I WANT TO BE A PART OF IT...NEW YORK!

IVIG, Pemphigus & Pemphigoid
Learn more about IVIG and its future in treating pemphigus and pemphigoid.
See page 4

Are You Registered?
The IPF’s 9th Annual Patient/Doctor Meeting will be held in New York City September 15-17, 2006.
See page 7

In this Issue
2 The View from Here
3 A Patient’s Story: We Can Make a Difference
4 Treating Pemphigus and Pemphigoid with IVIG
7 Annual Meeting Registration Form

Look inside for your REGISTRATION FORM (inside back cover) and mark your calendars! The IPF and PPS are heading to New York City for the biggest and most important Patient/Doctor meeting in pemphigus and pemphigoid history!

This year’s meeting is at the New York University School of Dentistry. This event is highlighted by a Gala Dinner (with a special celebrity guest event) on Saturday evening at Manhattan’s famed Water Club Restaurant.
WORDS FROM THE PRESIDENT AND EXECUTIVE DIRECTOR

GROWTH CAMPAIGN PAYING OFF

The IPF Growth Campaign is showing signs of continued growth! To date, the campaign has raised over $30,000 and we are striving for more! The more we raise the more we can enhance operations and increase efforts to provide support, fund research, and grow the Foundation as a whole. Thank you to all who have dug deep and given to a great cause - each other!

The Annual Meeting is nearly upon us. If you haven't made your reservations yet now is the time. This year's event will be the largest gathering of pemphigus and pemphigoid patients in history! Add to that the information offered, exciting events, and host of experts on hand you cannot afford not to go. Don't forget the Gala Dinner on Saturday night to celebrate the unification of the IPF and the Pemphigus & Pemphigoid Society under one umbrella. During the dinner we will present our Annual Awards, have some outstanding entertainment, and hear and honor some special guests. This is a true New York City celebration that will rival anything you have ever seen.

The cost for all of this is $125 and includes the most up to date information, weekend public transportation pass, interaction with patients, caregivers, and doctors, and the chance to grow as a "family." I look forward to seeing everyone there! If you can't attend help someone who would like to but can't by sending in a Scholarship Donation. Don't forget, without YOUR support we can't support others! ●

JOIN US IN NEW YORK!

Most of this issue of the Quarterly is dedicated to our upcoming meeting which will be held in NY on September 16-17, 2006. This is going to be an exciting meeting with a gala dinner at a wonderful club in New York along the water front. The meeting will include information and updates on the clinical trials now going on as well as 2 Q&A sessions with the doctors. Finally, we will have a half day on Sunday for patient to patient interaction.

Dinner will include a Jazz band, awards, and an honorary guest. We ask everyone supporting the Foundation to come and enjoy a wonderful weekend in NY. NY is a great city and one that everyone should experience at least once in their lifetime. The Museum of Natural History and the Metropolitan Museum of Art are two of the most prestigious museums in the world. Of course, there is Broadway with the best plays and musicals, Rockafeller Center, Lincoln Center and the hub of international finance: Wall Street. Visit the Empire State Building or ake a tour around Manhattan, head up the Hudson River, or visit the Statue of Liberty. And, in-between all that, learn about the IPF’s Growth Campaign.

I know I sound like a tour guide for NY, but I was born in NY and love the city - I just don't like the cold that much! This will be a great opportunity to help the IPF and the PPS celebrate their merger, learn from the experts and have a great time as well. Some people have been reluctant to come to the annual meeting because of the fear of being depressed from discussing the disease or seeing people with lesions, but I have been doing these meetings for 9 years, and accept for one or two, most of the comments about our meetings have been nothing but positive. Join us this year and you will leave with more hope than ever before! ●
HOW CAN YOU HELP IMPROVE TREATMENT: VOLUNTEER!
WE CAN MAKE A DIFFERENCE

by Alan Papert
AKA Alan PV Boston

Every one of us wants to be treated by the most experienced dermatologist, and collectively we aim to collate a list of these doctors in each region of the country. However, by sending every new pemphigee (shorthand for a pemphigus or pemphigoid sufferer) to an experienced doctor, there would be very few opportunities for the next generation to reach that level.

Amongst other things the IPF is working to increase awareness in the medical schools to prevent the above scenario, and there is a very real way that we, as patients, can make a contribution, and that is by volunteering to appear before classes as examples for the students.

