Conference sells out!
Last minute rush for seats overwhelms hotel.
See the photos, read what the experts said.
See pages one and seven

It has been a great year
President Jean Barish reports on the progress made this year.
See page three

Fundraising
There are many ways to give, some have tax advantages and one earns you income.
See page 16

Segall is back from England
This was a historic meeting between the NPF and the PV Network which produced important results.
See page five

Trick or treat?
NPF Treasurer tells of tricks and treats on his road to recovery.
See page four

Cicatricial pemphigoid
Dr. Joyce Rico explains the basics. Check it out.
See page 14

Contents:
2. The view from here
3. President’s message
4. Patient Profile
5. Report from England
6. Medical Observer
7-11. The SF Conference
12. Support group news
14. CP basic information
15. Subscription Form
16. Barish on fundraising

A record crowd of 90 patients and caregivers attended the “Summit in the City” conference in San Francisco on September 23, to hear the experts, socialize with fellow patients and get answers to pressing questions. An overview of the conference is below, reports on what was said start on page seven.

“Summit in the City” draws record attendance

Third conference a real charmer

By Sal Capo
Editor

A big hazy sun was well on its way to burning off the overcast when the first guests of the conference began to arrive at the hotel near Fisherman’s Wharf. The scent of the ocean air mixed with the call of the gulls, and sailboats dotted the bay. It was the kind of a clear, clean, crisp autumn day that makes for great tourist photos and exciting times. As more guests arrived the mood was part seminar, part class reunion and part fiesta! The buzz of conversation and occasional bursts of laughter filled the ballroom.

Two doctors join, one leaves Advisory Board

For the third time this year, Dr. Grant Anhalt, head of the NPF Medical Advisory Board has announced additions to the Board, while one member has decided to leave.

Dr. Joyce Rico will give up her teaching position with New York University and join a pharmaceutical company to head a research department.

“Dr. Rico has been a very valuable source of information and energy,” said Executive Director, Janet Segall. “She has been exceedingly generous to us and she will be missed. We send her our thanks and best wishes in her career.”

Continued on page 14

Officially the Third Annual Doctor/Patient Conference, “Summit in the City,” took place September 23, 2000 at the Holiday Inn at the Wharf in San Francisco. The conference sold out following a flood of last minute requests for tickets. The ballroom filled for the event was filled to capacity with 90 patients and caregivers, plus seven speakers and several staff members and volunteers.

Executive Director Janet Segall put the conference together all but
Continued on page six
We can be the principle organization worldwide

I recently spent two and a half weeks in London. The main reason for my trip was to help England's PV Network raise its profile, and to give me an idea of how we could be effective in becoming a more visible presence internationally.

In England many other diseases are represented by their own charitable foundation, but because of the rarity of pemphigus in countries like England, I came away with the sense that we can indeed be the principle organization with support groups or networks around the world.

Siri Lowe, the PV Network leader, has written a very thorough account of my visit. (See page five) A lot of what we are doing in the States, Siri is doing in England. She is working very hard to bring pemphigus to the foreground in England so that people with the disease and in the medical community are aware of what pemphigus is and also how serious it is. There, as here, diagnosis can take a while due to the fact that the disease is often the last one considered, and drug treatment protocols differ depending on who the physician might be.

The meetings I went to with Siri were very interesting from my perspective. The idea that doctors, patient groups, pharmaceutical companies and government representatives could meet on a regular basis to discuss important issues facing each group was very impressive. Of course, I realize how difficult that might be for us just, due to the size of our country and the differences in how the health care system works, but nonetheless, it was impressive.

I also found our meetings with Prof. Martin Black, England's leading pemphigus expert, and Dr. Fendela Wojnarowska to be very productive. Both physicians were very eager to support the efforts of the PV Network to raise the profile of pemphigus in England. Dr. Black is the only physician in England using the ELISA test. Dr. Black was also supportive of the PV Network's wish to be an adjunct of our Foundation with the NPF looking for more international support.

Prof. Black and Dr. Wojnarowska were very interested in collaborating on a pemphigus registry in England as well. As with the doctors in the U.S., both Prof. Black and Dr. Wojnarowska believe that a registry might help change the image of pemphigus in England. I indicated to both physicians that the Foundation, would support their efforts.

As a group that exists to help patients, I found great satisfaction with the success of the first patient meeting of the PV Network. One of the things that struck me was how similar the needs of individuals living here with pemphigus/CP as they are in England. I met people from all walks of life, all races and all ethnic groups some older, some younger. People came from different parts of England to meet others going through the same things, asking the same questions. With the weather windy and rainy, and the transportation system in turmoil, people came anyway. The most amazing story I heard was two women with PV who came to the meeting together. They had coincidentally met in the same knitting circle. What are the odds of that happening with such a rare disease?

From all this I realize that as rare as this disease may be, there are tens (maybe hundreds) of thousands of people around the world suffering from pemphigus or pemphigoid. The disease, as well as the drugs we all take (no matter where we seem to live), can cause us great pain, worry and fear. The stories I heard in England were as moving as any story I have heard from the U.S., or from anywhere else. We are a community of people from all walks of life living and learning how to deal with a serious, life-threatening, chronic illness.

Continued on page 14
It's been a remarkable year for the NPF

Our base of support has grown, and our financial position has improved

Dear friends,

This has been a remarkable year for the National Pemphigus Foundation. As I look back over the past twelve months I am thrilled at how much we have accomplished. There is so much good news to report it is difficult to know where to begin.

More and more people are learning about and supporting the Foundation, and we have connected with a growing number of people with pemphigus, their families, friends and the general public. This year we identified several hundred newly diagnosed patients. We were able to refer them to the best medical care possible, as well as provide the emotional support so vital when first diagnosed with pemphigus. The response to our support has been wonderfully heartwarming. It is a constant reminder of how important our work is.

We are reaching out to the pemphigus community in many ways. Our Third Annual NPF Meeting in San Francisco, “Summit in the City,” was a great success. It drew almost one hundred people, and provided a forum for the presentation of a great deal of invaluable information about living with and treating pemphigus. It was also a wonderful opportunity to meet old friends and make new ones.

