

IDF ADVOCATE

THE NATIONAL NEWSLETTER OF THE IMMUNE DEFICIENCY FOUNDATION



Immune Deficiency Foundation Mission

To improve the diagnosis and treatment of patients with primary immune deficiency diseases through research, education and advocacy.

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Soaring at the IDF National Conference

Spirits in St. Louis flew high June 28 as the city opened its gateway for three days for more than 1000 attendants at the IDF 2007 National Conference. This was the first time IDF had ventured west of the Mississippi and participants from 44 states, Washington DC, Mexico, Canada and even as far as New Zealand flocked to the “Show Me” state.

Marcia Boyle, President & Founder of IDF, warmly welcomed all patients, families, healthcare providers, and industry representatives at this gathering of the primary immunodeficiency community. She discussed the purpose of the Conference, “As patients and family members, the more you understand your diagnosis, the treatments, and issues such as health insurance, the better you can

protect you and your children’s future. We’re here to provide you with the tools to empower you individually, as well as to focus on issues that affect the community.”

Starting on a high note, Marcia introduced the keynote speaker, Alan Hobson. Alan, a Mt. Everest climber, summiteer and cancer survivor, presented a riveting talk on “The Triumph of Tenacity.” Weaving his compelling story of triumph and perseverance into the everyday struggles we all face, he demonstrated that with the right attitude we can achieve the goals we set and live the life we want. He offered practical steps as well as encouraging advice to manage the unexpected and turn obstacles into opportunities.

The audience responded with a standing ovation to this inspiring and motivational presentation, providing the perfect kick-off to this remarkable conference.

continued on pg.8

Handbook Chapters

The Immune System and Primary Immunodeficiency Diseases
Common Variable Immune Deficiency
X-Linked Agammaglobulin Selective IgA Deficiency
Severe Combined Immune Deficiency
Chronic Granulomatous Disease
Wiskott-Aldrich Syndrome
Hyper IgM Syndromes
DiGeorge Syndrome
IgG Subclass Deficiency and Specific Antibody Deficiency
Ataxia Telangiectasia
Hyper IgE Syndrome
Complement Deficiencies
Other Important Primary Immunodeficiency Diseases
Inheritance
Laboratory Tests
General Care
Specific Medical Therapy
Infants and Children with Primary Immunodeficiency Diseases
Adolescents with Primary Immunodeficiency Diseases
Adults with Primary Immunodeficiency Diseases
Health Insurance
Glossary
Resources

PATIENT & FAMILY HANDBOOK FOR PRIMARY IMMUNODEFICIENCY DISEASES

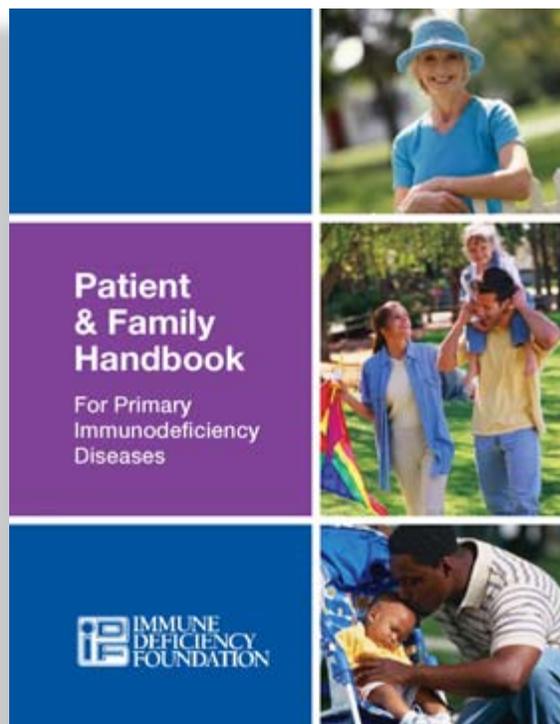
Fourth Edition Now Available

IDF is proud to announce that the Fourth edition of the Patient & Family Handbook for Primary Immunodeficiency Diseases is now available. It's been over two decades since the first edition, the result of an appeal from IDF patients, their families, and their physicians. Since that first edition, there have been two editions, translations into Spanish and French, and tens of thousands of copies distributed throughout the world. The handbook's goal has always remained the same—to provide the most current information on primary immunodeficiency diseases for our community.

This fourth edition was inspired by the many new and exciting advances in the diagnosis and therapy of the primary immunodeficiencies. Six new chapters have been added to this full color edition, and all of the existing chapters have been revised to include information on new diagnostic tools, more precise clinical information and new therapies. Chapters have been expanded and updated to reflect new findings and convey common sense guidelines. The resource section includes information available in both print and on the Internet and the glossary offers definitions of possibly confusing medical terms.

The Patient & Family Handbook is the result of the efforts of a large team of people who generously offered their time and knowledge. Both Drs. Michael Blaese and Jerry Winkelstein did a remarkable job as the editors for the handbook. Dr. Winkelstein also served as the editor of the first three editions and his experience and dedication were extremely valuable. More than twenty contributors wrote specific chapters that were reviewed by members of the IDF Medical Advisory Committee. Special recognition goes to Baxter Healthcare Corporation for their generous support of all three previous editions and the generous grant, which made this fourth edition possible.

To receive a free copy, simply contact IDF at 800.296.4433 or e-mail your request to idf@primaryimmune.org.



IDF Advocate

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Live Your Dreams!

By Nancy Hoffman, IDF Volunteer

When I was a young girl I used to bring home books about Africa. I would stare at pictures of the incredible animals and at the beautiful brown-eyed children and dream about one day actually being in Africa and seeing all this and more.

Unfortunately, things don't always go as planned and I was diagnosed with CVID in 1995 after nine years of serious illness. My doctor at that time said that I would "live a bubble existence." That meant no outside social activities and no travel. I thought that my dream might be lost. However, I am a very determined lady, and don't give up easily. I started checking out my options to figure out how I could travel and live life as I wanted. I contacted IDF and they were so helpful! Their practical advice and assistance encouraged me to follow my dream.