Two years ago, I had such an opportunity. A class of dental students at Harvard Medical School was being introduced to pemphigus. After his introduction, the professor asked me to explain how the disease first manifested itself. The students then took over asking in depth questions about how I was diagnosed, my treatment regime, how I felt at different stages of my illness, etc.

In the interim between my visit being arranged and the actual day my symptoms had practically disappeared, so the students still had to rely on pictures to see the physical effects.

As I was leaving the lecture theatre, I was approached by a number of the students wanting to know how much pain I had felt, the emotional experience I had been through, etc. I believe that many members of that class will, when confronted with a pemphigee, be more likely to recognize that fact now. They also know that there is an up-to-date source of information available to them on the IPF web site, which, I am sure, was visited by many of them after the class. Hopefully, they were also sensitized to the emotional needs of the patients.

I urge those pemphigees who are being treated by a doctor associated with a Medical school, to volunteer themselves to be used as living examples for classes. Maybe we can set up a mechanism through the IPF, its support groups, and/or the medical professional societies, to put the professors in touch with people with active flares.

All this still leaves those patients who live far from the research centers and in small communities where the doctors have had no chance to treat pemphigees. If these patients are not in a position to travel to an experienced doctor, their salvation should be in the spread of Tele-medicine which enables their doctor to consult electronically with an expert. As an engineer who worked on some of the early technology to make this possible, I am disappointed that it has taken so long to spread. I know that the issue of patient privacy is a major stumbling block, but, hopefully, all the roadblocks are being addressed, since the benefits are considerable.

We all should work to have this sort of collaboration built into our medical support system. It will never be as good as being there, but it is far preferable to the following scenario. A pemphigee, for whom I have been providing support, was told by her doctor that there is no cause or cure for pemphigus and that she was “mad to suggest that stress could be making it worse.”

All Rights Reserved.

Don't forget to mark your calendars for the Patient/Doctor Meeting in New York City! September 16-17, 2006!
IVIG HOME TREATMENTS ARE ON THE RISE. IS IT THE RIGHT CHOICE FOR YOU?

TREATING PEMPHIGUS & PEMPHIGOID WITH IVIG

Michelle Greer, RN, MBA
Michael T. Rigas, PharmD
Crescent Healthcare, Inc.

“The information provided in this article has been edited by Drs. Jean-Claude Bystryn and Sergei Grando. There are several companies that provided IVIG at home. The decision to utilize IVIG at home and what company to use, should be made between you and your treating physician.”

IVIG stands for intravenous immunoglobulin. Immunoglobulins are also known as antibodies, which are produced by the body to activate the immune system to attack virus and bacteria. IVIG is a medication that is a blood product. It is made from the plasma of thousands of carefully screened donors. 90 – 98% of IVIG is made up of Immunoglobulin G (IgG). IgG is a Y-shaped protein that is the main antibody found in blood plasma. Its role is to neutralize toxins and cause bacterial and viral inactivation. There are small amounts of other antibodies (IgA, IgE, IgM) found in IVIG products. There are many different brands of IVIG. The main differences are the IgA content, the type of sugar found in the solution and the methods use to protect the patient from viral transmission. The pH also varies depending on the brand.

There are several FDA-approved or labeled uses for IVIG. They include:
- Primary Immune Deficiencies
- Chronic Lymphocytic Leukemia
- Children with HIV
- Allogeneic Bone Marrow Transplants
- Kawasaki Syndrome
- Idiopathic Thrombocytopenia Purpura

There are even more diseases and disorders that are considered standard treatment that aren’t FDA-approved, also called off label use. The majority of these are autoimmune diseases and disorders, including pemphigus and pemphigoid.

When IVIG is given for immune deficiencies, it is known as replacement therapy. This simply means the person lacks or doesn’t produce enough antibodies, and IVIG is given to make up for the deficiency.

Autoimmune diseases occur when the body fails to recognize itself and produces antibodies that activate immune response to attack one or more systems of the body. These antibodies are also called autoantibodies. Autoimmune diseases are also known as immune-mediated diseases or antibody-mediated autoimmune diseases.