We continue to publish The National Pemphigus Foundation Quarterly, the only newsletter dedicated exclusively to pemphigus. We also continue to provide up-to-the-minute information on our website, as well as links to other websites with valuable information about bullous disease, drugs, autoimmune disease and other subjects of interest to our members. Our online and local support groups are a wonderful source of information and assistance to so many of you.

Our presence outside the United States is growing. We have subscribers to the Quarterly and online support group members from around the world, including Europe, Australia and the Middle East. We also have a network of medical professionals outside the U.S. We are affiliated with an active organization in Canada. And our Founder/Executive Director Janet Segall, has just spent several weeks meeting with people in England to help build affiliated organizations.

We have also been very successful at reaching out to the medical community. Last year we again were represented at the annual meeting of the American Academy of Dermatology. Our table drew a great deal of interest from many medical professionals from around the globe. Additionally, we participated in an annual educational meeting in Washington, D.C., meeting with members of the National Institutes of Health as well as Senators and Congressional representatives on Capitol Hill.

Finally, we continue to support other organizations interested in pemphigus, such as the American Autoimmune Related Disease Association, the Coalition for Patient Advocacy for Skin Disease Research, and the Children’s Skin Disease Foundation.

Our efforts to support research continue. We are working with two pharmaceutical companies, Genzyme Corp. and Alexion Pharmaceuticals, who are applying state-of-the-art research techniques to gain a better understanding of the cause and treatment of pemphigus and pemphigoid. We have also created an NPF research fund to help support the pemphigus research of a young scientist.

The leadership of the Foundation has also grown. We have added a new member to the Board of Directors, NY Supreme Court Justice Janice Taylor.

We have also added two new members to our Medical Advisory Board, Dr. Sergei Grando from the University of California at Davis and Dr. Hosein Nousari, from Johns Hopkins. Finally, our base of support has grown, and our financial position has improved during the year.

In addition to our past accomplishments and successes, we have exciting plans for the future. This April, with the support of the American Academy of Autoimmune Related Diseases, the Foundation is sponsoring a two-day medical conference entitled “Pemphigus as a Model of Humoral Autoimmune Disease.” This meeting, to be held in Bethesda, Maryland at the National Institutes of Health, will bring together scientists and clinicians from all over the world to discuss the causes and treatments of pemphigus/pemphigoid. It is our hope that this meeting of experts will increase their understanding of this disease and bring us closer to the day when there will be a cure.

We are working to improve our educational material. We are increasing our list of physician referrals, in order that anyone with pemphigus will be assured the best possible medical care. A revised NPF Welcome Book should soon be available to help newly diagnosed patients better understand the disease. We are producing a short documentary video that will introduce pemphigus to a larger audience of both lay people and medical professionals. And we are starting to look into ways of supporting public policies that affect people with pemphigus, most notably healthcare issues.

This is a chance to say “Thank You” to everyone who has been so helpful in making this year so successful. Our staff, the Board of Directors, and the Medical Advisory Board have all worked tirelessly and selflessly. Our online support group administrators have put in countless hours making sure everything runs smoothly. The wonderful medical professionals who generously donated their time to make “Summit in the City” such a success deserve special thanks. To our supporters in the business world who have helped, all our thanks. And to so many of you have helped with your financial support, support-group efforts, and emotional support to one another, I want to extend one huge “THANK YOU ONE AND ALL.”

The National Pemphigus Foundation exists to give everyone affected with this illness the tools to live their lives as fully and successfully as possible. Just as important, we are here to give you the hope that is needed to help you deal with your disease. Keeping this hope burning may be the most important thing we can do. At this holiday time of renewal it is my wish that the coming year brings each of you better health and continued success in living with pemphigus. May we all hope for new treatments, ongoing research, and greater success at finding a cure.

Warm regards,
Jean Barish
President
By Lee A. Heins

My story began six years ago on Halloween evening, 1994. I was 41 at the time. My family and I were attending our annual church Harvest Festival. As I walked around the carnival, I enjoyed all of the foods one would expect to eat at such an event. However, this time the popcorn hurt my mouth and the tomato sauce on the pizza stung like acid.

Later that week, I made an appointment with my internist. My throat looked eroded and it was suspected that I had an infection. Over the next two months, we tried several different courses of antibiotics, all without success. By the middle of December, I started losing weight, as I had a difficult time eating due to the pain of the oral lesions.

By New Year's Day, my wife was frustrated by the lack of progress with the internist and quickly enrolled me in a new internal medicine group which had an outstanding reputation. My new primary physician was on vacation so his partner examined me. He said that he “did not know from what I was suffering.” Guessing it to be some type of an autoimmune disorder, he prescribed Prednisone, which he believed would stabilize the condition. In addition, he referred me to an ENT (Ear, nose and throat) Specialist.

Meanwhile, I began to contact numerous other physicians whom I had met through the banking business as clients and friends. In all, I believe that I spoke on the phone or in person with no less than 12 physicians. The consensus seemed to be that I should have a biopsy of the tissue in my mouth. The ENT Specialist performed the biopsy and forwarded it to a lab for analysis. The Medical Director of the lab, sensing that something was unusual, sent over a memo to the dermatology pathologist asking, “This isn’t pemphigus vulgaris is it?” I was fortunate that the biopsy was sent to the best dermatology pathologist in Orange County, California. He quickly replied that it was pemphigus vulgaris, and I finally had a diagnosis after four months!

Over the following months, I started to research the disease and look for answers. One of my clients was a dermatologist with Kaiser Hospital. He called me with the unbelievable news that he had just received a letter (this was in the days before the Internet was widely in use) from Janet Segall at the National Pemphigus Foundation, which was still in its infancy.

I contacted Janet and she was able to answer my questions and give me the support I needed to face the disease. The most exciting news was that she was organizing a support group in Los Angeles.

Later that year, we had our first meeting. I was so excited to meet others who could share my experience. At the first meeting, I was struck by the fact that when the disease is under control, pemphigus patients actually look healthier than the general population!

During a subsequent meeting of the Los Angeles support group, I asked Janet Segall how the Foundation supported itself and how it planned to grow to serve additional patients. Janet, at that time, had no real experience in fund raising, so I volunteered to be the Fund-Raising Chairman.

One of our first projects was to initiate the Annual Holiday Fundraiser in which individual patients approach family and friends during the Thanksgiving to New Year’s Holiday Season to provide support for the Foundation. This has been a very successful program over the past several years, as people are anxious to help friends and associates who are suffering from this serious and unusual disease. The Holiday Fundraiser has allowed the Foundation to provide support to patients who need assistance with the disease.