I had to make several changes in my life. This meant finding a new doctor who could help me pursue my interests, so I found a new doctor who encouraged me. My new physician suggested I try a new treatment method, subcutaneous immune globulin replacement therapy, better known as Sub Q. Sub Q is usually self-administered and allows for more flexibility. It gave me more control over my treatment schedule and made my travel plans possible.

My new doctor gave me advice about traveling with a primary immunodeficiency disease. He suggested I take Malaria Pills, an extra supply of antibiotic (just in case), drink bottled water and eat cooked food. Then he encouraged me to "go for it!"

That was all I needed to get going! I contacted the company that supplies my gamma globulin and supplies for the Sub Q treatments and asked them to help me prepare for my trip. They labeled all my bottles and supplies with my name, doctor's name and contents just in case there were questions about what I was carrying. I put all of this in a small black bag along with a letter from my doctor stating my diagnosis. I just put this bag along with my personal things through the x-ray machines at each airport, from Palm Springs to Dallas to London to Nairobi and back. Never once was I questioned...but I sure was ready!

My month long trip in January to Africa was an amazing journey! I experienced all that I'd hoped and dreamed of! I worked with the Kenyan Children's Foundation in an aids orphanage in Karen, Nairobi, visited homes and schools for children in the slums of Kibera and had the opportunity to personally hear and see these beautiful children with the big brown eyes. Then, while on



two incredible safaris in Samburu and Masai Mara, I had the opportunity to be only 10-15 feet away from amazing animals--the ones I had studied in the picture books as a child. This time, I was the photographer, taking pictures to my hearts content. I have breathtaking photos!

My Sub Q treatments were easily done in the clean little room I had at the convent in Karen, and then in the very nice tents (with a bathroom) on the safaris. I only needed to do my usual sanitary set up and all went well.

Since my adventure in Africa, I've also just returned from a month long trip with my husband and our cat "Boo" in our motor home. We saw eleven states, five national parks, and biked along the river in Durango, Colorado. Doing my Sub Q each week was so easy, even though every time it was in a new location.

I lived my dream and I have more planned! Yes, those of us with a primary immunodeficiency disease have a health situation to deal with, but it does not have to dictate how we live. We can work around it, and with it. It is important to "be aware and be prepared, but not afraid."

Nancy Hoffman, an IDF Volunteer from Big Bear Lake, California, was diagnosed with CVID in 1995. She is active as a peer support volunteer and grassroots advocate and was named Peer Support Volunteer of the Year in 2007. Her travels have taken her to Africa; however, her adventures with IDF have taken her to Capitol Hill to visit Congress as well as plasma centers to personally thank donors for their generosity and for making a difference in patients' lives.

IDF Applauds Legislation to Improve Access to IVIG

The Immune Deficiency Foundation (IDF) applauds Congressman Kevin Brady from Texas who introduced H.R. 2914, known as the Medicare IVIG Access Act of 2007 on July 28, 2007. With bipartisan support, the bill seeks adequate reimbursement for Intravenous Immunoglobulin (IVIG) products and accompanying care in the management of patients with primary immunodeficiency diseases (PIDD) and other serious conditions that require IVIG therapy.

“Through my work with IDF and the patients and their families living with PIDD, this issue has become very close to me personally,” adds Rep. Brady. “This bill, along with the strong endorsements of my colleagues on the Hill, shows that we have heard their voices in Washington, and they are not alone.”

The good news is that the problem of IVIG patient access came up during the House Ways and Means Committee’s consideration of the Children’s Health and Medicare Protection Act of 2007 (CHAMP) on August 1, 2007. Rep. Brady, as original sponsor of the Medicare IVIG Access Act of 2007, H.R. 2914, offered the bill as an amendment to be included in the CHAMP Act. Rep. Pete Stark, Chairman of the Health Subcommittee, responded that he believed that Mr. Brady had correctly identified the problems with IVIG. He mentioned his personal experience using IVIG and added that he would be pleased to work with Mr. Brady toward a solution.

Rep. Jim McCrery, ranking Member on the Committee, spoke in support of the amendment and specifically reiterated that IVIG is a real problem and emphasized its importance. He stated that everyone keeps saying that there’s a problem, but nothing gets done to fix it. He urged that they continue to work on it together. Rep. Charles Rangel, Chairman of the full committee, assured him that the commitment to work on the issue is there and that he recognizes the urgency to get something done for the patients.

The current IVIG access and care issue began in January 2005 as a result of the Medicare Modernization Act under Part B, which changed the way physicians and hospital outpatient departments were paid under Medicare. The law reduced IVIG reimbursement rates such that most physicians in outpatient settings could no longer afford to treat Medicare patients requiring IVIG, and either delaying regular treatment or sending many patients to a hospital clinical setting for therapy. Patient access was further complicated in 2006 when similar payment changes went into effect for hospital outpatient departments.

“For most patients living with primary immunodeficiency diseases, IVIG is their only treatment option,” said Marcia Boyle, IDF President and Founder. “The delay or denial of treatment which is occurring under current Medicare reimbursement standards is putting patients at tremendous risk for serious, life threatening complications.”

Specifically, the bill seeks to improve health outcomes and quality of life for patients by:

- Improving reimbursement for IVIG products as well as ancillary services
- Allowing for home infusion provisions to cover administrative services
- Maintaining pre-administration fees that allow providers to obtain IVIG products

This bill is supported by multiple patient, professional and manufacturer organizations, including IDF, the Clinical Immunology Society, Jeffrey Modell Foundation, GBS/CIDP Foundation International, the Plasma Protein Therapeutics Association, Platelet Disorder Support Association, National Patient Advocate Foundation, ASD Healthcare and Accredo Health.

CALL TO ACTION

Now is the time for the entire IVIG community to contact their Representatives and Senators to urge them to act now to fix the IVIG problem.

Contact your Representative: Contact your Representative and express your pleasure that Democratic and Republican leaders of the Ways and Means Committee pledged to work on getting something done on IVIG issues and ask them to do so this year. Set up a meeting when your Representative is in the district office. Write to your Representative through IDF Action Alert and explain the importance of finding a solution this year. Ask the Representative to press the leaders of the Ways and Means Committee to include an IVIG fix in any compromise legislation that emerges this year, and co-sponsor the legislation.

