New perspectives on the cause of blister formation

Grando seeks volunteers to test nonhormonal substitute for Prednisone

By Sergei A. Grando, M.D., Ph.D., D.Sci.
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The goal of my research is to develop a safer and more rational treatment for pemphigus. I am deeply concerned that we, as physicians caring for patients with pemphigus, have to accept the risk of severe side effects related to the use of long-term, high dose corticosteroid therapy.

Despite recent progress in developing nonhormonal therapy for other autoimmune conditions, the treatment of pemphigus remains largely dependent on corticosteroid hormones. The lack of progress in developing new therapies for pemphigus is ironic because we thought we understood the basic mechanisms responsible for the development of this disease. But, perhaps our understanding was wrong and possibly this misunderstanding has hampered advancement in treatment.

Traditionally, dermatologic research workers have held that pemphigus occurs when one's own antibodies attack and disrupt the desmosomal attachment points of epidermal keratinocytes—the cells comprising the superficial layer of the skin wherein the blisters emerge. It is further believed that these autoantibodies specifically bind to, and block the function of, the desmosomal adhesion molecules of the desmoglein family. Then, when these desmosomal attachment points fail, keratinocytes separate allowing interstitial fluid to infiltrate into the space around them. Clinically this is reflected by the formation of fragile blisters which quickly break down leaving the red, raw erosions that we recognize as the typical lesions of pemphigus.

Corticosteroids, such as Prednisone, are thought to control pemphigus by inhibiting the ability of lymphocytes to produce the anti-desmoglein autoantibodies. Although this is the currently accepted view of pemphigus and its treatment, my research leads me to conclude that these traditional beliefs are too simplistic and that they may even be erroneous.

Lesson one: Antibodies to desmoglein 3 may not cause pemphigus vulgaris

We have primarily studied the variety of pemphigus known as pemphigus vulgaris. In this disease the antibodies specifically and directly responsible for the

Segall named to CPA-SDR committee

NPF President Janet Segall was named to a special committee during the annual telephone conference call of the Coalition for Patient Advocacy for Skin Disease Research (CPA-SDR). The call was held November 17, 1998.

"With members spread across the country, this is the best way to meet and coordinate our efforts," reported Segall. "I was named to a committee which will bring to Dr. Steven Katz, CPA-SDR Director, the NPF's concerns about the small amounts of money allocated to skin disease research and the kinds of research that are funded.

Dangerous trends

First, I want to thank everyone involved in this year’s holiday fund-raiser. Yet another record amount was gathered and the Foundation will be able to expand its program a little more this next year. The final sum, along with the donors list, will be published in the next issue. I hope everyone had a wonderful holiday season, and may this year be a good one for you. Now, I’d like to discuss one of the most important issues each of us must face sooner or later: health care.

Improved health care for those living with pemphigus is one of the Foundation’s most important goals and one which is an active and ongoing struggle. Presently there are two trends we find disturbing; the continuing reduction of basic health care, and the reallocation of research money away from the academic community.

There are some of us who have very good jobs and very good health care, but many others do not. In the first what is needed is provided when and where needed, no questions asked, and that’s as it should be. But a growing number of Americans are noticing that health care providers are increasing the number of obstacles and restrictions, sometimes significantly.

The movement toward HMOs has caused more and more services to be restricted, or denied altogether. Services and specialists which pemphigus patients need are delayed, sometimes for weeks, while paperwork is processed and approval determined. We all know what pemphigus can do in a week without the proper treatment or drugs.

In some cases some classes of drugs, especially the newer drugs, are not covered by the HMO, which leaves the patient to incur the expense or go without the most effective medicines. One researcher told us that a vaccine for pemphigus may cost $10,000 per year or more. Will your insurance cover this or will you?

This is especially true of Medicare patients with supplemental insurance packages, who are finding alarming changes in coverage and in many places, coverage is refused altogether. In Florida, a half dozen insurance companies are being investigated for anti-trust violations after dumping 50,000 seniors from their roles and other states are experiencing similar troubles.