Two years ago, I was asked to be on the Board of Directors of the National Pemphigus Foundation. Due to my background as a Bank of America loan officer, I was elected as Treasurer of the Foundation.

Over the past six years, I have struggled with osteoporosis as a result of Prednisone use. Several years ago, before Fosamax and Miacalcin nasal inhalant, I sneezed (one of those suppressed sneezes) and instantly compressed my spine four inches. This took many months to heal.

In September 1999, I had a serious pemphigus flare, which was so bad that my local dermatologist resigned from my care, stating that he did not have adequate experience to deal with the disease. I contacted Janet Segall, who in turn contacted Grant Anhalt M.D. at Johns Hopkins.

Dr. Anhalt referred me to David Woodley M.D., who is co-chair of the University of Southern California Department of Dermatology. Dr. Anhalt and Dr. Woodley have been friends for the past 20 years. The Foundation’s referral to Dr. Woodley was worth every minute of my volunteer work over the years.

During my flares, I was hospitalized at the Norris Cancer Hospital, which is USC’s facility authorized to provide chemotherapy treatment. I was being treated with Cytoxan, which is a chemotherapy agent. The “Cytoxan Protocol” included 575 mg a day of I.V. Cytoxan for four days (the dose is half of that given to a cancer patient), 100 mg of I.V. Prednisone, as well as daily plasmapheresis (blood plasma exchange).

Upon release from the hospital, I continued the Cytoxan Protocol on an interval of once every three weeks for four months, and then once a month for four additional months.

Plasmapheresis was done three times a week for two months and then gradually extended to be done two days prior to the monthly Cytoxan protocol. My daily Prednisone dose peaked at 120 mg a day and has been significantly decreased at this point. I continue to recuperate at home, and am hoping to return to work within eight months.

During the long course of this illness, I have been blessed to have the unwavering support of my family, as well as much encouragement from my friends in the Foundation. My goal is to keep myself healthy so I can continue to help the Foundation reach out to others who are in need of information and support.
Advancing the cause of English PV patients

By Siti Lowe
Founder of The PV Network
London, England

So, how was it, this first ever visit of the National Pemphigus Foundation’s Executive Director, Janet Segall, to The Pemphigus Vulgaris Network in England? Well, it was great! Exhausting, very hard work, and very, very successful.

We had a busy programme over two weeks. Meetings with the two main blistering disease dermatologists in this country, the first ever UK meeting for people with pemphigus who’d been in touch with the PV Network, and several meetings of the umbrella groups that we campaign with. It also gave Janet and I a chance to meet and find out about the similarities and differences in our work.

The basic work of letters and phone calls, supporting people with pemphigus, answering questions, helping people get information; all of that is of course exactly the same. Just as in America, we campaign here with other similar patient groups in order to get a better hearing for the small number of people with pemphigus. Although the structures of the political and health systems are very different in our two countries, it became clear that we work in fundamentally similar ways each adapting ourselves to these different structures.

We work with two main umbrella groups here. One is the Skin Care Campaign (SCC), which campaigns about things that affect us as pemphigus patients (e.g. the closure of specialist dermatology wards in hospitals). The other is the Long-Term Medical Conditions Alliance which represents patients living with chronic disease and campaigns on the issues which we have in common (e.g. getting a patient voice within the National Health Service). We are also a member of the Associate Parliamentary Group on Skin (which is linked to the SCC and makes reports to government via parliament).

The SCC links up patient groups representing various skin conditions. It is led by patient groups but also has amongst its members dermatologists, dermatology nurses, GPs, pharmacists, representatives from our Health Service (e.g. this meeting had two people from the Department of Health) and representatives from the pharmaceutical companies.

This was one of the differences that Janet commented on, that there was an umbrella group covering such a wide spread of interested parties, and particularly that members of the Department of Health could be present at a meeting voicing patients’ concerns. If I’d been from a different country and trying to make sense of it all I don’t think I’d have had a clue about the government and health service structures that were referred to but the spread of issues covered gave an idea of the range of SCC’s work. Some issues (like the successful campaign in part of London to keep dermatology services within a hospital setting) are clearly of very great importance to pemphigus patients.

Professor Martin Black, of St. John’s Institute of Dermatology in London, and Dr. Fenella Wojnarowska of the Churchill Hospital in Oxford are the two doctors most interested in pemphigus in this country and support the PV Network. We had a really interesting meeting with Professor Black, and a visit to his laboratory where they diagnose biopsies from all over the country and are currently using ELISA testing (the only place to do so at the moment in England). I was enormously grateful to have Janet with me, she was able to talk to Professor Black about the international work of the NPF and gave him a really good idea of the parameters of that work particularly links with dermatologists worldwide.

Our meeting with Dr. Wojnarowska had to be via a three-way telephone conversation. Again, Janet was able to give the doctor a really good idea of the scope of Foundation’s work. He and I sorted out a future campaign on the two projects that Zoe (the cofounder of the PV Network) and I want to try in the immediate future. One is project the word “remission,” more of that later. The other is project “patient population.”

It was fascinating to find that the Foundation is supporting a Pemphigus Registry in America. The question of how many pemphigus patients are there in the UK is one that we have been trying to look at. We think it’s really important to find out and start some sort of Register here.

So it was terrific to be able to widen this whole issue into an international one. Dr. Wojnarowska will try and spearhead a proper study of patient population in the future. In the meantime we’re hoping to get some provisional figures via the British Association of Dermatologists.

We’re also hoping to start doctors thinking about the way they use the term “remission” when we may still be on drugs and indeed still have some disease activity. Dr. Wojnarowska has kindly offered to write an article. I hope this will get things rolling.

The other big feature of Janet’s visit was the first ever meeting in the UK for people with pemphigus, and mucous membrane pemphigoid (circular pemphigoid) which we cover since there is no support group in this country for people with CP. We’d had to hold it in London which inevitably meant that people in Scotland and the far north of England were unable to get there.

Hopefully some time in the future a PV Network member from the north may be able to host a similar, smaller regional meeting. Anyway, despite the problems of distance, 25 people were able to come with apologies from another 14 who were unable to make it.