Contact your Senators: As you did with your Representative, contact both of your Senators and ask them find a solution to IVIG problems this year. Visit them in their home offices when they are home. Write to them through IDF Action Alert and request that they go to leaders of the Senate Finance Committee (which has jurisdiction over Medicare and its provisions dealing with payment for IVIG) and ask that the IVIG problem be included in the Senate’s Medicare package of amendments and in any agreement that emerges from discussions with the House.

IDF Action Alert- www.primaryimmune.org

For more information, call Christine Belser, Senior Director, Programs & Communications at IDF, 410-321-6647.

Three Reasons to Support H.R. 2914, the Medicare IVIG Access Act of 2007

Reason One

H.R. 2914 gives the Secretary of Health and Human Services authority to adjust Medicare's payment for IVIG.

The bill does not tell the Secretary what that rate should be. Rather, it directs the Secretary to review existing data and to collect additional data related to the unique characteristics of IVIG. Then, if appropriate, the Secretary can adjust the payment to cover the costs of IVIG within seven months of enactment of the bill. This provision will help ALL Medicare beneficiaries who use IVIG.

Reason Two

H.R. 2914 will authorize Medicare to pay for the costs of administering IVIG in the home for PIDD patients.

Currently there is a Medicare home infusion benefit specific to patients with primary immunodeficiency disease (PIDD) that is covered under Medicare Part B. Unfortunately, this provision only covers the purchase of the product, and does not cover the costs for administering the product in the home, including nursing and equipment. When Medicare reimbursement was lowered in Medicare Part B, companies could no longer afford to treat PIDD patients, leaving this a hollow benefit. All other Medicare patients who need IVIG are eligible under Medicare's prescription drug program under Part D, and many providers can still afford to treat those patients. So, only PIDD patients are excluded from receiving home infusion, and this is a patient population that must avoid unnecessary exposure to infection in other care settings.

This bill would authorize Medicare to begin paying for the costs of administering IVIG in the home for PIDD patients.

Reason Three

H.R. 2914 will evaluate access to assure that all IVIG users have access to this life saving treatment.

Regardless of how the Secretary may change Medicare's payment for IVIG under H.R. 2914, it is necessary to monitor access for beneficiaries to determine if existing problems are ameliorated, persist, or become worse. The bill requires the Secretary of Health and Human Services to contract for two beneficiary surveys over three years to measure changes in patient access to IVIG and providers, as well as changes in health outcomes.



Thank You...to these Members of Congress for their Support of the H.R. 2914, the Medicare IVIG Access Act 2007

Rep. Kevin P. Brady (R, TX)
Rep. Marsha Blackburn (R, TN)
Rep. Michael C. Burgess (R, TX)
Rep. Barbara Cubin (R, WY)
Rep. Phil S. English (R, PA)
Rep. Luis G. Fortuno (R, PR)
Rep. Wally Herger (R, CA)
Rep. Brian M. Higgins (D, NY)
Rep. Maurice Hinchey (D, NY)
Rep. Steve J. Israel (D, NY)
Rep. Sam Johnson (R, TX)
Rep. Nicholas V. Lampson (D, TX)
Rep. Michael R. McNulty (D, NY)
Rep. Gregory W. Meeks (D, NY)
Rep. Devin Nunes (R, CA)
Rep. Ron E. Paul (R, TX)
Rep. David E. Price (D, NC)
Rep. Jim Ramstad (R, MN)
Rep. Steven R. Rothman (D, NJ)
Rep. Lucille Roybal-Allard (D, CA)
Rep. Pete Sessions (R, TX)
Rep. Christopher H. Smith (R, NJ)
Rep. Ellen O. Tauscher (D, CA)
Rep. C.W. Bill Young (R, FL)

WHO IS JACK?

By Roy and Charlotte Hagelin

At the 2007 IDF National Conference, when Carol Ann Demaret and Dr. Jordan Orange dedicated their comments to the memory of “Jack” the question on many minds was “Who is Jack?” We are Roy and Charlotte Hagelin from Fairfield, New Jersey, and we would like to share the story of our son, Jack, with you.

On September 4, 2002 our family was blessed—Jack was born. He was a beautiful baby, but shortly after his birth, Jack began spitting up and experiencing diarrhea. At five weeks, Jack was malnourished, dehydrated and almost down to his birth weight. He had to be hospitalized.

We were alarmed because Jack’s uncle and male cousin had suffered from these same conditions. There was never any diagnosis for these boys and unfortunately, they died at young ages. Did our Jack have the same condition?

After a 10 week battle of trying to find out what was wrong, the only diagnosis the doctors offered was a “severe milk protein allergy.” Jack’s formula was changed and he improved. However, his lower extremities started to swell, initially starting in his right foot, then spreading through both legs and scrotal area.

Infection after infection, hospital stay to hospital stay, Jack’s “milk allergy” just didn’t seem to be the answer. We saw some of the best doctors on the east coast, traveling from one state to another on a quest for the right diagnosis.

Then another odd symptom appeared. At ten months, when Jack cut his first two teeth, they were pointed (cone shaped). A

pediatric dentist felt this was a result of all Jack’s medications and medical issues. But, after we remembered that the cousin who died as an infant had similar teeth, we grew even more concerned.

A turning point came during one hospital stay when we asked for a dental x-ray. A dental resident suggested that our son had Ectodermal Dysplasia. This led us to a genetic specialist and an immunologist who explained that patients with Ectodermal Dysplasia often also have primary immunodeficiencies. Jack’s medical history fit into the diagnosis. We were then told about Nuclear Factor Kappa B Essential Modulator (NEMO), a rare x-linked primary immunodeficiency.

When Jack was threatened with pneumonia, he had an emergency hospitalization which led us to the Children’s Hospital of Philadelphia (CHOP) and Dr. Jordan Orange. That hospital stay changed our lives. The tests confirmed the NEMO diagnosis and Jack began a regimen of immune globulin therapy. While it was an incredible feeling to finally have a diagnosis, we were also told that most boys with NEMO have a life expectancy of only ten years. When we heard about stem cell transplants, we felt it was the only option for our son to live a long healthy life.