Pemphigus is a rare autoimmune disorder in which antibodies are produced resulting in damage to cells in the upper part of the epidermis. This causes lesions or blisters to appear on the skin and/or mucous membranes of the mouth, nose, and eyes. Pemphigus refers to this group of blistering diseases known as Vulgaris, Foliaceus, and Paraneoplastic.

Pemphigus Vulgaris (PV), is the most common and accounts for about 70% of all cases. It tends to be more common in people of Mediterranean descent. Pemphigus Foliaceus (PF) has six subtypes, and its incidence varies, but overall it is very rare and sporadic worldwide. Pemphigus, Paraneoplastic is found in patients with underlying neoplasms. Pemphigus can occur in any age group, but most commonly in ages 50 – 60.

Symptoms of pemphigus depend on the type and where the lesions are. In PV, mucous membranes are frequently affected first, and it may be months before skin blisters appear. A lot of people with PV might actually be seen and diagnosed by an oral surgeon. PV on the skin starts out as very fragile blisters that rupture quickly, causing painful, raw erosions on the skin. In PF, the mucous membranes are rarely involved, and the blisters usually appear on the trunk first. They are small and superficial and also rupture quickly causing skin erosions which become scaly and crusted. Pain is more localized.

Pemphigoid is another rare autoimmune disorder in which the body perceives skin proteins between the dermis and epidermis as foreign and attacks them. This also causes lesions or blisters to appear on the skin and/or mucous membranes of the mouth, nose, and eyes. There are two types of pemphigoid: Bullous and Cicatricial. In Bullous Pemphigoid (BP), it’s mainly the skin that’s affected. It occurs more in people age 50 – 70, although there have been cases in children and people of any age. Lesions tend to clear up with treatment without scarring. Sometimes lesions can clear up without any treatment. In Cicatricial pemphigoid (CP), the mucous membranes are frequently affected in addition to the skin, and scarring is more common. Because of this permanent damage to the eyes (visual problems) and throat (hoarseness, airway obstruction) can occur. CP occurs most frequently in the early to mid-60’s.

Again, symptoms of pemphigoid depend on the type and location of the lesions. Patients with BP may present in several different ways, the most common being generalized, where the lesions can show up on any skin surface, with tendency toward the joint flexure areas. They can be painful and itchy. If mucous membrane involvement occurs in BP, it is less severe than in CP. Patients with CP experience painful erosions of mucous membranes. Depending on where these erosions are, symptoms include pain or a “gritty” sensation in the eyes due to decreased tear production, mouth blisters, hoarseness, difficulty swallowing, bleeding after blowing the nose, as well as skin lesions.

Besides assessing the skin and

continued on page 5...
mucous membranes lesions, there are two tests that are done to diagnose pemphigus and pemphigoid. The first is a skin biopsy from the edge of a blister. The second test is called immunofluorescence studies, which can be direct and indirect. This involves shining a special light onto the microscopic samples of skin around blisters and the skin around them looking for telltale responses. Sometimes it is a dentist or oral surgeon that sees these symptoms first.

Once the diagnosis is made, pemphigus is treated with steroids and/or drugs that curb immune system response, or immunosuppressants. Because of the damage to the skin, there is a lot of inflammation. Steroids are very powerful anti-inflammatory agents. Immunosuppressants are prescribed to prevent or decrease the production of the autoantibodies. In many cases, this is all that is needed to remit and even resolve the condition. However, these drugs have side effects and with long term use, can create other problems.

Studies have shown that IVIG is effective in treating pemphigus. Several treatment courses are usually needed for the desired therapeutic effect. In fact, one study concluded that treating mucous membrane pemphigoid with IVIG can actually end up being less expensive than treating with steroids and immunosuppressants because there were fewer side effects with the patients receiving IVIG. The side effects associated with immunosuppressants created other issues that made the course of the disease more expensive in the long run.

Why IVIG works in autoimmune diseases is much more complex than simply replacing a deficiency. When IVIG is administered in these instances, it is known as immune modulation. In other words, by administering IVIG, the goal is to stop the immune system from producing autoantibodies. There are several theories why IVIG is effective in doing this. IVIG is given in high doses in these instances. The theory is that if enough healthy IVIG is given the person’s autoantibodies will not have a chance to cause the response in the immune system that causes the autoimmune disease.