We held the meeting in the headquarters of the British Association of Dermatologists, who were enormously supportive of us. So this meant it was in a proper, comfortable lecture theatre with good facilities. It was a really lively meeting which I enjoyed enormously and I’ve had lots of letters and phone calls since telling me how much people appreciated hearing Janet speak and having a chance to swap experiences, ask questions and talk about the many things we have in common as people living with pemphigus. I’m pleased to say that two members have offered to hostess a similar, south of the country regional meeting next year.

So that was what happened during our schedule of meetings, plus, of course, loads of chats and a chance for me to find out lots more information on pemphigus. I came to the conclusion that apart from necessary differences of style and approach we’re more or less doing the same things, with some very important exceptions. We’ve got a registered medical charity (owing to my own health limitations and the smallness of the patient body here), and that would really limit our activity but for the fact that our mother organization, the NPF, is an internationally respected medical charity with a superb medical advisory board. So the things we don’t have the resources to do (i.e. fundraise, support research, international work) are covered by The Foundation.

To sum it all up, I had an exhausting but great time, and Janet’s visit has meant that we’ve been able to raise the profile of The PV Network with doctors and start what will hopefully be a regular feature of patient meetings.
Innovative therapies and immunology

By Edward Tenner M.D.

Because pemphigus is a classic autoimmune disease, many researchers have been investigating the basic mechanisms that cause it, hoping to find treatments not only for pemphigus but for other autoimmune diseases that affect many more people. The meeting next spring at the National Institutes of Health on pemphigus sponsored by the National Pemphigus Foundation and the American Autoimmune Related Disease Association will look into, among other things, current and novel treatments and treatments of the future. I want to highlight two articles that talk about some of the innovations in immunology that may lead to more specific treatments for pemphigus that will be more efficacious and have less side effects than what is presently available.

The first appeared in The New England Journal of Medicine on June 3, 1999 as an editorial by Robert S. Schwartz titled “The New Immunology—The End of Immunosuppressive Drug Therapy?” In it are summarized the amazing advances that have been made in elucidating the basic science mechanisms of our immune systems. It is truly remarkable how the specific chemicals and reactions that lead to activating our immune systems have been determined. In learning how T cells, which are the fundamental cells driving our immune system, are activated in a two signal activation process call costimulation, a mechanism that control and even stop this activation have been learned. One paragraph near the end of the editorial said, “Looking to the future, one can see that the new immunology—harnessing the body’s own mechanisms for regulating the immune system—could be a far better solution to the problem of... transplant...rejection and even to the treatment of autoimmune diseases than nonspecific immunosuppressive drugs.”

Next let’s look at an editorial by John R. Stanley in the British Journal of Dermatology (Feb. 2000; 142:208-209) titled “Understanding the pathophysiology of pemphigus suggests innovative therapeutic approaches.” Dr. Stanley outlines historically the progress that has been made in understanding pemphigus. First clinical and histologic features of pemphigus were studied, distinguishing the major types of pemphigus. Further advances determined that pemphigus is an autoimmune disease. As the autoantibodies have been identified and characterized it has been possible to identify the antigens attacked in the disease, desmoglein 1 in pemphigus foliaceus and desmoglein 3 in pemphigus vulgaris. Studies have shown that interfering with the function of these desmoglein proteins causes desmosomes that hold skin cells together not to work and leads to blister formation.

Using this information Dr. Stanley notes how Dr. Masayuki Amagai and his colleagues have performed experiments that suggest innovative treatments for pemphigus. Knowing the antigen i.e. desmoglein 1 or 3, which the autoantibodies attack, is it possible to clone the antigen in a way that it retains its immunoreactivity. If a patient’s sera is mixed with these cloned antigens, the disease causing autoantibodies attach to the antigen and are removed from the patients blood. Removing the pathologic antibodies should stop their destructive effects on the desmosomes and stop blisters from occurring.

Another futuristic therapy from Dr. Amagai’s group is then discussed based on an article in the same issue of the British Journal of Dermatology (142:321-330). This potential treatment would destroy the B cells that produce autoantibodies against desmoglein that go on to cause pemphigus. In mice part of desmoglein 3 (the antigen) is combined with a toxin. This antigen-toxin combination molecule targets the autoantibody-producing B cells by attaching to receptors on the surface of the B cells. When these combined complexes are brought into the cell the toxin destroys the disease causing B cell. This was shown in individual cells and also in animals. The beauty of this possible treatment is that only B cells that produce pathologic autoantibodies are destroyed. The remainder of the immune system is unaffected.

I share Dr. Stanley’s excitement in the possibilities of these innovative treatments. Being so specific they would avoid many of the side effects of the medications used in treating pemphigus at present. The excitement must be tempered with the realization that getting to the point where they could be used in people may be many years away.
The speakers covered a great deal of ground during the day-long conference in San Francisco. The basics of pemphigus and various ranges of treatment were covered in detail, and new subjects were presented which have not been covered in previous meetings. These included basic oral hygiene and Dr. Sergei Grando's new research using a nonsteroid drug called Mestinon.

Here are some of the highlights of what was said by the doctors in the order in which they spoke.

**Dr. Kari Connolly**

University of California at San Francisco. Professor of Dermatology. Specialties include Scleroderma, fibrosis Lupus and other autoimmune diseases.

The basics of pemphigus was Dr. Connolly’s topic. She explained that pemphigus is “an aberrant immune reaction leading to production of antibodies that target cells in the skin (keratinocytes). This causes the skin to fall apart.”

Pemphigus affects men and women equally, and usually in the fifth or sixth decade of life. In her practice, she has patients from 11 to 90 years of age. Some races are more likely to get pemphigus and she cited Jews and those living around the Mediterranean Sea as having higher incidence. It is not an inherited disease, but there are genetic markers which increase one's chances of developing pemphigus. PV patients outnumber PF patients by about five to one. D-penicillamine (not related to penicillin) and Captopril (a blood pressure medicine) have been shown to cause pemphigus in some people. There is no difference between drug induced pemphigus and the naturally occurring disease. Pemphigus can cause death if not treated.

The frequency of occurrence is not well documented and she urged a study to attain accurate numbers.

When asked about the dangers of a mother giving pemphigus to children during pregnancy, she said, “The short answer is no. Statistically this does not run in families to any great degree. There is very little cause to worry that your children will develop it later in life.