In March 2006, we prepared for Jack’s transplant. A generous stranger was a perfect unrelated donor match, and on March 25, the transplant was done, but did not progress well. Jack’s blood count was not what it should be and Jack wasn’t making enough of the “right” cells to cure his immune issues. Thankfully, the donor again provided bone marrow and Jack was re-transplanted on May 17. This time, all went well and Jack was released from the hospital on June 10th. We were now beginning a new life as a family with a child who we hoped would soon be “cured” from his immunodeficiency.

The next year was filled with weekly visits to the hospital lab to check Jack’s blood count and deal with gastrointestinal issues that stayed in the way of his getting healthy. Then, on March 16, 2007, Jack was hospitalized again. He had been taking medications to suppress his new immune system and developed a cough that led to pneumonia. On March 27, Jack’s little body decided not to fight.

So, this is Jack, our Hero. He gave us life and love. He will remain in our hearts and his legacy will be a part of all of our lives, forever.

This is a short account of Jack’s diagnosis and some of his treatments—there is so much more to the remarkable life of this young boy. You can learn more about Jack and the Hagelin family at their Web site, www.cure4jack.org.



Jack and Sister Christina

What is NEMO?

By Jordan Orange, MD, PhD

Mutation of the gene called NEMO can be a serious cause of primary immunodeficiency. NEMO stands for the “nuclear factor of kappa light polypeptide gene enhancer in B cells essential modulator.” It is more commonly called the “NF- B essential modulator”, or just “NEMO.” Although the name is a mouthful, it is justified by the importance of this gene. NEMO is a key part of a master switch used to “turn on” function in a cell. Many valuable receptors in the immune system use NEMO as a switch to access immune responses. This is sort of like a switchboard operator connecting the caller to the person they are trying to speak to.

The NEMO gene is on the X chromosome and one healthy NEMO gene is all the immune system needs to work. A mutation in the NEMO gene causes immunodeficiency only in boys, because they only have one X chromosome.

Boys who have a mutation in their NEMO gene can have different immune and non-immune problems depending upon where the mutation in the NEMO gene is located. One common non-immune problem affecting about 60% of boys with a NEMO mutation is a condition called ectodermal dysplasia. Boys with ectodermal dysplasia have fine sparse hair, extremely pointy and missing teeth and do not sweat because they lack sweat glands. NEMO mutation is not the only cause of ectodermal dysplasia, but is the main one that is linked to primary immunodeficiency.

The immune systems of boys with a NEMO mutation can be deficient in a number of different ways. This includes effects on the B cells and T cells as well as some of the pre-programmed “innate” defenses of the immune system. For this reason, the NEMO mutation can lead to infections very early in life, as well as some more unusual types of infection. Some of the more unusual infections include “atypical” mycobacteria (bacteria that live inside of cells only and are related to the bacteria that causes tuberculosis) and severe cytomegalovirus (a herpes virus related to the virus that causes “mono” which can cause serious illness in individuals with primary immunodeficiencies). The effects on B cells also frequently cause major gaps in antibody responses and can lead to repeated bacterial infections. These can be very serious and include pneumonia, meningitis (bacteria around the brain) and septicemia (bacteria in the blood). Although we do not completely understand why, a number of boys with NEMO mutations also have very bad diarrhea and inflammation in their intestine. This can look like inflammatory bowel disease.

Since 2000 when NEMO mutations in boys were first discovered, we have learned a great deal. We have come a long way in understanding how to screen for and confirm a diagnosis. We

have also gained valuable insight into treatments that can be useful. These treatments include using immune globulin therapy (IVIG) to provide an antibody defense, and prophylactic antibiotics to help protect against the mycobacteria and other bacterial infections. In addition, certain treatments that try to suppress the inflammation of the intestine and prevent severe diarrhea may be used. In a few boys, blood stem cell transplantation (from cord blood or bone marrow) has been successful in curing the primary immunodeficiency. This, however, is still in its early stages and there is much to learn.

Importantly, we have learned a great deal about what to expect and how to stay “one step ahead” of the complications of NEMO. We now know some preventative measures to use to try and keep boys who have NEMO mutations healthy. Collaboration with patients and families that have been affected by this disorder has been instrumental. They have taught us important lessons and we truly appreciate their openness and generous sharing of experiences. Although we still have ground to cover to gain the upper hand over NEMO, we hope to do better and better.



Jack with Dr. Orange

Dr. Jordan Orange is an Assistant Professor of Pediatrics at the University of Pennsylvania School of Medicine and the Children’s Hospital of Philadelphia. For the past two years, Dr. Orange has served as the chairman of the Primary immunodeficiency Committee of the American Academy of Asthma, Allergy and Immunology and is an active member of the IDF Medical Advisory Committee.



Back row (L to R) Dr. Hans Ochs, Dr. Jennifer Puck, Heather Smith, and John Smith, Front row, Taylor Dahley

Major Announcement—Legislation Introduced!

During the panel discussion on Access and Reimbursement for IVIG, it was announced that U.S. Representative Kevin Brady, along with 23 cosponsors, introduced legislation, the Medicare Immune Globulin Access Act of 2007, H.R. 2914, to the Committee on Ways and Means and the Committee on Energy and Commerce. Spearheaded by IDF, this bipartisan effort seeks adequate reimbursement for IVIG products and accompanying care in the management of patients with primary immunodeficiency diseases and other serious conditions that require IVIG therapy. Thunderous applause and delighted shouts sprung from the audience as participants signed long thank you lists to the bill's cosponsors. (For more information on H.R. 2914, please see page 4.)

First and Foremost—Education and Information

One component of all IDF conferences is the amazing involvement and commitment of our physicians. World renowned physicians shared their knowledge and experience with patients and their families and gave participants not just the opportunity to learn from the best, but viable, important information to take



Carol Ann Demaret and Mike Richardson



Dr. Tony Bonilla, and Dr. Ramsey Fuleihan

home and use. With over 30 scientific and medical sessions to choose from, conference attendees learned about advancements in diagnosis and treatment and often had the chance to personally ask questions of expert clinical immunologists—a rare opportunity for most of us. In addition, two special breakfast symposiums were held, Immune Globulin Therapy on Friday and New Advances: A Molecular and Genetic Approach on Saturday.