IVIG is given intravenously, directly into the veins. The dose depends on what the IVIG is being given for and is also based on body weight. To treat pemphigus, the doses are as high as 2000mg/kg. Because the doses are higher, they are divided into infusions that are given over the course of up to five days, and they can be consecutive or nonconsecutive days.

IVIG has a half life that is on average 30 days, depending on the brand and the person. Half life means the point at which the IVIG concentration in the blood falls to half of its original value. That is why IVIG doses are repeated at regular intervals of monthly or every two to four weeks. Many times this regimen is indefinite or even life time. In other cases, the condition resolves and the IVIG can be discontinued.

If side effects are intolerable or severe, or if the veins are such that getting an IV in place is not possible, IVIG may be administered subcutaneously. In this case a needle is placed under the skin and the medication is given. These infusions are given more frequently and take longer because the skin cannot handle the volume of fluid that the veins can. This route is more common in children where the dose is low enough that the skin can absorb the volume successfully. However, for any patient, if the IVIG route is no longer viable, and the patient needs IVIG, the subcutaneous route may be a viable alternative.

In general, IVIG is considered to be safe, and the majority of people tolerate it without problems. The adverse reactions occur only in less than 1% of patients. Patients with pemphigus and pemphigoid who suffer from side effects of steroid therapy have a higher risk. Most of the side effects occur because it’s administered too quickly. Because of this, it is gradually infused, starting at a very low rate and increased at intervals until the maximum rate is reached. Many times, if side effects are experienced, simply reducing the infusion rate will eliminate them. Side effects can also be offset by taking premedications, such as diphenhydramine (Benadryl®), acetaminophen (Tylenol®), aspirin, and even IV hydration fluids. In addition, side effects usually subside as the body adjusts to the IVIG. Other more severe reactions can be minimized but not always prevented by careful prescreening and choosing the appropriate brand.

Side effects are manageable and may include:

- Headache
- Fever
- Fatigue
- Chills
- Flushing
- Dizziness
- Urticaria
- Chest Tightness
- Nausea and Vomiting
- Muscle cramping
- Blood Pressure changes

There are other reactions that more severe but rare. These include:

- Anaphylaxis – a severe allergic reaction. People at higher risk for this type of reaction would be premedicated. At least one dose would need to be given in a medically supervised setting, such as a doctor’s office or a hospital infusion center. The level of IgA should be checked to avoid anaphylactic reaction to IgA present in the IVIG preparation.
- Aseptic Meningitis Syndrome – severe headache that resolves when IVIG is stopped.
- Thrombotic Event – a blood clot forms because the IVIG is slightly thicker than blood. Certain conditions or something in the medical history may make a person susceptible to clot formation. People at higher risk would be premedicated with aspirin, or a stronger prescription blood thinner. Some people may benefit from hydration before IVIG administration.
...continued from IVIG, page 5

- Renal Failure – potential problem for people with underlying kidney disease or diabetes. An IVIG brand without sucrose can minimize this complication. The IVIG with underlying kidney disease or diabetes. An IVIG brand

companies have access to all brands, which give people receiving IVIG the option of getting the best brand for their clinical status, or switching if they react to one. Another benefit of receiving IVIG at home for treatment of pemphigus and pemphigoid is less risk of being exposed to infection that may occur in a hospital or clinic setting. Wound care management in people with blistering diseases must be impeccable, and whenever possible exposure to potential infections must be avoided.

Insurance coverage for pemphigus and pemphigoid can be somewhat difficult, and in some cases, there is no guarantee of coverage. However, these days, more off label uses of IVIG are being accepted as standard when conventional treatments have failed. Therefore, many insurance companies and medical groups are covering IVIG at home for pemphigus and pemphigoid. In fact, until 2006, Medicare only covered IVIG at home for people with immune deficiencies. With the Part D changes, there is now home IVIG coverage for autoimmune disorders, including blistering diseases. Copays and deductibles can exist depending on the health plan, including the Medicare Part D plans. In some instances, the setting for IVIG administration may be influenced by amount of the patient’s copays and deductibles which may be less in one setting compared to the other.

In conclusion, IVIG is a relatively safe and effective steroid-sparing therapy that appears successful in treating pemphigus and pemphigoid in many, but not all cases, where steroids and immunosuppressants given alone have failed. IVIG can be safely administered at home, and many insurance companies have recognized that IVIG is a standard (although not proven) therapy in treating these conditions. All of this gives people with autoimmune blistering disorders hope and additional treatment options.


