic supplements before using them. Some things to consider are: effectiveness, dosage, safety record, manufacturer practices and strain. Finding the right bacteria for the right patient, at the right dose and the right time is not as easy as drinking sour milk.

Editor's note: Although there may be a useful role for probiotics in PIDD, no quality studies have been performed. Because some PIDD patients may be at risk from even the bacteria in probiotics, you should check with your physician regarding the utility of probiotics in your healthcare regimen.

References


Boyle, R.J., Robins-Browne, R.M., Tang, M.L.K. Probiotic use in clinical practice: what are the risks? Am J Clin Nutr, 2006; 83(6); 1256-64.


On the surface, the Scott family seems like a typical American family: three kids, a dog, a nice house in a suburban neighborhood. And, it is typically hectic in their home. In the mornings, making sure everyone is dressed, has his or her lunch packed, and gets out the door on time is reminiscent of any family with kids in school. In the afternoon, juggling Andrew’s soccer practice, Millie’s piano lessons and Claire’s ballet sometimes leaves the Scotts feeling overwhelmed. In addition, Mr. Scott runs his own business and Mrs. Scott works part time at the children’s school. All pretty typical.

Add to all of this activity planned trips to the hospital every three weeks, constant hand-washing to avoid bacterial infections and frequent doctor visits for anything that resembles a potential illness. Still sound pretty typical?

Yes, if, like the Scotts, you have a child with an immune deficiency disorder. And, to complicate the scenario, a new research study on the psychosocial functioning of children with immune deficiency diseases indicates that these children may have elevated behavioral and emotional difficulties.

This research study was initiated when Dr. Carrie Piazza-Waggoner was working with children with asthma at Ruby Memorial Hospital in West Virginia. A physician on staff noticed that children with immune deficiency disorders often displayed an array of psychological difficulties and suggested Piazza-Waggoner investigate their increased risk of behavioral and emotional problems.

The recognition that children with chronic illness are at an increased risk for both behavioral and emotional difficulties is not new. But, children with immune deficiencies and their families had yet to be the focus of research. Piazza-Waggoner’s research attempted to determine if children with immune deficiency diseases and their caregivers have altered psychological function and if the severity of the disorder is related to psychological functioning.

Piazza-Waggoner and her colleagues collected data from 40 children and their caregivers. Twenty of the children had a diagnosis of pediatric immune deficiency disorder, and 20 had asthma, serving as a comparison group. The children and their caregivers completed a variety of psychological questionnaire forms.

The findings from Piazza-Waggoner’s study seem to confirm many perceptions parents of children with pediatric immune deficiency disorders express: The caregivers of immune deficient children reported more behavioral problems in their children than did caregivers of children with asthma. The caregivers also reported their own elevated psychological distress.

Furthermore, the severity of the immune deficiency disorder was significantly associated with several behavioral adjustment issues. More specifically, results from this study show that children who received intravenous immune globulin or immunomodulatory treatments were reported to have more problems than children not receiving them, i.e., children with very severe immune deficiencies were more likely to have a psychiatric diagnosis and receive special education services.

Piazza-Waggoner had hypothesized that illness severity may impact the degree of psychological functioning. She clarified the study’s results, “It's not necessarily the [disorder], but perhaps it's all the complications of the medical issues of having multiple diagnoses—there's a lot to manage.”

“There is absolutely a need for more research,” she continued. Additional research on the psychological functioning of pediatric patients with immune deficiencies could help identify interventions that promote the children’s overall adjustment.

In the meantime, what can families like the Scotts, their teachers and doctors glean from the study’s findings? The research should heighten awareness that psychological distress may be elevated in individuals with immune deficiency disorders. If doctors are not aware of this, caregivers can raise the issue and request referrals. If families notice that their children display increased sadness, fear, worry or withdrawal; high levels of social distress; interpersonal difficulties; low self-esteem or other distress, they should consider seeking such assistance.

Another important point to remember: Parents and other caregivers should recognize their own distress as well. The heavy burden of disease can take its toll on the entire family, even in families as typical as the Scotts.

References
There may be as many as 500,000 people in the United States with diagnosed and undiagnosed primary immune deficiency diseases (PIDDs). For many, the diagnosis of PIDD occurs after a medical work-up prompted by problems with chronic and unresolved sinusitis. Even with regular immune globulin therapy, many patients are just dreading the next sinus infection.