“On the other hand, there is a chance that a baby will be born with blisters for a short period of time. This is caused by antibodies crossing the placenta. This will resolve by itself in six months and does not require treatment of the baby.”

There are only 23 such cases in the medical literature.

“Pemphigus tends to be a very chronic or long lasting disease but there are a wide range of useful and effective treatments.” Treatment requires the patients to take an active part in the recovery plan. “I do think people go into spontaneous remission and you be hopeful about that,” she said.

**Dr. Grant Anhalt**

Acting Head of Dermatology, Johns Hopkins University Medical Center. Vice President of Scientific Affairs for the NPF and Chairman of the Medical Advisory Board.

He is one of the world’s foremost experts on pemphigus. He and his team were the first to identify the sub strain known as paraneoplastic pemphigus in 1990.

Dr. Grant made two presentations. The first discusses the differences between PV and PF. In the lab, PF is differentiated from PV by the Desmogleins. Desmosomes are the sites of attachment between adjacent cells. The transmembrane proteins that form these attachments are called desmogleins. Desmoglein 1 is associated with PF and Desmogleins 1 and 3 are associated with PV. PF has blisters closer to the surface of the skin.

With pemphigus “the first two or three years are the more difficult than in subsequent years. Once in a good durable remission, the next decade should be easier to manage the disease.” he said. Generally, if the initial outbreak of pemphigus is mild, subsequent outbreaks should also be mild. There are exceptions.

PF, known as Fogo Selvagem in Brazil is unique as there are tens of thousands cases in dense clusters. There appears to be environmental triggers. Dr. Grant showed slides taken on his trip to Brazil with Dr. Luis Diaz of the University of North Carolina at Chapel Hill, another NPF medical advisor, who has been studying this variant for decades.

Ninety percent of cases in Brazil occur in rural areas near water. Patients removed from the rural environment improve, but relapse when returned to the original areas.

A tribe of natives is being watched in a remarkable study. These natives were relocated to an area in which Fogo Selvagem is common. They were evaluated medically before they moved and are being visited often in the five years of the study. There is a 3.2% prevalence of FS and there appears to be a very long incubation period. Some patients have pemphigus antibodies up to five years before the first sign of blistering. No medical personnel in contact with FS patients have ever developed the disease.

Paraneoplastic pemphigus is the most problematic of the subsets. It affects the mucous membranes, palms, soles of the feet, lips and some internal organs. It is always associated with one of several uncommon tumors of the immunologic or blood systems. If the tumors are benign, as in the case with Castleman’s Disease (commonly found in younger patients) and if they can be removed entirely, the pemphigus will often resolve itself and chances of survival are increased.

Pemphigoid was differentiated from pemphigus in the 1950s. It commonly involves large, itchy areas, large blisters and hive-like rashes. Treatment for pemphigoid involves blocking the inflammation factors. There are numerous medicines which are ef-
...WHO SAID WHAT
Continued from page seven

ective. Treatment usually includes steroids. Tetracycline and niacinamide may work instead of steroids in mild cases. It is not life-threatening in most cases.

Cicatricial pemphigoid, (CP) affects the mucous membranes, eyes, esophagus, larynx and may lead to blindness and breathing difficulties. It needs to be treated aggressively with steroids. (See related story on page 14.)

Dr. Anhalt’s second presentation was on the treatment of pemphigus.

Some of the problems in treating pemphigus arise from a lack of scientific studies due to the small patient populations which preclude traditional double-blind crossover trials.

Much of the current information about treatment comes from small studies, anecdotal evidence (some of questionable value) and “favorite recipes” of the doctors confronted with the disease.

Pemphigus can be reproduced “with precision” in mice and studies using mice are going on in several places.

Pemphigus can be difficult to treat for a variety of reasons. Over the 18 years he has studied pemphigus, he has developed a set treatment regimen.

First he uses Prednisone alone, one milligram per kilogram of body weight per day, up to 60 mg and no higher. If the patient continues to flare, an immunosuppressant is added. (Only 20% to 30% of patients respond to Prednisone alone.)

Imuran and CellCept are his drugs of choice in this case. He prescribes up to 300 mg per day of Imuran or up to 3000 mg per day of CellCept.

Anhalt tapers patients to five or ten milligrams of Prednisone every other day over nine to 12 months. He puts patients on 1500 mg of calcium per day and 400 to 800 mg of Vitamin D.

He orders bone density scans and watches for osteoporosis. Depending on the patient he may suggest Calcitriol, Fosamax, testosterone replacement or steroid

Continued on next page

Top: “Statistically (pemphigus) does not run in families. There is little reason for concern.” says Dr. Kari Connolly of the University of San Francisco in describing the basics of pemphigus. Bottom: The lively Question and Answer session featured responses from, (L to R) Drs. Sergei Grando, Francesca Losada Nur, Grant Anhalt and Kari Connolly.
replacement.

If that fails to control the outbreak of blisters, Anhalt turns to Cytoxan which is extremely effective. Cytoxan is not for everyone as the side effects can be severe, including adverse affects on fertility.

The fourth step, if all else fails, is plasmapheresis which rapidly removes the antibodies from the blood stream.

Ideally, patients should not go up and down on their doses of Prednisone. This is "harmful because flairs cause pemphigus to harden and become more and more difficult to knock down," he said. He increases the dose of immunosuppressants instead.

Dr. Anhalt believes IVIG therapy loses its beneficial affect over time and is extremely expensive. He believe pemphigus requires daily medication for optimum effect. Photopheresis reduces side effects but does not control antibody production.

Of dubious value in the treatment of pemphigus are gold injections (25% rate of severe side effects), cyclosporine (works better on T cell diseases), methotrexate (an antiinflammatory drug), photopheresis (unknown how it works) and Dapsone, (useful to treat pemphigoid but does not stop antibody production in pemphigus patients).

Pemphigus is considered under control when 90% of blistering are gone. Occasional blisters, such as after eating pizza, are not a serious concern.

Patients often flair after the flu or other virus attacks, because the immune system steps up, but he waits two weeks to see if further treatment is necessary.

Continued on page 10
Dr. Sergei Grando

University of California at Davis. Professor of Dermatology. Among his specialties are biochemistry, molecular and cellular biology and immunology. He began his study of medicine in Kiev and Moscow, Russia. He was certified in California in 1996.