In the publication Strategic Medicine, Diane Archer, executive director of the Medicare Rights Center says “Even if (seniors and disabled beneficiaries) are able to get supplemental insurance to fill gaps in Medicare, many beneficiaries will experience serious financial difficulty and an inability to get their need for prescription drugs satisfied.” Switching to another provider may only postpone the inevitable.

This trend in HMOs also has the insidious side-effect of drastically reducing the number of dermatology specialists for any but the most common diseases, because time-intensive (read: money losing) services are reimbursed at a lower rate than for simpler, “in-and-out” services and the so-called “big ticket” items such as acne surgeries.

Pemphigus must be treated by knowledgeable doctors with the proper drugs in a timely manner but unfortunately, this is not the situation in a growing number of cases.

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...new perspectives on the cause and treatment of pemphigus

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development of blisters are currently believed to be directed against the desmosomal adhesion molecule, desmoglein 3. However, I now believe that antibodies to desmoglein 3 may be only incidental and that antibodies directed to other antigens may be more directly responsible for causing the disease. Several lines of evidence, both from our laboratory and from other laboratories, support this belief.

First, desmoglein 3 is localized solely to the desmosomal portion of the keratinocyte cell membrane [1], whereas in electron microscopic and immunofluorescent studies, the antibodies present in patients with pemphigus vulgaris bind to many other sites on the keratinocyte cell surface [2, 3]. Second, pemphigus vulgaris antibodies bind not only to a 130 kD protein (the molecular weight of desmoglein 3) but also to proteins of various other weights [4-12]. Third, in the laboratory, antibodies which bind to a non-desmoglein protein of 66 kD, can cause pemphigus-like blisters when they are injected into mice [9].

Fourth, antibodies to desmoglein 3 are found in many relatives of patients with pemphigus but the presence of these antibodies is not associated with the development of blisters in these relatives [13]. Fifth, neonatal mice injected with antibodies specifically directed to desmoglein 3 do not develop pemphigus-like lesions as one would expect if these antibodies were the actual cause of the disease [14]. Sixth, mice which are bred to have defective [15], or absent [16, 17], desmoglein 3 do not spontaneously develop pemphigus-like lesions on the skin as would be expected if this adhesion molecule were solely responsible for the cause of pemphigus vulgaris.

In addition, in my laboratory, we have developed collateral evidence which argues against a causative role for antibodies to desmoglein 3 in the development of pemphigus [18]. Specifically, we are able to create pemphigus-like blisters in mice lacking desmoglein 3 ("desmoglein 3 knockout mice") when we inject them with serum from patients with pemphigus vulgaris. Since these mice possessed no desmoglein 3, some other antigen(s) must have been the target of the disease-causing pemphigus antibodies. It could be argued that the antibodies from these patients cross reacted with desmoglein 1 (the antigen said to be responsible for pemphigus foliaceus) and that this cross reaction caused the blisters but we tested for the presence of anti-desmoglein 1 activity in the serum used in these experiments and found none.

Taken altogether, these data indicate that antibodies to a protein other than desmoglein 3 are responsible for the development of blisters in patients with pemphigus vulgaris.

Lesson two: identification of disease-causing antibodies to antigens other than desmoglein 1 and 3 in the serum of patients with pemphigus vulgaris.

Recently, our laboratory has identified a keratinocyte-derived, non-desmoglein, protein that can absorb the disease-causing antibody from the serum of patients with pemphigus vulgaris [19]. The potential to create blisters in mice can be restored by adding this antibody back to the serum. Both normal and desmoglein 3 knockout mice develop blisters. Since desmoglein 3 knockout mice were bred have no desmoglein 3, and since we documented that there was no anti-desmoglein 1 antibody in the serum, we conclude that the blister causing antibody is not directed toward a desmoglein protein.

However, this non-desmoglein antibody, when administered alone, is not sufficient to cause blisters in mice. Thus we believe that there are "primary" and "secondary" antibodies which must act cooperatively to cause pemphigus. The primary antibody initiates the pathological process leading to blisters. The secondary antibody is produced by the body in order to get rid of the cell debris left after assault by the disease-causing primary antibody.