Sinusitis Basics

Sinusitis involves inflammation and infection of the sinus cavities, and it usually follows a cold or an allergy exacerbation. Some infections occur because mucus can’t drain through the nasal passages. Chronic sinusitis is defined as sinusitis lasting for at least three weeks.

Sinus cavities are air spaces located in the skull surrounding the nose. There are four pairs of sinus cavities known as the paranasal sinuses, and one or more of them are the usual culprit when a person complains of sinus pain:

- Frontal (above the eyes)
- Maxillary (inside the cheekbones)
- Ethmoid (behind the nose, between the eyes)
- Sphenoid (in the upper part of the nose, behind the eyes)

Sinuses perform their job by providing an opening into the nose, allowing air and mucus to pass. The sinuses are connected by a mucus membrane lining the pathway. Understanding this connection is important, as it explains why any event creating swelling or inflammation in the nose can affect the sinuses. The inflammation can be caused by allergens, infection or environmental irritants.

If the sinuses get blocked with drainage, it may create pressure on the walls of the sinus cavities, in turn creating “sinus attacks.” If there is a blockage caused by inflammation, in turn preventing air from entering the paranasal sinus cavities, a vacuum may result, creating sinus pain.

You Already Know the Symptoms

Any individual who suffers from PIDD, or the parent of a child with PIDD, is likely quite aware of sinusitis symptoms. Common symptoms include facial pain, sinus tenderness, headaches, nasal drainage that is green or yellow, itchy cough, post-nasal drip, bad breath, upper jaw pain, sore throat, sensitivity to light, fatigue and swollen face and eyelids. Adults who are afflicted with sinus infection in the maxillary sinuses specifically may complain of pain in the upper jaw and sore teeth. Children who suffer from sinusitis may be less likely to complain of headaches than adults. The children will, however, be more likely to suffer from foul breath, irritability, coughing, fever and facial and eyelid swelling.

Many patients who suffer with chronic sinusitis may have an ongoing inflammatory condition due to allergies. Sinusitis may result from mucus blockage caused by allergic rhinitis. A good place to start resolving allergic symptoms is through avoidance of allergens (substances that cause allergic symptoms). Minimize exposure to pollens, mold, pet dander, dust mites and even certain foods to reduce allergies. Of course, it is difficult to avoid all allergic triggers so your healthcare provider may suggest different types of allergy medications to relieve the symptoms. Some types of medications your healthcare team may recommend include corticosteroids, antihistamines, decongestants, leukotriene modifiers and mast cell stabilizers. Never begin any new course of therapy without consulting your healthcare team.

Many patients find that sinus irrigation relieves the inflammation of sinus passages. Sinus irrigation involves the infusion of normal saline solution into the nasal passages. Patients can do it daily to reduce medication use and improve breathing. Discuss nasal irrigation with your healthcare provider prior to attempting it.

1 Cooper, Megan PhD, Pommering, Thomas, DO, Koranyi, Katalin, MD. “Primary Immunodeficiencies.” American Family Physician. Nov 2003.
What’s Bugging Me?
For most episodes of sinusitis, the origin is a bacterial infection that may have been preceded by a cold or a viral infection. The most common organisms causing the infections are streptococcus pneumonia and haemophilus influenza. These organisms are normally present in the respiratory system, and usually don’t create a medical complication unless a person’s immune system is compromised.

People with an immune deficiency are also at increased risk for sinusitis from other organisms, such as a fungus or other bacteria. The most common invasive fungal sinusitis is caused by aspergillus, and, if left untreated, can lead to destruction of the sinus cavities and bone and encroachment of the infection into the eye sockets and brain. For these reasons, it is imperative to receive treatment for sinusitis by a trained practitioner, well informed about the care of patients with PIDD.

What Type of Sinusitis Do I Have?
The healthcare provider will determine if the diagnosis of chronic sinusitis applies, through in-depth medical history and a physical. This will involve questions about specific symptoms and their duration, along with an MRI or CT scan and comparison of X-rays.

The results of X-rays are not considered the best diagnostic tool for chronic sinusitis, so a CT scan will usually be requested. After review of all of this information, the healthcare provider may begin treatment with antibiotics or other medications, or may request that the patient consult with an ear, nose and throat (ENT) specialist. A traditional nasal swab is not considered to be very helpful in determining the cause of a chronic sinusitis. The only way to get a more detailed specimen from the sinus cavity is through an invasive culture collected while under anesthesia.