Grando said that he was “fooled” by the simplicity of pemphigus and it was easy to accept the prevalent theories of treatment. He “presents (an) alternative explanation to the existing facts. I believe Prednisone works in pemphigus because of an unknown mechanism to counter the action of antibodies.”

He is searching for a safer drug which will treat pemphigus without the many side effects of Prednisone. His present work revolves around the nonsteroid medication Mestinon (pyridostigmine bromide) which is starting to be used to treat myasthenia gravis (Weakness of the skeletal or voluntary muscles). Traditional treatments include Prednisone, cyclosporin, and azathioprine.

A report in the Archives of Dermatology that pemphigus was improved by cigarette smoking supported his earlier research that nicotine may play a role in the mystery of pemphigus antibodies. Nicotine stimulates the production of adhesion molecules.

(Note: He does not recommend pemphigus patients take up smoking, quite the contrary. Cigarettes contain thousands of chemicals many proven harmful. It is only nicotine he is interested in.)

Antibodies to the nicotinic receptors of
muscle tissue has been shown to be a cause of myasthenia.

In general, when pemphigus antibodies are found in the skin, blisters occur. When Prednisone is used the blisters stop. The binding of antibodies to the cell cause a biochemical process called "receptor mediation." He parts with traditional theories about the mechanism of pemphigus because the they cannot adequately explain why, in pemphigus, the cells detach but the bridges between the cells remain intact. He believes that there are not one or two triggers of pemphigus, but many, which is why there is so much variation from one pemphigus patient to another.

"No two of you in this room have the same disease," he said.

There are many receptors, viewed as locks, involved and each has its specific key. Only some receptors cause pemphigus, and he is looking to identify them. Prednisone works because it blocks all receptors, not just those related to pemphigus.

Recently, his early success with a patient who has both pemphigus foliaceus and myasthenia gravis using Mestinon (without Prednisone), lead to thoughts that he had found a new and more effective treatment. Continuing research has shown that there are more pieces to the puzzle. He has developed the "Multiple Hit Theory" in which the results of the cumulative effect of various antibodies to different targets which concern cell shape and adhesion. The severity of the disease depends on the ratio of these various antibodies in each patient. His research continues and he is presently in drug trials using Mestinon. To date he has six patients in the trial; two have responded, two have not and it is too early to tell about the others.

Mestinon may cause cramps, diarrhea and hypersalivation, (and others), which can be mitigated by taking the medicine before sleep. I comes in a syrup, extended release tablet and injection. This drug is not for use by pregnant women.

If you are in the Sacramento, California area, or can get there, and would like to participate in the study, contact Dr. Grando at 916-734-6057 or fax 916-734-6793. Further information can be found on the NPF website at www.pemphigus.org/k_grando-nonhor.htm.

**Dr. Francesca Lasada-Nur**

University of California at San Francisco, School of Dentistry, Stomatology Clinic. She has been treating pemphigus and pemphigoid patients for 15 years.

"Oral hygiene is critical to treating pemphigus in the mouth," said Dr. Lasada-Nur at the beginning of her talk. The reasons have to do with the delays common in the diagnosis of pemphigus. She points to the rarity of the disease, inexperienced dentists who often see patients

Continued on page 15
Toronto group shares experiences

By Dan Goodwill
Chapter Leader

The Toronto Support group held its latest meeting on November 8 at the North Toronto Community Centre. Each of the attendees reported on their current state of health. As always, there was lively discussion on where each patient was on the bumpy road to improved health.

One of the attendees explained her journey that has taken her from pulsing to plasmapheresis to IVIG. She explained to the group the way each of these treatments work and each of their side effects.

The pulsing is received five times over a ten day period. The IVIG took place over two days. It produced major headaches. None of these aggressive treatments worked particularly well. In addition to these treatments, she continues to take Prednisone and CellCept. Progress is slow but steady.

Another PV patient spoke about the relationship between her allergies and PV. Her PV flares during allergy season. She talked about the consistent problems she is having with swelling and bleeding under her nails. There was much discussion about her continuing to take five to 10 milligrams of Prednisone over many years with little effect. The patient was encouraged to speak to her doctor about her drug regimen to explore the idea of raising the Prednisone dosage and/or adding an immunosuppressant, to bring her illness under control.

One patient discussed his experience with lesions all over his body. He stressed the importance of visiting a “Wound Clinic” to consult with people who are experienced in bandaging. This is an art unto itself and not an area of expertise for many doctors, including dermatologists. The patient explained the importance of using rubber pads such as those used for burn victims. It is most important to use bandages that do no skin to the skin.

The patients talked about the need to get flu shots at this time of the year. One patient told a particularly touching story about his young son, who, upon seeing his dad get emotional, under the influence of Prednisone, would say “you are acting like ‘Pemphigus Man’, not like my dad”. Everyone in the room could relate to the mood swings, the anxiety and paranoia, that so many of us have experienced under the influence of Prednisone.

There was also a discussion on the dangers of using homeopathic treatments. Some may contain a form of natural Prednisone and can add an unwanted boost to the patient’s Prednisone levels. Other homeopathic drugs can work at cross purposes with the traditional pemphigus drugs. Most of the attendees believe that taking traditional pemphigus drugs is still the best method of treating the illness until a vaccine is developed.

Los Angeles group presents dental hygienist

By Carol Goren

The October 15 meeting of our Los Angeles Support Group was called to order at 1:10 PM with 11 people in attendance which included two new members, Barbara Roller, Chairperson, had all members introduce themselves and share their medical history.

Guest speaker, Pamela Cady, member of the American Dental Hygiene Association, made a presentation to the group.

She emphasized that each person should consult with three doctors: periodontist or dentist, dermatologist and an autoimmune specialist.

She stated the importance of oral hygiene and getting your teeth cleaned every three to four months. “Biotene” and “Tom’s of Maine” toothpaste was recommended along with “Glide” floss. Toothbrushes should be ultra soft using only gentle pressure. An electric toothbrush should not be used.

Vitamin B complex and a multi vitamin may help with tongue irritation which is a build up of bacteria and plaque. An anti bacterial tongue gel was recommended to relieve discomfort. Whitening gels and tartar controls should not be used. Before dental work you may want to increase the dose of steroids or take 2 to 3 days before your visit an anti inflammatory medication such as 800 mg of Advil.