We then attempted to observe under the microscope how this non-desmoglein, primary antibody initiates a process of keratinocyte separation from one another and rounding up which is termed "acantholysis" and is unique to pemphigus. When we placed this non-desmoglein antibody with cultured keratinocytes we noted that the keratinocytes began to shed desmosomes and associated small portions of plasma membrane from their surface. This suggests to us that binding of this antibody to the cell surface alters the stability of the keratinocyte plasma membrane, which then breaks up releasing desmosomes into the intercellular space. From these observations we have developed a new hypothesis regarding the pathophysiology of blister formation in pemphigus vulgaris.

Hypothesis one: Blister formation in pemphigus occurs in two phases. First, disease causing, primary, antibodies bind to non-desmoglein antigens. This antibody binding causes injury to the cell membrane and subsequent shedding of desmosomes into the intercellular space. Second, during the next phase, these detached desmosomes expose desmoglein antigens. These antigens then lead to the formation of secondary, anti-desmoglein antibodies. Thus while desmoglein antibodies are not primarily responsible for the blister formation, they may play a role in the scavenging of the defective, shed desmosomes.

Based on this hypothesis, the treatment of pemphigus will be efficient if it can: 1) stop production of primary antibodies; or 2) protect keratinocytes from the deleterious effects of primary antibody binding. The first goal can be achieved by developing "anti-pemphigus vaccine" using as immunogen the keratinocyte protein targeted by primary antibody. To achieve the second goal, one need first to elucidate the mechanism of acantholysis.

Lesson three: Corticosteroid therapy improves pemphigus by a novel pathway.

As indicated above, conventional wisdom suggests that corticosteroids such as prednisone improve Pemphigus by suppressing lymphocyte production of disease-causing antibodies. However, in 1983, Swanson and Dahl first demonstrated that methylprednisolone (the activated form of Prednisone) added to skin cultures prevented acantholysis that would otherwise occur when Continued on page 12
Houston

By Richard M. Schwartz

On November 19, 1998, the Houston support group was fortunate to host Dr. Robert Jordon at its second meeting. Dr. Jordon is not only a well respected dermatologist specializing in the research and treatment of pemphigus, but is also a new member of the Medical Advisory Board for the NPF. We were pleased he agreed to be our guest speaker.

Thirteen people attended the meeting, including those of us with pemphigus and our relatives. After a brief introduction, during which Dr. Jordon talked of his long professional involvement with pemphigus, and the fact that his father had also done pemphigus research, Dr. Jordon spent the next hour and a half answering questions on subjects from diet to current research, treatment modalities, etc.

After Dr. Jordon left, we held a brief meeting during which it was decided that our first task should be to raise money for the National Organization. We’ll meet again to discuss ways of doing that.

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New York

By Joan DeLucie

September-On September 10, 1998, 22 members and guests attended the New York support group meeting at New York University Medical Center. The meeting began with the introduction of new members Melissa Gluck, Glenn Kressner and Esther Laks. The group then shared personal experiences and challenges, as well as individual treatment regimens for pemphigus, vulgaris, pemphigus foliaceus and pemphigoid.

An overview of the Chicago trip was presented by Dr. Joyce Rico, Joan DeLucie, and Eli Ben-Dor. Dr. Rico summarized the research project currently being conducted by Pepticum, Inc. All agreed that the most rewarding part of the trip was meeting Foundation members and the physicians that continue to support us.

DeLucie continued by discussing preliminary plans for the Foundation’s second annual meeting to be held in New York City the first weekend in August. She thanked Miriam Weiss, Marcia Pepper, Matthew Koening and Peg Schreder for volunteering to help with the project.

The meeting continued with Marcia Pepper discussing a fund raiser given by her company through an alliance with a coat manufacturer. In telling the success story, it is Marcia’s hope that this concept can be turned into a profitable fund raiser for the NY group.

The group discussed establishing a refreshment committee and Alex Segoura graciously accepted the leadership responsibility. In addition, the group expressed interest in purchasing the Baltimore tape, which Joan will look into and report on at the November meeting.