IDF collaborated with Washington University in St. Louis and the Missouri Nurses Association to offer continuing medical education program for healthcare professionals. Providing landmark research and breakthrough information on primary immunodeficiencies, leaders in immunology lectured on recent developments in diagnosis and treatment to the 73 attendees. First indications of the medical education program's success came almost immediately from attendee evaluations. Based on information from these lectures, the majority of attendees stated they were now more likely to begin screening for primary immunodeficiencies—which is great news for improving early diagnosis!

On Saturday, life style sessions and discussions offered advice on managing healthcare as well as dealing with social and emotional needs. From family dynamics to employment rights; parenting to travel considerations, the presentations offered new skills and covered a myriad of interests.

And the Conference wasn't just for adults-- nearly 200 young people, ages 6 – 17, attended the youth programs that featured the benefits of learning while having fun. Children, aged 6 through 11 became doctors for a day at the Kid's Clinic where they performed "surgery" on a model of a teddy bear. But, they also became the super heroes, "Germ Busters" and learned valuable skills to cope with their unique challenges of growing up with primary immunodeficiencies. Older youth, 12 to 17 year olds, learned how to thrive as they emerge into adulthood with "Primary Immunodeficiencies and Treatment

Options,” “Be Your Own Advocate,” “Deal with It! Coping Skills” and “Ask the Expert Seminar.” Finally, the Science Center opened its doors to all youth on Friday, where the participants could experiment with science activities.

We are all in this together!

Individuals and families living with primary immunodeficiency often describe feeling isolated because there are few people who can understand what they are going through, and for many, the National Conference was the first time they had ever met anyone else from the primary immunodeficiency community. However, feeling isolated was hardly a problem at the National Conference. Packed with people who all shared experiences with dealing with primary immunodeficiency disease, new relationships were formed as attendees exchanged ideas, experiences and support. As one first time attendee stated, “GREAT! As a newly diagnosed patient, it was very helpful to talk to other people who are going through the same thing. Topics for the breakout sessions were a nice variety.”

On the other hand, many old friendships were rekindled. The exclamation of one returning participant was representative of many, “What a wonderful conference again! You know I honestly thought that I might not attend because I went to the last three, but I’m glad I went. There is always something new to learn, contacts to be made and old friends to see again.”

Always Time for Fun

Throughout the conference, there were opportunities to relax and have a little fun. All IDF Conference participants were treated to “Catch the Spirit...Celebrate IDF!” on Friday night. This very special evening of music, dinner, and riverboat “gambling” in celebration of the IDF community, was sponsored by Talecris Biotherapeutics. Then on Saturday, Baxter Healthcare hosted “Zoobelation,” an evening touring the Saint Louis Zoo, riding the zooline railroad and the conservation carousel, complete with a Missouri barbecue dinner and music.

As a patient-focused organization, IDF is proud of the patients and their families, medical and nursing communities and everyone in the primary immunodeficiency community. From the overwhelmingly positive feedback we have received from conference attendees, it appeared that most attendees felt the same way about the fourth National Conference. “Inspiring”, “terrific” and “awesome” are some of the adjectives used to describe the conference; however, one attendee summed it up best:

“Invaluable information from speakers and met other families exactly like mine... priceless!”

*Maggi Dodd, CPNP
and
Debra Sedlak, CPNP*



*Dr. Michael Blaese,
Sonia Vohnout, and
Steve Fietek*



Lindsey Nerem, Barb Cobler and Dallas Jackson

IDF NATIONAL CONFERENCE AWARDS

The Boyle Achievement Award pays tribute to a member of the medical/scientific community who has extensively contributed toward improving the diagnosis and care of patients with primary immunodeficiency diseases. This year, it was awarded to **Charlotte Cunningham-Rundles, MD, PhD**, who has spent much of her career investigating the causes and treatment of primary immunodeficiency diseases. She is world renowned for her research on the syndrome of common variable immunodeficiency. Currently, Dr. Cunningham-Rundles is Professor of Medicine and Director of the Allergy and Immunology Training Program at Mount Sinai School of Medicine in New York City. She also supervises both the Immunodeficiency Clinic, a referral service for children and adults with primary immunodeficiency disease and the Clinical Immunology Infusion Service where patients with CVID and other antibody deficiencies are treated. IDF is particularly proud that Dr. Cunningham-Rundles serves on the IDF Medical Advisory Committee and pleased to honor her with this prestigious award.

Sponsor Awards

Baxter Healthcare Corporation and **Talecris Biotherapeutics** were honored for their Legacy Sponsorship, the highest commitment to the conference. **CSL Behring** was recognized as a Platinum Sponsor and **Grifols, USA** was acknowledged for their Silver Sponsorship. In addition to supporting this conference, these companies have generously supported IDF throughout the year as Core Sponsors, providing IDF with a base of funding to help support programs that are integral to its mission and goals. These include our direct services for individuals and families, essential medical and scientific programs, public policy program and survey research.



Award winners Ray Liu; Paul Perreault, Sr.; Bruce Bunyan; Nancy Hoffman; Marcia Boyle; Jordan Orange, MD, PhD; Pete O'Malley

Volunteer Awards

The first volunteer award was for Fundraising and was awarded to **Evelyn Maselli** and **Terri Lowell**. Although they could not attend, they were honored for their dedicated and enthusiastic fundraising activities in Connecticut over the past seven years.

Nancy Hoffman, an IDF Volunteer from California, was recognized for her outstanding Peer Support. Nancy is always willing to provide peer support to anyone who needs it. She seems to never get tired of requests from IDF and is always happy to help in any way.

The final volunteer award was given for Advocacy to **Jordan Orange, MD, PhD**, Assistant Professor of Pediatrics at the University Of Pennsylvania School Of Medicine. IDF applauds Dr. Orange for taking an active role in the ongoing education and advocacy efforts to ensure that patients have access to IVIG in all sites of care, as well as access to the most appropriate care.