Barbara Roller attended the conference in San Francisco and gave a report. The meeting was adjourned at 4:00 PM.
**SUPPORT GROUP NEWS**

**NPF search for logo ends**

The National Pemphigus Foundation is pleased to present the new logo! It was back in 1997 when work on the logo began, but not until late last year was a suitable design was created and agreed upon. (See cover for color version.) The square shape is symbolic of stability and equality. Two undulating bands cross the square to represent layers of skin. Some might notice the two p-shapes, one each for pemphigus and pemphigoid.

The Foundation's colors will be skyblue and black on a white field. Skyblue was chosen because it reflects unity and the worldwide scope of the Foundation. The logo was designed by Sal Capo.

**Delucie decides to stay on in NY**

*Reiki Master addresses group*

By Matt Koenig

Over 20 people attended the latest meeting of the New York support group, held September 21 at New York University in the Orentreich Conference room.

We are happy to announce that Joan DeLucie will stay on as leader of the New York group, with many of her administrative responsibilities to be shared by a committee of supporting members.

The group heard a presentation from Rita Davies, a certified Reiki Master and Transformational Breath Facilitator. She discussed stress and pain management through breath control and Reiki techniques. Reiki is based on ancient Chinese spiritual practices and has become popular in modern times with people who are interested in relaxation methods.

It is truly a “hands-on” approach, in which Ms. Davies applied touch and meditation to improve the mental state of several of our members. While there is no scientific proof behind her methods, many of our test subjects did report that they felt more relaxed. We all enjoyed her presentation!

Although the golf outing was cancelled, due to the slow sign-up, we are trying again with a Sunday Brunch, to be held on December 10 at the Sag Harbor restaurant. The cost is $12 per person. Anyone interested should contact me at 516-825-4594 for more information.

**It's a girl and she's beautiful!**

Isabel Tidly Byrne was born on Saturday, October 21, 2000 at 9:07 PM. She weighed seven pounds, 12 ounces, and was 19.5 inches long. Mother and child are both home and doing well.

“The labor was worth every second, to see her beautiful face and body, and she is quite healthy.” Erica reports, “Isabel is really a blessing to us. Thank you to everyone for your kind thoughts and support.”

Erica has PV and recently underwent experimental treatments with Dr. Anhalt at Johns Hopkins with excellent results.

**The Anhalt Tape**

A video lecture by Dr. Grant Anhalt of Johns Hopkins University on the current views surrounding the basic biology of pemphigus

Dr. Anhalt is Chairman of the Dermatology Department at Johns Hopkins University Medical Center in Baltimore, Maryland. He is NPF Vice President of Scientific Affairs and Chairman of the Medical Advisory Board. He is one of the world's leading researchers into pemphigus. This illustrated material is not available in a similar format anywhere in the world.

- Professionally produced
- Available in VHS format only
- Approximately 60 minutes
- A 100% of proceeds used to cover production costs and generate funds for additional NPF educational materials.

Suggested donation of only $150.00

Copy or cut out this form, print clearly, and return with donation.

The National Pemphigus Foundation
P.O. Box 9606, Berkeley, CA 94709-0606

Please send me ______ copies(s) of Dr. Anhalt's lecture and slide show.

Payment (Do not send cash): [ ] Check [ ] Visa [ ] Master Card

Card no: _______ _______ _______ _______

Expiration date: MO: ______ YR: ______ Amount enclosed: ______

Signature: __________________________ Print name: __________________________

Address: __________________________

City: __________________________ State/Province: __________________________

Country: __________________________ Postal code: __________________________ Date: __________________________

*U.S. FUNDS ONLY PLEASE. THIS PRICE INCLUDES POSTAGE AND HANDLING. IF THIS AMOUNT IS A HARDSHIP, PLEASE CONTACT THE FOUNDATION.
Two join, one leaves MAB

From page one

Joining the Medical Advisory Board are Dr. Sergei Grando from the University of California at Davis, and Dr. Hossein Nousari from Johns Hopkins University.

Dr. Grando is a professor of Dermatology and long-time friend of the Foundation, hosting several support group meetings and making himself available for questions and consultations.

...We can be principal organization

From page two

The Foundation is committed to helping anyone it can from anywhere in the world. After my trip to England, I encourage anyone reading this in a community with others living with pemphigus/pemphigoid, to form a group (whether it is 5 people or 100 people). The empowerment we get from each other is so positive and meaningful. I saw how excited people were to meet others who knew what they knew and felt what they felt. For many of us the course of our lives changed with our diagnosis. Talking with others who know much of what we feel can hopefully help us learn to live with pemphigus/pemphigoid as opposed to suffer from it.

CP has numerous manifestations

By Dr. Joyce Rico

Cicatrical pemphigoid is an autoimmune blistering disease that predominantly effects mucous membranes, including the mouth and throat, eyes, and genitalia. Patients with skin involvement present with tense blisters or erosion, often on the head and neck, or at sites of trauma. Blisters heal with scarring, which can lead to vision loss or blindness, hoarseness or airway obstruction.

Patients with other autoimmune blistering diseases may also have mucosal involvement. There are wide differences in clinical manifestation of this disease, with some patients presenting with eye disease and others presenting with nose, mouth and throat involvement. Most patients with CP have onset in the seventh decade. CP has been rarely diagnosed in children under age 10.

CP is an autoimmune blistering disease associated with autotropin antibodies directed against normal skin antigens present at the junction of epidermis and dermis. The two major antigens associated with CP are collagen XVII and epiligrin (laminin 5).

There is no clinical differences between patients with anti-collagen XVII and anti-epiligrin reactivity. Determination of the specific antigen that a patient is reacting against is not readily available, except in specialized research laboratories, but will not change the patient's treatment.

Cicatrical pemphigoid has been treated with many different therapies including topical agents, intranasal steroids, antiinflammatory agents such as Dapsone and Prednisone, and immunosuppressants such as azathioprine (Imuran), cyclophosphamide (Cytoxan), cyclosporin (Neoral), mycophenolate mofetil (CellCept). Intravenous immune globulin (IVG) has been used for patients with eye disease. Treatment of this condition requires a coordinated approach by the patients primary care physician, and specialists such as ophthalmologists, gynecologists, otolaryngologists (ENT), and dermatologists.
first (since pemphigus routinely affects the mouth with skin involvement coming later) and doctors who "treat patients without knowing exactly what they are treating."