November-On November 5, 1998, 18 members and guests attended the New York support group meeting at New York University Medical Center.

Our guest speaker was Mitchell R. Anderson, Regional Director of Agencies, Presidential Life Insurance Company. Presidential is a 31 years old New York based life insurance company with estimated assets of $2.4 billion. Its mainstream focus is in the life and fixed annuity marketplace. The Presidential Guaranteed Issued Life contract has been helping people all over the country for over 15 years. Presidential is an industry leader in creating products for special market niches such as ours.

Mr. Anderson discussed the benefits of the Presidential Guaranteed Issue Graded Benefit Life Plan. He informed us that the Presidential Life Plan is a true guaranteed issue with no health questions or underwriting. The issue age is 40 to 80 in most states and up to $25,000 face amount except in New Jersey where it is up to $15,000. Mr. Anderson further explained the death benefit from other than accidental causes. For more information about Presidential Life’s Guaranteed Issue Graded Benefit Life, please contact Mitchell Anderson at 69 Lydecker Street, Nyack New York 10960, or Lou Schwimmer, President, Louis Universal Ltd., 170 Broadway, Suite 609, New York, New York 10038.

I want to thank Louis Schwimmer, Mr. Anderson’s partner and a member of the NY support group, for generously offering to donate his commission to the NPF for each member who purchases a life insurance policy.

I also want to thank Janet Segall for attending the our support group meeting. Janet updated us on Foundation activities and discussed the necessity of funding raising to keep the Foundation viable.

Our next meeting is scheduled for Thursday, January 21, 1999.

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San Francisco

The San Francisco chapter has tentatively scheduled it’s next meeting in Sacramento in April. Notices will be mailed out directly once details are finalized.

Contact: The Foundation
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Toronto

By Dan Goodwill

October-The Toronto support group met on Sunday, October 18 at the North Toronto Memorial Community Centre. There were 12 people in attendance. The group welcomed three new members, Zaibeen Ismail, Mayuri (May) Patel and Rose Bradshaw. Each member of the group discussed their current state of health. The fact that May’s pemphigus has gone into full remission was a morale booster for the group.
The group discussed the types of guest speakers that they would-like to see at the coming meetings. Harriet McCabe agreed to invite a fitness instructor to discuss exercise programs. In view of the possible link between stress and PV, the group agreed to invite a person with expertise in stress management and relaxation techniques to a future meeting. There was also agreement to invite an endocrinologist who can address the subject of hormones and drugs later in the year.

Dan Goodwill agreed to present a budget at the next meeting. Dan and Brian Shell will meet to develop a draft program on fundraising for the group.

**January:** The Toronto support group held a local meeting on January 10 at the North Toronto Memorial Community Centre. The meeting consisted of two components. During the first part of the meeting the group reassessed the role of the support group and whether it was meeting the needs of the group. Those people who attended the meeting on a cold, snowy winter day, agreed that there was value in having an ongoing forum for discussion, and in trying to help each other during this difficult period in our lives.

The group also agreed that meeting every six weeks was too frequent. Starting after the next meeting, we agreed to meet every two months. One of the attendees agreed to ask a fitness instructor to attend the March meeting to offer their thoughts on PV and fitness.

During the second half of the meeting, each of the attendees discussed their current state of health, and in particular, the impact that the drugs have on our lives. The daily regimen of Fosomax, Prednisone, calcium, vitamin pills, CellCept or Imuran, along with our own food and beverage restrictions, are a constant preoccupation.

In view of the side effects (e.g. bone fractures, gum erosions etc.) that some of us have experienced from a prolonged use of Prednisone, there is much wisdom in trying to keep the dose of this drug as low as possible, and to try to come off this drug as quickly as possible. However, for many of us it is quite difficult. The use of immunosuppressive drugs should be considered if there is difficulty in reducing one’s dose of Prednisone.

**Midwest**

**By Arlene Strauss**

The Midwest Chapter is still promoting the Foundation’s “Awareness Program.” We have t-shirts, bumper stickers and buttons available. This is terrific exposure to create public awareness of the disease and the Foundation. Your participation will help our special cause. Anyone interested in participating please see the order form on page 13.