SPECIAL THANKS TO IDF 2007 NATIONAL CONFERENCE PRESENTERS

Loris Aro, RN
Barbara Ballard
Mark Ballow, MD
Troy Balthazor
Melvin Berger, MD, PhD
Kim Bernstein, RN
Julie Birkofer
R. Michael Blaese, MD

Francisco (Tony) Bonilla, MD, PhD
Erin Boyle
Rebecca H. Buckley, MD
Sharon Byrd
Janet Christensen
Lisa Codespoti
Mary Ellen Conley, MD
Richard Darby

Maggi Dodds, CPNP
Kimberly Duff, RN, BSN
Kristin Epland RN, CNP
Carol Ernst, RN
Thomas Fleisher, MD
Michael Frank, MD
Katie Franks
Ramsay Fuleihan, MD

Erwin Gelfand, MD
Terry Halper
Theresa Halper
Leslie Harman, MA, NCC
Terry Harville, MD, PhD
Mike Hershfield, MD
Nancy Hoffman
Steven Holland, MD

YOUTH LEARN LIFE LESSONS

Not only were there scheduled youth activities at the National Conference, there were also plenty of opportunities for young people to learn in other ways. In one case, a four-year old girl approached a climbing wall with her dad. When asked if she wanted to climb it, she looked up at the 12 foot wall and said, "Well, maybe a little later, like when I am sixteen." However, she was intrigued--she wanted to climb to the top and ring the bell.

After a little encouragement, she was in the safety harness and began her climb. About a third of the way up, she started to get scared and wanted to get down. Even though she didn't get to the top, everyone clapped and cheered for her.

Later, she came back and wanted to try again. This time she got about two-thirds of the way up and came down to much cheering and applause. Finally, near the day's end, she returned for her third try. Slowly and with some degree of difficulty, she climbed to the

top and rang the bell. Well, you never saw a child more proud and happy--she had the biggest, widest smile!

Once she got down, she ran over to her dad and shouted, "I did it! I did it! Did you see me Dad? Did you see what I did? I climbed all the way to the top Dad! I rang the bell!" Needless to say, everyone cheered and applauded her achievement.

This little girl wasn't alone. In climbing the wall, many children gained self confidence. It also taught them to feel strong and independent and believe in themselves. There is a lesson we can all learn from this, find your own personal Mt. Everest and conquer it!

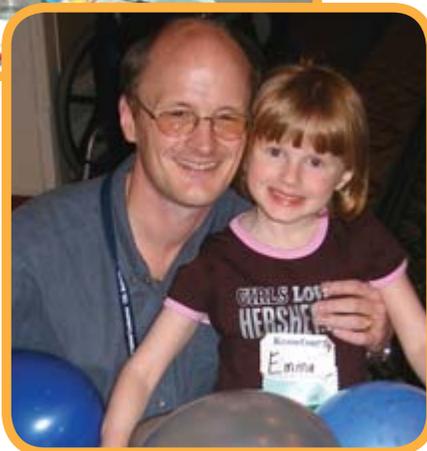
Face painting at the St. Louis Zoo



Towma Rastad, with her children Alec and Alissa



Rayni Million-Worley posing with penguins at the St. Louis Zoo



Arlan and Emma Sprague



Front row (L to R), Izabelle Harville, Gabrielle Harville, Ray Ballard, Forest Madden. Back row (L to R), Jean Harville, Dr. Terry Harville, Amanda Madden-Dyal, Barb Ballard, Don Ballard

Janet Hoskins
 Roger Kobayshi, MD
 Donald Kohn, MD
 Howard Lederman, MD, PhD
 Harry Malech, MD
 Peter Mannon, MD
 Diane Mato
 Barbara Meade

Susan Nissen, MD
 Hans Ochs, MD
 Jordan Orange, MD, PhD
 Jennifer Puck, MD
 Sue Rantham
 Towma Rastad
 Brian Rath
 Terry Rayburn, RN, BSN,ACRN

John Routes, MD
 Richard Schiff, MD, PhD
 Quentin Seals
 Debra Sedlak, CPNP
 John Seymour, PhD
 Ralph Shapiro, MD
 Yvette Shorten
 Jermaine Shorten

Gloria Slater, CSL
 E. Richard Stiehm, MD
 Kathleen Sullivan, MD, PhD
 Lynne Szott, RN
 Lisa Waugh
 H. James Wedner, MD
 Scott Zeigler, RPh

CALENDAR OF EVENTS

September 8, 2007

Patient Education Meeting
Hoagland Pincus Conference Center
Shrewsbury, MA
9:00 am-2:00 pm
Speaker: Francisco (Tony) Bonilla, MD, PhD,
Boston Children's Hospital
For more information contact Towma
Rastad at 508.596.3746 or email
allissalec@yahoo.com

September 15, 2007

IDF Center of Excellence Patient Education
Meeting
Duke University Medical Center
Searle Center, Room E
Saturday, September 15, 2007
9:00 a.m. to 2:30 p.m.
Speakers: Rebecca Buckley, MD, Laurie Lee,
MD and Joseph Roberts, MD, PhD
For more information, contact Diana Gill
at 443.632.2545 or email
dgill@primaryimmune.org

September 15, 2007

IDF Milwaukee Conference
Milwaukee County Zoological Gardens
Zoofari Conference Center
Milwaukee, WI
Time: 9:00 am-3:00 pm
Speakers: William Grossman, MD, PhD
and John Routes, MD, Medical College of

Wisconsin
For more information contact Linda
Keegan at lindakeeganwi@aol.com.

October 10-11, 2007

LeBien Visiting Professor
University of South Florida
Tampa, FL
For more information contact Tamara
Brown at tbrown@primaryimmune.org

October 13, 2007

Operation Outreach Patient Meeting
Nashville Airport Marriott
Nashville, TN
9:00 am-2:30 pm
Speakers: John Fahrenholz, MD and Keegan
Smith, MD, Vanderbilt University Medical
Center and Lynne Szott, RN, Manager
Reimbursement Ig Products, CSL Behring
Please RSVP no later than October 8, 2007
by contacting Diana Gill at 443.632.2545 or
email dgill@primaryimmune.org

October 18-19, 2007

LeBien Visiting Professor
University of West Virginia
Charleston, WV
For more information contact Tamara
Brown at tbrown@primaryimmune.org

October 27-30, 2007

American Association of Pediatrics

San Francisco, CA
IDF will be an exhibitor

October 27, 2007

IDF Family Conference Day
Science Museum of Minnesota
St. Paul, MN
For more information contact Kathy
Antilla at 763.444.6840, or
kantilla@primaryimmune.org

November 1, 2007

LeBien Visiting Professor –
Mary Ellen Conley, MD
Children's Hospital of Pittsburgh
Pittsburgh, PA
For more information contact Tamara
Brown at tbrown@primaryimmune.org

December 5, 2007

Milwaukee Area Support Group for
Parents of Children with Primary Immune
Deficiency Disorders
Milwaukee, WI
6:30 p.m. to 8:30 p.m.
For more information contact Linda
Keegan at lindakeeganwi@aol.com

June 18-20, 2009

2009 IDF National Conference at
Disney's Contemporary Resort in
Lake Buena Vista, FL

With Gratitude

The Immune Deficiency Foundation gratefully acknowledges those who generously contribute to the organization to celebrate the memory of someone who has died or to honor someone special.