She says that after two weeks a skin biopsy should be automatic, but many dentists and doctors fail to respond appropriately. This allows pemphigus time to develop, and requires more treatment to control the disease. This problem is compounded by the use of improper medications as doctors treat the wrong disease.

She says that pemphigus has "messy" lesions while pemphigoid has "neat" ones.

A major concern is plaque buildup caused by poor brushing due to blisters, bleeding and pain. Dentists are not as thorough as they should be when cleaning a patient's teeth because they do not want to risk hurting the patient, but this can make pemphigus harder to control in the mouth.

Losada-Nur "treats the patient, not just the disease." She tailors her treatments to each patient by taking extensive medical and drug histories. She places her patients on Prednisone and azathioprine (Imuran), (and lately Cellcept) immediately, and prescribes topical steroids with which she has had great success. In fact, if a patient's flairs are confined to the mouth she prefers to use topical steroids rather than start or increase Prednisone. Dental trays are very useful to put the medicine where it will be most effective. This allows her to treat the mouth while the dose of Prednisone is being reduced. Pemphigoid patients usually have a harder time with trays as it causes the gums to peel, so she uses systemic steroids in these cases.

When a patient needs dental surgery, she gives them 20 mg of Prednisone the day before, the day of, and the day after as a precautionary measure, unless the patient is already on a high dose. A temporary dose of 20 mgs does not require tapering. If flaring, she prefers patients postpone surgery if possible.

Losada-Nur puts her patients on a salt-free, low carbohydrate diet and recommends smaller, more frequent meals to control appetite. She is a firm believer in healthy nutrition, physical exercise and yoga to make the body stronger to better deal with any disease. She said that "patients who are overweight suffer side effects more often and sooner." She recommends a weight loss program.

Biotene tooth brushes, the softest available, are recommended and Biotene mouthwash that does not contain alcohol which burns. A weak solution of (3%) of hydrogen peroxide in Luke warm water and a pinch of salt is also an excellent, and inexpensive, mouthwash.

Immediately upon diagnosis she sends her patients to a dental hygienist for a thorough cleaning of the teeth, unless the mouth is in very bad shape in which case she waits a few weeks until the mouth improves. She suggests getting your teeth cleaned every three or four months to control plaque and avoid complications. Patients should brush immediately after meals or snacks to help stop plaque from forming.

CLARA FELIX
Freelance nutritional journalist

Felix contends that modern man has gotten away from the "evolutionary diets" which allowed humans to thrive as a species.

The result is a long list of common ailments including allergies and depression. Over the past 70 to 100 years humans have begun to eat more protein without balancing the diet with other essential ingredients including sea food and vegetables.

Primarily she is concerned with Omega-3 fatty acids which she say should be consumed in equal amounts of Omega-6 fatty acids.

High protein diets make the body acidic which is problematic.
You make it all happen, day after day

By Jean B Barish
NPFF President
The record level of contributions last year fueled record achievements by the NPFF this year. (See “President’s message” on page three). We reached more doctors, more caregivers and more patients, including a growing number of children, than ever before and provided them with a wider range of services last year.

For that we want to thank those who have given and urge them, and everyone who has yet to contribute, to help us continue to grow and achieve our mission to ease the suffering of those afflicted with pemphigus and pemphigoid.

Private and corporate donations are the life blood of our organization. We hope to not only continue the services we provide now, but to expand our programs to be even more effective and beneficial.

Our “wish-list” includes increasing our patient outreach and support programs both nationally and internationally; expanding our ability to refer patients to qualified physicians; assisting our current list of local support groups and expanding into other cities; to increase the number and quality of our educational publications for patients and physicians; to increase our exposure to the medical community; and, of course, to support research into a cure for these dreadful diseases.

The Foundation would also like to have the resources to participate more actively with patient advocacy coalitions and other organizations that deal with health care and insurance issues that are vitally important to our members. With your support, these wishes will be made realities.

There are countless ways to contribute, one of which will surely fit into your budget and financial planning. Here are a few suggestions:

1. Respond to our fundraising letter campaign currently in progress. This is the easiest. Simply write a check or make a contribution on your credit card.

And while you’re at it, ask your family, friends and coworkers to help as well. Any contribution, regardless of size, will join with the contributions of others to insure our good work continues.

2. Check with your employer to see if they have a matching gift program or some other mechanism for charitable donations. This is a great way to multiply your donation and make others aware of our cause.

3. Check with your financial planner or tax advisor and evaluate the various ways to contribute and, at the same time, reduce your taxes. Furthermore, in some cases, such as the Charitable Gift Annuity, there is a way to earn income for yourself at the same time.

Two investment companies which offer such funds are Charles Schwab at 1-800-746-6216, or www.schwabcharitable.org, and Fidelity Investments at 1-800-682-4438 or www.charitablegift.org.

The Foundation is prepared to accept gifts of stocks, bonds and other securities which is yet another good way to reduce your taxable income.

4. Another easy way to give to charity is through your Will or Living Trust. This is perhaps the most common and productive way to donate and it has tax advantages also.

The list goes on. In short, there are no limits on ways to provide contributions. Give in honor of a patient or caregiver; give because you have, yourself, been helped by the Foundation; give because the needs are so great; or give because it simply feels good to help the less fortunate.

Before 1994 there was no Foundation, only a black-hole of need. There was no way to reach patients and share crucial information on treatments or tips to make living with pemphigus and pemphigoid less maddening. There was no one to educate the medical community or work with other patient advocacy groups to advance the cause of our patients. Before 1994 there wasn’t anyone to talk to about pemphigus at all.

Won’t you help us continue to reach doctors, legislators, the public, and all those patients around the world who, except for the Foundation, would have nowhere to turn for education and support?

Wouldn’t you enjoy the feeling of knowing that every dollar you give goes directly to reduce ignorance about pemphigus and pemphigoid and lessen the suffering our patients endure? Please, act now. There is so much we have done, and so much more to do.

We thank you in advance for your contributions. They mean so much to so many.

More information at www.pemphigus.org