**Contact:**
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**Baltimore**

The Baltimore Area Chapter is seeking members to expand its ranks. Those in the area should contact Erica Byrne.

**Contact:**
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Phone: 410-964-1099
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**Philadelphia**

Contact Barbara Sipe for information concerning the next Philadelphia area support group meeting.

**The NPF Website**

Sal Cappo, Webmaster

The NPF website contains a large selection of articles from previous issues of the newsletter on a range of topics. Other articles are added when available between issues, so check the website often.

Also offered is a large selection of links to sites of interest to those living with pemphigus and care-givers.

It is located at www.pemphigus.org. Please let us know how we might improve our site.

**Amazon.com**

Satisfy your book, video and music needs by taking a trip to the largest selection online and help the Foundation earn money. The NPF earns 15% of the price of products listed on our website or 5% of books purchased at Amazon.com’s website (proving you enter via the NPF website). Just click on the Amazon.com icon on the Foundation’s home page.

**Online discussion**

Sandra Frank & Sal Cappo, Managers

The National Pemphigus Foundation Online newsgroup is open to everyone interested in pemphigus.

To join the list send a blank message to discussion@pemphigus.org. No other information is required. Your email address will then be added automatically.
Diagnostic patterns in pemphigus vulgaris

By Dominik Ettlin, MD, DMD
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As most readers know, the National Pemphigus Foundation held its first annual meeting in Chicago on August 1, 1998. Attendees of the meeting were given a questionnaire. The purpose of this survey was to shed light on the diagnostic process of each participant, and to analyze the collected data for possible trends. The questionnaire was also posted on the Foundation’s website for a few weeks. The collaboration between patients and clinicians is critical to gain more insight into the clinical and basic research aspects of the disease. I would like to take this opportunity to thank everyone who took the time to answer the questionnaire!

Pemphigus vulgaris is an autoimmune disorder that typically presents with sores in the mouth (oral), on the skin, or both. In clinical practice, dental and medical professionals are often challenged by the ill-defined and variable appearance of sores that present in the mouth. We were interested to answer several questions retrospectively:

1) Where does pemphigus vulgaris start: on the skin or in the mouth (oral sores)?

2) When pemphigus vulgaris starts on the skin: what is the time interval between the onset of sores and the final diagnosis? How many clinicians were consulted before the diagnosis of pemphigus vulgaris has been established?

3) Similarly, when pemphigus vulgaris starts in the mouth: what is the time interval between the onset of sores and the final diagnosis? How many clinicians (dentists or physicians) were consulted before the diagnosis of pemphigus vulgaris has been established? Thirty-seven individuals responded to the questionnaire. The analysis of the data revealed some interesting results.

Answer to question 1-In 30 individuals (81%), the first presentations of pemphigus vulgaris were mouth (oral) sores. Only 7 people or 19% experienced their first sores outside of the mouth.

The observation that the disease starts in the mouth in the majority of persons confirms other reports in the medical and dental literature.

Answer to question 2-(See illustration lower left) Of the 30 patients with mouth sores as the earliest disease manifestation, the time interval to diagnosis was less than 6 months for 17 patients (57%), 6-12 months for 4 patients (13%), and greater than 1 year for 9 patients (30%). By comparison, the diagnosis was established within six months in all patients (100%) experiencing skin lesions first. In other words, the proper diagnosis for the same disorder had taken different amounts of time, depending if the disease started with mouth sores or skin sores, respectively.

Answer to question 3-(Illustration above) When the skin was the first manifestation of pemphigus vulgaris, most patients were diagnosed by one or two clinicians. This was very different for patients who experienced only mouth sores. 21 people, i.e. the majority (70%), had to consult with three or more clinicians before the early mouth sores were properly diagnosed as pemphigus vulgaris.