Gifts In Memory Of

Roy M. Davis	Nadine F. Hersh
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	Eric Marder
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Paula Rottman
Jacquiline Routman
Jerome Routman
Doris Singletary
Kevin Song
Lee Zlotowicz

Gifts In Honor Of

John G. Boyle	Celina Frelinghuysen	Eugene Solar
Cheryl Cario	Christopher Kluxen	Scott Solberg
Irene Chen	Gregory Kendall Pillon	Aaron Staebell
Lillian Feeley	Arye Rubinstein	Elliot Uecker
Landon Feniger	Sarah Rumsey	John Wettstein, Jr.
	Helen K. Sexton	



These donations help IDF to improve the diagnosis and treatment of patients with primary immune deficiency diseases through research, education and advocacy. If you would like to make a donation, please go on our Web site, www.primaryimmune.org, click the "How You Can Help" tab and use the secure online server. You can also contact us in any of the following ways:

Phone: 800.296.4433 or 410.321.6647

E-mail: idf@primaryimmune.org | Mail: IDF, 40 W. Chesapeake Avenue, Suite 308, Towson, MD 21204

Marcia Boyle visits New Zealand Immune Deficiencies Foundation Meeting

During July 2007, IDF President and Founder, Marcia Boyle traveled half-way around the world to be with the Immune Deficiencies Foundation (IDFNZ) and KIDS Foundation of New Zealand for their annual meeting in Auckland, New Zealand. As keynote speaker, Marcia presented information on IDF and the issues and challenges of U.S. healthcare. The three day conference featured educational and life skill sessions, presented by immunologists and other experts, activities for children, and even a little tenpin bowling.

Marcia commented, "New Zealand is a long way from home, but the concerns and issues of IDFNZ are not very different from what IDF faces in the U.S. It really is all about the patients and families living with primary immunodeficiencies-- advocating for their needs and helping them get the services and information they need. It was a wonderful experience to be able to spend time with these dedicated people."

IDFNZ supports children, teenagers and adults with primary immunodeficiency disorders in New Zealand and the Kids Foundation is the welfare arm of IDFNZ and is responsible for caring for suspected or diagnosed children, teenagers and their immediate families, as well as bone marrow transplant children. The Reverend Stephen Baxter, Chairman of IDFNZ, serves with Marcia Boyle on the Board the International Patient Organisation for Primary Immunodeficiencies (IPOPI).

Over the years, IDF has worked closely with IDFNZ and IPOPI in support of the efforts of the Asia Pacific Alliance to develop strong patient organizations in the Asia-Pacific region of the world. In the last two years, this partnership has helped develop The Immune Deficiencies Foundation of Australia.



Marcia Boyle hands out an award to a student. Joining Marcia are Sarah Perry and Stephen Baxter from the IDFNZ.

Volunteer Activities



Stephanie Bush, IDF Volunteer, along with family and friends, represented IDF and helped create awareness about primary immunodeficiency diseases in the Fourth of July parade in Chelmsford, MA.



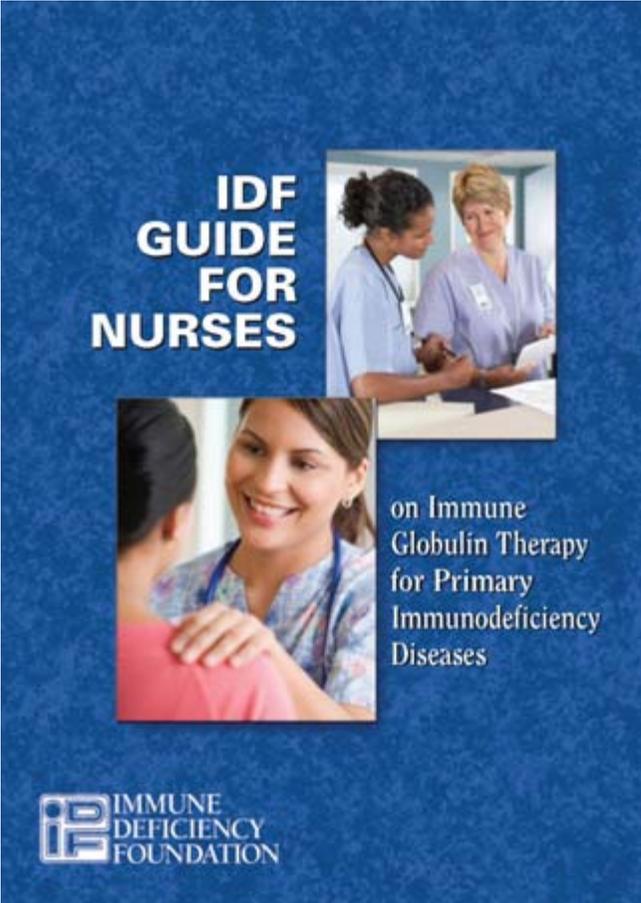
The Steffes family (l to r) Ken, Liz, Lisa and Danny organized a special "Under the Sea Benefit" at the Minnesota Zoo. Over 200 family members and friends enjoyed a wonderful evening that featured a silent auction, music and fun. Congratulations to the Steffes for hosting this joyous event that raised \$22,000 for IDF!