**MEETING REGISTRATION FORM**

**NAME:** ___________________________________________  **AGE:** __________

**ADDRESS** ____________________________________________  # of Previous Conferences Attended _____________

**CITY** _____________________________________________ **STATE** __________ **POSTAL CODE** __________

**COUNTRY (if other than United States)** ____________________________

**EMAIL** ____________________________________________

- [ ] I require special assistance (please attach a written description for all persons with special needs)

Please list names and ages of other guests who are attending.

<table>
<thead>
<tr>
<th>Name</th>
<th>Age</th>
<th>Special assistance?</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**MEETING REGISTRATION FEES**

**NUMBER OF PEOPLE ATTENDING** _______  x $125.00 = $ __________

**GROWTH CAMPAIGN DONATION** $ __________

**MEETING SCHOLARSHIP DONATION** (help someone in need attend this year’s meeting) $ __________

- [ ] I request scholarship assistance. Please call me immediately and accept this donation to help defray costs.

**GRAND TOTAL** $ __________

---

- [ ] Check
- [ ] Money Order
- [ ] Visa
- [ ] Master Card

Card # __________ - __________ - __________ - __________  Expiration Date: ___ / ___  Postal Code: __________

**Name on Card** ___________________________________________  **Signature** ___________________________________________

This donation is in HONOR / MEMORY of _____________________________

- [ ] Please notify them at the following address:

  **ADDRESS** ____________________________________________

  **CITY** _____________________________________________

  **STATE** ___________________________________________

  **POSTAL CODE** ______________________________________

  **COUNTRY (if other than United States)** __________________

**PAYMENT CAN BE MADE IN YOUR HOME COUNTRY’S CURRENCY AND MUST ACCOMPANY THIS FORM. You can also fax it to us at (916) 922-1458.**

Send form and payment to: IPF, 1540 River Park Dr Ste 208, Sacramento CA 95815

---

**PLEASE INDICATE THE NUMBER OF MEALS YOU WILL WANT FOR THE GALA DINNER**

(This should match the number of people attending identified in the **NUMBER OF PEOPLE ATTENDING** block above)

<table>
<thead>
<tr>
<th>MEAL</th>
<th>QUANTITY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Filet Mignon</td>
<td></td>
</tr>
<tr>
<td>Florida Red Snapper</td>
<td></td>
</tr>
<tr>
<td>Vegetarian Plate</td>
<td></td>
</tr>
</tbody>
</table>

**TOTALS**
IPF & PPS INVITE YOU!

You are invited to join us as we celebrate the unification of two organizations geared towards the pemphigus and pemphigoid communities! The International Pemphigus Foundation and the Pemphigus and Pemphigoid Society have led the way for patient advocacy and advances in patient services.

Mark your calendars, pack your bags, and get ready to head for New York! The IPF and PPS are celebrating in grand New York City style as the Big Apple plays host to our Patient/Doctor Meeting. This year’s event will feature a gala celebration dinner at The Water Club on Saturday evening. This spectacular celebration is nestled between two days of exciting and informative discussions presented by some of the foremost experts in the fields of pemphigus and pemphigoid.

For those arriving Friday night there will be a reception welcoming everyone to the meeting. The meeting itself will be held at the New York University’s College of Dentistry and refreshments will be available during breaks. The meeting will conclude around noon on Sunday allowing attendees plenty of time to return home from a weekend of support, knowledge, and friends. More information is available at www.pemphigus.org.

DONATE YOUR CAR TO THE IPF!

If you live in the United States you can donate your vehicles to the International Pemphigus Foundation! Your tax-free donation helps us continue to provide services and information to patients, their families, and caregivers around the world.

We will complete the initial paperwork for you.

Call or email us today!
(916) 922-1298
pemphigus@pemphigus.org

IPF Vehicle Donation Program
1540 River Park Drive • Suite 208 • Sacramento, CA 95815

Shop Online!
http://www.pemphigus.org/affiliates.html
The IPF now offers a shopper’s affiliation page on our site! Merchants such as Amazon.com, Walmart, GiftBaskets.com, Easy Click Travel and others! We are excited to offer this service to you - services we all use, only now the IPF can receive a portion of your purchase towards our fundraising efforts.