In summary, pemphigus vulgaris invariably affects oral tissue at some time. This study confirms previously published data that the disease typically starts in the mouth. In this sample, the diagnosis was considerably delayed in patients presenting with initial oral involvement, as compared to patients who experienced skin lesions first. More clinicians were involved before oral sores were properly diagnosed as opposed to skin lesions. Hence, health professionals should consider pemphigus vulgaris as a possible cause for mouth sores. If a physician or a dentist is unfamiliar with oral diseases, they should consider prompt and proper referral when encountering oral sores. There is a need to enhance the knowledge of clinicians regarding the oral manifestations of systemic diseases. As a further step in this direction, the data of this study will be presented at the next meeting of the International Association of Dental Research on March 11, 1999 in Vancouver, Canada.
By Dan Goodwill

My first symptoms of PV appeared in August of 1995. After visiting five physicians, I received a definitive diagnosis in December of that year and began taking Prednisone in January of 1996. Within a year, I began to see a noticeable deterioration in my gums. To be specific, in some areas, the outer layers of skin tissue at the gumline disappeared. The thin outer layer of skin tissue became pink. Even with gentle brushing and flossing, I would experience bleeding.

Initially my dentist consulted with my dermatologist, Dr. Daniel Sauder at Sunnybrook Health Science Centre in Toronto. My dentist suggested that I get my teeth cleaned twice a year and take a mouthwash called "Peridex" to prevent infection. This had no impact. From my participation in the pemphigus newsgroup, I learned that other people had similar problems.

Some people suggested that I purchase a topical steroid and apply it with my finger to the affected areas. I consulted my dermatologist, received a prescription and began to follow this routine. This also produced minimal results.

About a year ago I lost one of my fillings. This caused me to go to another dentist, Dr. Paul Belzycki who is a periodontist and does restorative surgery. He took one look in my mouth and was horrified. He had never treated a pemphigus patient, but he knew that I had a major problem that, if left untreated, could result in infection and the possible loss of my teeth.

He then consulted with my dermatologist, and with Dr. Jim Main, his former Professor of Dentistry at the University of Toronto, and one of Toronto's leading specialists. Together they came up with the idea of preparing a set of trays. These are plastic moulds that fit snugly over my gums and teeth. This would allow me to apply the topical steroid to the affected areas and then place the trays over the gums so that it would keep the topical steroid in place.

This worked reasonably well and I saw some significant improvement. Once my gums began to look better, I stopped the treatment. The result: Within a couple of weeks, my eroding gums were back.

At this point I asked Dr. Belzycki to consult with Dr. Dominik Ettlin, who is an Assistant Professor, and Director, Division of Oral Medicine/Oral Diagnosis at Northwestern University, and who has considerable knowledge in treating pemphigus patients. I met Dr. Ettlin at the first national conference of the Pemphigus Foundation in Chicago in August. From this conference call I learned the following:

First, to be truly effective, I must dry the affected areas with a tissue before applying the topical steroid. Second, I must follow a consistent pattern during and after my treatment. I must use the trays three times a day, seven days a week. Once the skin tissue on my gums has returned to more normal levels, I can then revert to using the trays three times a day, three days a week. In other words, I must continue this procedure indefinitely and consistently, or risk a relapse. I am now, finally, seeing a significant improvement. While I have a way to go before my gums look the way they did prior to the onset of PV, I am very happy with the progress I am making.

Here are a few other tips I have picked up along the way. Make sure the trays fit very snugly. If they don't come out properly the first time, have them done again. Since my gums have been so fragile, brushing my teeth has been an adventure. To keep your teeth and gums in good shape, use an extra soft toothbrush and use "dental yarn" instead of dental floss. It's much more gentle.

Also, have your teeth cleaned every six to eight weeks. This will prevent the buildup of tar- tar along the gumline and reduce the chances of infection. It also allows the skin tissue at the gumline to affix itself to your teeth. Ultraviolet works better than Kenalog in Orabase to treat this type of problem.

The rehabilitation of my gums is still a work in progress. It has been a constant learning experience, with input from four dentists, my dermatologist, and many people on the online pemphigus newsgroup, who have been kind enough to share their experiences with me and the group. I gratefully acknowledge those who have helped me come up this learning curve and have put me on the road to recovery.

Note: As always, check with your medical practitioners before making changes to your health regimen.
When the eyes have it

By Edward Tenner M.D.
Hoffman Estates, Ill.