What If the Meds Don’t Work?
Sinus surgery sometimes helps reduce the frequency of infections and symptoms for people with primary immune deficiencies. If the healthcare provider believes sinus surgery is necessary, the patient will be referred to an ENT specialist. The specialist will determine if sinus surgery is the best option after reviewing the imaging studies, such as a CT scan, the patient’s history and physical exam, and other possible test results, such as blood work.

Sinus surgery is conducted as an endoscopic procedure, meaning through the nose without cutting the patient’s skin to reach the sinus cavities. If it is necessary, polyps can be removed during the procedure and the septum can be straightened. The procedure lasts from one to three hours, and is conducted under general anesthesia. Sinus surgery is usually performed as an outpatient procedure, unless there are other health concerns.

There is usually no scarring, as the scope is passed through the nose. The ENT surgeon opens the natural nasal passages, allowing them to drain and ventilate. During this time, it is possible to obtain a true culture of the organism causing the sinusitis. The surgeon may use the visual scope to remove polyps or diseased tissue in the area. If the passages appear to be open and able to ventilate well, the patient may have the area “washed,” which means irrigated in order to cleanse the cavity of the organisms causing the inflammation and infection.

A follow-up visit to the ENT specialist usually is scheduled one to two weeks after the sinus surgery,
depending on the individual patient outcome. After the procedure, there may be mild facial swelling, discomfort in the nose, difficulty breathing through the nose, and some difficulty swallowing. These symptoms usually resolve in two or three weeks. If symptoms do not resolve or get worse, the ENT specialist and the primary healthcare provider should be contacted.

Follow-up care after sinus surgery involves dedicated nasal sinus irrigations to keep the tissues clean and moist. This step often determines the length of positive results from the procedure, so it needs to be taken seriously. The irrigations may be required from seven days to many weeks, depending on the individual.

Although sinus surgery may be effective in treating sinusitis, therapy and treatment for the underlying cause must be continued after surgery.

**Kids and Sinus Surgery**

A child’s frontal sinus does not fully develop until somewhere between ages 8 and 12. During a sinus surgery, the area known as the anterior ethmoid will be opened and ventilated, which in turn opens the area called the frontal sinus recess. The ENT specialist will avoid the frontal sinus recess in order to prevent damage or obstruction of this area.

A child should be prepared for some nasal pain following a sinus surgery. Inform the child that it may be hard to breathe through the nose. There may be a small gauze dressing under the nose to catch the brownish drainage after surgery. Nasal irrigations can be challenging for children who do not want anything placed into their noses. Consult the child’s healthcare provider for alternate ways to keep the nasal passage moist, such as sodium chloride sprays or vaporizers. Try using a reward system for successful irrigations or nasal sprays, such as sticker charts, prizes or special treats.

Let the child know that after a sinus surgery, he or she may need to wait a few days to resume all normal play activities and school attendance. Consult the child’s ENT specialist for more details on follow-up care.

**Prevention**

People with PIDD have no real way to prevent sinusitis, although there are ways to maintain a healthier lifestyle and keep a vigilant watch for sinus complications. If the patient is on immune globulin therapy, it is important to follow through on all treatments and doctor appointments. It might help to use an air filter at home to reduce allergens and air pollutants, which cause irritation.

Adults with PIDD can help prevent sinusitis in several ways:

- Limit alcohol consumption, as it can inflame nasal passages.
- Try using a humidifier at home and at work, and be sure to maintain a clean filter in the unit as directed, and change water daily to avoid the creation of mold.
- Wear a mask around construction areas to limit exposure to fungus and molds.
- Discuss the use of decongestants with your healthcare provider—air travel can cause pain because of the changes in cabin pressure, and the use of decongestants may make the flight more comfortable.

Home remedies will not treat sinusitis, but some things can be done to minimize pain and discomfort. Some patients find the use of saline sprays helpful in keeping nasal passages from becoming dry and irritated. The use of warm steam from facial vaporizers or a bowl of hot water can also help. Warm compresses applied over inflamed sinus areas have been reported to reduce pain and discomfort.

Many patients find acupuncture helpful in reducing inflammation as well as decreasing pain and headaches associated with chronic sinusitis. Always consult your healthcare team before beginning any new treatment modality.