NEW IDF GUIDE FOR NURSES

IDF is pleased to present the new, updated and expanded IDF Guide for Nurses on Immune Globulin Therapy. Based on recent advances in nursing care and treatment, our Nurse Advisory Committee revised the popular guide, which was originally developed in 2004. The new Guide provides direction on the administration of immune globulin replacement therapy in the safest, most effective way. Since most IVIG are administered by nursing professionals, nurses are in a unique position to improve the treatment experience and provide an improved quality of life for patients. The Guide also covers the important role nurses play in providing education and evaluation for patients who choose subcutaneous immunoglobulin replacement therapy (SCIG).

Guide highlights include clinical uses for immune globulin replacement therapy; product selection and characteristics; infusions, complications and adverse events of IVIG and SCIG; nursing interventions for adverse events; concomitant medications; and nursing responsibilities. The reference and resource listings underscore relevant information and where to find it.

The Guide is available on the IDFWeb site at www.primaryimmune.org. Nursing professionals may contact IDF at 800.296.4433 or idf@primaryimmune.org to order this free booklet. The IDF Guide for Nurses was made possible by an unrestricted educational grant from Talecris Biotherapeutics.



**IDF
GUIDE
FOR
NURSES**

on Immune
Globulin Therapy
for Primary
Immunodeficiency
Diseases

IMMUNE
DEFICIENCY
FOUNDATION

Is it just an infection?

You should be suspicious if you have an infection that is...

Recurrent
keeps coming back

Persistent
won't completely clear up or
clears very slowly

Severe
requires hospitalization or
intravenous antibiotics

Unusual
caused by an uncommon organism

Runs in the Family
others in your family have had a
similar susceptibility to infection

If any of these describe your infection, ask your physician to check for the possibility of a primary immunodeficiency disease. These diseases are caused by genetic defects and can affect anyone, regardless of age or sex. People with primary immunodeficiencies are more susceptible to infections and health problems that lead to serious and debilitating diseases. It is critical to get an early diagnosis and proper medical care.



IDF Can Help

Immune Deficiency Foundation (IDF) is the national organization dedicated to patients living with primary immunodeficiency diseases. Contact IDF to learn more.

800.296.4433 • idf@primaryimmune.org • www.primaryimmune.org

New Poster Fosters IDF Awareness and Call to Action

IDF is pleased to offer a new poster that cautions people to be suspicious if they have a recurrent, persistent, or severe infections or if a particular infection appears to be hereditary. The flyers alert individuals about the possibility of a primary immunodeficiency and encourage them to check with their physician.

The colorful posters are perfect for bulletin boards in physician offices, schools, or other public places that will catch the attention of people in the community.

To receive free posters and help spread awareness about primary immunodeficiency diseases, please contact IDF at 800.296.4433 or idf@primaryimmune.org.

INDUSTRY NEWS

Talecris Biotherapeutics Institutes a Portability Policy for Gamunex®

In response to evolving needs expressed by the patient community and healthcare providers, effective immediately, Talecris Biotherapeutics, Inc. has implemented a Portability Policy for Gamunex®. This policy better reflects and responds to current market dynamics by accommodating changes in customer relationships, sites of service and points of care. Talecris' goal in implementing the Portability Policy for Gamunex is to help ensure patients who rely upon Gamunex continue to receive it reliably and at their preferred location in situations where their product supply channels or sources of distribution have changed.

To help ensure continuing availability of Gamunex, there are several delivery options for patients and their physicians that will result in fewer treatment disruptions. Although the exact details of the portability process vary from customer to customer and are subject to various conditions, purchases of Gamunex generally can be transitioned from one Talecris contracted organization to another, or to a direct contract with Talecris. *Excerpted from Talecris Biotherapeutics News Release, Aug. 2, 2007*

CSL Behring Announces FDA Approval of Privigen™

CSL Behring announced that the U.S. Food and Drug Administration (FDA) has granted marketing approval for Privigen™ (Immune Globulin Intravenous (Human), 10% Liquid), an intravenous immunoglobulin (IVIg) for treating patients diagnosed with primary immunodeficiency (PI). Privigen is also indicated for the treatment of chronic immune thrombocytopenic purpura (ITP) to rapidly raise platelet counts to prevent bleeding.

A 10 percent liquid preparation of polyvalent human immunoglobulin, Privigen offers healthcare professionals convenience and ease-of-use. It is the first and only proline-stabilized IVIg that is always ready for immediate use, requiring no refrigeration or reconstitution. CSL Behring plans to launch Privigen in the first quarter of 2008. *Excerpted from CSL Behring News Release, July 27, 2007*

Baxter To Launch Gardian™ To Help Provide Continued Access To Gammagard For Current Patients

Baxter International, Inc. announced that it will launch GARDian, an innovative program that helps provide continued access for patients relying on Gammagard Liquid and Gammagard S/D [Immune Globulin Intravenous (Human)] (IGIV). IGIV products like Gammagard Liquid and Gammagard S/D are immunoglobulin (IgG) replacement therapies containing broad-spectrum immunoglobulins and are indicated for patients with primary immunodeficiency. Primary immunodeficiency is a group of genetic disorders in which the immune system fails to produce adequate amounts of antibodies, thereby predisposing individuals to increased risk of infection.

IGIV users and physicians can experience changes involving their providers or site of care that can sometimes impede access to their preferred IGIV therapy. Baxter's GARDian program, which is expected to start early next year, will provide patients and physicians with continued access to Gammagard Liquid and Gammagard S/D even when such changes occur. *Excerpted from Baxter International Inc. News Release, August 22, 2007*

IDF Visits Biomat Center



The Biomat Center in Reading, PA participates in the IDF program Blue Jeans for Healthy Genes. Attending a press conference to celebrate their participation in Hanging-Up Jeans are (left to right) Corey Stein, Reading's Biomat Center Facility Manager; Marcia Boyle, IDF President and Founder; and Chris Healey, VP, Government and Public Affairs, Grifols.

NEW IDF ACTION ALERT--

Encourage your Representative to cosponsor H.R. 2914, the Medicare IVIG Access Act 2007, and ask both your Senators and Representative to include a Medicare IVIG provision in Medicare Legislation this year.

Visit our Web site, www.primaryimmune.org, and click on "IDF Action Alert" to personally send out the new alert to Congress.

Circle of Support 2007

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The Circle of Support consists of organizations committed to supporting the Immune Deficiency Foundation at the highest level.

The Circle provides vital resources to fulfill the vision, mission and core services of the organization.



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