Unfortunately, chronic sinusitis is often a constant and major healthcare concern for people with PIDD. By being aware of the prevention methods, symptoms, and how to aggressively treat sinusitis, you might be able to forestall if not avoid that next sinus infection.

**References**


Department of Otolaryngology; Lucile Packard Children’s Hospital, Stanford. 2006. Endoscopic Sinus Surgery.


...Guillain-Barré Syndrome (GBS)

Websites and Chat Rooms
1. The GBS/CIDP Foundation International, 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/.

Online Pamphlets
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
2. GBS Support group—UK Chat room—requires registration: http://www.jsmarcusen.com/gbs/uk/chat.htm
3. GBS Foundation Discussion Forums www.guillain-barre.com/forums

...ITP (Idiopathic Thrombocytopenic Purpura)

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

Online References
4. Infusion Network Systems Article: The Expanding Use of IVIG provided by ZLB Bioplasma, Inc.: http://www.infusionsystems.net/article-ExpandingUseofIVIG.html

...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
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/.

Online Peer Support Groups
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/.

Online Peer Support Groups

...Myositis

Websites
1. The mission of The Myositis Association, www.myositis.org, is to find a cure for inflammatory and other related myopathies, while serving those affected by these diseases. 202-887-0088
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
Wanted to Know About...

Online Peer Support Links

...Peripheral Neuropathy (PN)

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

Online Peer Support Links
2. MSN Support Group: Discussion Board: http://groups.msn.com/PNPARTNERS
5. Yahoo Support Group—Australia Discussion Board: http://au.groups.yahoo.com/group/LifeWithPN/

...Primary Immune Deficiency Disease (PIDD)

Websites and Chat Rooms
1. The Immune Deficiency Foundation (IDF), www.primaryimmune.org, is dedicated to improving the diagnosis and treatment of PIDD through research and education. 800-296-4433
2. The Jeffrey Modell Foundation, www.info4pi.org, is dedicated to early and precise diagnosis, meaningful treatments and, ultimately, cures for primary immunodeficiency. 212-819-0200
3. The National Institute of Child Health and Human Development (NICHD), www.nichd.nih.gov, is part of the National Institutes of Health. Go to the “Health Information and Media” tab on the website and do a search under “primary immunodeficiency.”

Online Peer Support Links
1. Chat with parents of children affected by PIDD http://health.groups.yahoo.com/group/PedPID/
2. Chat with peers with PIDD http://health.groups.yahoo.com/group/PIDsupport/
4. Jeffrey Modell Foundation Message Board www.info4pi.org

...General Resources

Product Information
1. To learn more about Vivaglobin—the subcutaneous immune globulin (SCIG) go to: www.vivaglobin.com.
2. For more information about the 10% IVIG solution Gammagard Liquid, go to www.gammagardliquid.com.
4. For information about influenza and the influenza vaccine, visit www.cdc.gov/flu or call 800-CDC-INFO (800-232-4636).

Online Support Links
1. To learn more about Vivaglobin—the subcutaneous immune globulin (SCIG) go to: www.vivaglobin.com.
2. For more information about the 10% IVIG solution Gammagard Liquid, go to www.gammagardliquid.com.
4. For information about influenza and the influenza vaccine, visit www.cdc.gov/flu or call 800-CDC-INFO (800-232-4636).
Resource Directory

Other Organizations
1. 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.

2. 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

3. The nonprofit Patient Advocate Foundation, www.patientadvocate.org, seeks to assure patient access to care, maintenance of employment and financial stability. 800-532-5274

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

5. WebMD, www.webmd.com, is a handy medical reference that helps consumers take an active role in managing their health by providing objective healthcare and lifestyle information.

6. For a pediatrician’s guide to your child’s health and safety, visit www.keepkidshealthy.com.

7. The National Organization for Rare Diseases, at www.rarediseases.org, provides links to numerous other organizations that have disease-specific support groups and virtual communities for patients and caregivers.

8. American Autoimmune Related Diseases Association (AARDA) www.aarda.org brings national focus to autoimmunity through research, support groups and virtual communities for patients and caregivers.


Education and Disability Resources


3. California State Disability Insurance (SDI): www.edd.ca.gov (Please note that each state has a different disability program.)


5. The National Disabilities Rights Network: www.ndrn.org. This website offers a search tool to find resources in your state to assist with school rights and advocacy.


9. 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.

Books and Articles


3. "Anatomy of an Illness," by Norman Cousins, is a bestseller about overcoming illness and the triumph of the human spirit. The premise is that the human mind is capable of promoting the body’s capacity for combating illness and healing itself even when faced with a seemingly hopeless medical predicament.


5. "Bed Number Ten," by Sue Baier, provides a view of long-term care through the eyes of a patient totally paralyzed with GBS.


8. "Coping With a Myositis Disease," by James R. Kilpatrick, is written by myositis patients telling their personal stories.

9. "The Everyday Guide to Special Education Law," by Randy Chapman, Esq., makes the law accessible to parents so they can more effectively advocate for their children. Available at http://www.thelegalcenter.org/thelegalcenter-cgi-bin/shop?item=15

10. "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.

11. "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.

12. "Living Creatively With Chronic Illness: Developing Skills for Transcending the Loss, Pain and Frustration," by Eugenia G. Wheeler, is a self-help book specifically designed to help the chronically ill, their families, friends, counselors, medical personnel and the clergy.


14. "Managing Pain Before It Manages You," by Dr. Margaret A. Caudill, is a wellspring of wisdom and practical approaches that can help transform your life and your pain.
15. "Medifocus Guide to Peripheral Neuropathy," is a guide to current and relevant PN research, organized into categories for easy reading.


17. "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.

18. "Not Dead Yet: a Long Strange Trip From Doctor to Patient and Back Again," by Dr. Robert Buckman, an oncologist and comic writer, is a witty account of his life as a doctor and autoimmune disease survivor.

19. "Numb Toes and Aching Soles," by John Senneff, discusses the symptoms, causes, tests, treatments and coping strategies for peripheral neuropathy.

20. "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.

21. "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.

22. "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.

23. "Pride and the Daily Marathon," by Jonathan Cole, describes how Ian Waterman was suddenly struck down at work by a rare neurological illness that deprived him of all sensation below the neck, and how he reclaimed a life of full mobility.

24. "Pronoia Is the Antidote for Paranoia," by Rob Brezsny, explores the best way to attract the blessings that the world is conspiring to give us.

25. "When You’re Ill or Incapacitated" comprises one-half the booklet it shares with "When You’re the Caregiver," both written by James E. Miller, suggesting 12 things to remember or do in each role.

26. "YOU the Smart Patient: An Insider’s Handbook for Getting the Best Treatment," by Michael F. Roizen, MD, and Mehmet C. Oz, MD, with the Joint Commission on Accreditation of Healthcare Organizations, shows you how to tackle such healthcare decisions as picking the best doctors and hospitals for you, knowing when to get a second opinion, and more.

IG Manufacturer Websites
- Baxter: www.baxter.com
- Grifols: www.grifolsusa.com
- Octapharma: www.octapharma.com
- Talecris: www.talecris.com
- ZLB Behring: www.zlbbehring.com

Pump and Needle Websites
- Intra Pump Infusion Systems: www.intrapump.com
- Graseby Marcal Medical: www.marcalmedical.com
- Norfolk Medical: www.norfolkmedical.com

Medical Research Studies
Check out the official website for the National Institutes of Health patient recruitment program. This site provides summaries and criteria for studies as well as the ability to search for studies being conducted for a specific disease or disorder. http://clinicalstudies.info.nih.gov/

This website provides a wealth of information about clinical trials and volunteer participation. It gives you the ability to specify the disorder you are interested in, the location of the study, and the medication names or research protocols. www.centerwatch.com

This site has a registration form to request that you be notified about recruitment for future studies. www.clinicaltrials.com

WebMD has a service that matches volunteers with trials. There is an online questionnaire to complete and you will be notified via email of upcoming studies that match the criteria of your questionnaire. You can also search for specific studies. www.webmd.com

…Nutrition and Food Safety Information
2. American Dietetic Association: http://www.eatright.org
6. Childhood choking prevention:
   http://www.ffc.org/publications/brochures/index.cfm
7. American Academy of Allergy, Asthma & Immunology (AAAAI): www.aaaaai.org
8. The Food Allergy & Anaphylaxis Network: www.foodallergy.org
10. USDA Meat and Poultry Hotline: www.IsItDoneYet.gov

…Resources Just for Kids
1. “Germs Make Me Sick,” by Melvin Berger, explains with colorful illustrations how your body fights germs.
2. “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.

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

In 2006, FFF delivered 98% of MyFluVaccine orders on or before customers' selected delivery dates.

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