Each type of pemphigus and related diseases has differing kinds and differing percentages of ocular involvement. Also, the treatments for these diseases have many ocular side effects. Therefore it is very important that an ophthalmologist examines the eyes of patients with pemphigus. This is especially true at the beginning of symptoms and treatments so any eye problem can be promptly handled.

Ocular involvement in pemphigus vulgaris is uncommon according to the medical literature. However, a group of patients have been described with eye irritation, excessive tearing, and foreign body sensation where the eye symptoms preceded the appearance of mouth and skin lesions.

Blisters can involve the skin surface of the eyelids. Since the skin around the eyes is very thin this area must be cared for gently, usually with warm compresses and a mild steroid cream. Blisters may also form on the eyelid margins, but are less common on the bulbar conjunctiva (covers the white part of the eye).

These rarely cause scarring. More common is a purulent or catarrhal (lots of mucus) conjunctivitis with some redness of the conjunctiva. Since this is usually not infectious only warm compresses are needed. Blepharitis (crusted matter along eyelid margins) can occur. Treatment entails warm compresses and eye lid scrubs. Dry eye irritation can be a problem which is treated with preservative-free artificial tears.

Eye involvement in pemphigus foliaceus is more common. The skin around the eyes and eyebrows is often affected. A quarter of all patients lose part or all of their eyebrows. Pemphigus foliaceus often affects the skin of the eyelid and eyelid margins. If severe the eyelid can be scarred causing entropion (the eyelid turns inward) and trichiasis (eyelashes grow in abnormal directions, can rub against eye). While blisters on the palpebral (inside of eyelid) or bulbar conjunctiva are rare, conjunctivitis with redness and mucous secretion is often noted. Blisters of the cornea (clear "windshield" front of eye) usually involve the middle or lower part of it.

They can lead to inflammatory changes and pannus (scarring) formation of the cornea. The iris (colored part of eye) can be involved, with a degenerative reddish lesion on the surface of the iris. Cataract (cloudiness of the clear natural lens of the eye) is also commonly noted with variable size, shape, and color in the anterior lens cortex (soft outer part of lens).

Paraneoplastic pemphigus (a distinct type of pemphigus associated with an underlying cancer) commonly has eye involvement. A marked conjunctivitis with erosions was present in a majority of patients described. In one article this was treated with eyelid scrubs, antibiotic ointment, and steroid drops. Conjunctival biopsy has been used to help confirm this diagnosis.

Bullous pemphigoid is an autoimmune blistering disorder sharing some common features with pemphigus. Occasional involvement of the conjunctiva and eyelid margins with inflammation and scarring has been reported.

Cicatricial pemphigoid is a related disorder with common involvement of mouth and eyes. In the two thirds of cases when the eye is affected, it is also called ocular cicatricial pemphigoid. Both eyes are usually affected with a chronic conjunctivitis with thick stringy discharge, and burning and tearing. Fibrosis (scarring) beneath the conjunctival epithelium (surface cellular layer) is typical in this disease. As the scarring progresses symblepharons (fibrotic bridges between the conjunctiva of the eyelid and the eyeball) are characteristic. Also dry eye symptoms are created by destruction of tear component forming cells and tear duct openings. As the scarring gets worse the cornea is involved with new blood vessels growing in the previous clear tissue. Eventually the eyelids and eyeball fuse together (ankyloblepharon).

With the eye not able to close, the cornea becomes opaque and blindness ensued in almost one third of patients before there were effective treatments. Immunosuppressive drugs e.g. Cytoxan, Prednisone, and Dapsone are usually effective in controlling the disease along with aggressive use of lubricating drops and ointments. Any eye surgery must be undertaken with care because it can reactivate the disease.

As to side-effects from medications, Prednisone is the major source. Its two major eye side-effects being cataract and glaucoma. The cataracts are a special type called posterior subcapsular cataract. They refer to a cloudiness of the back part of the lens. They tend to grow slowly, usually after using Prednisone for along time at high levels.

If the Prednisone is stopped, the cataract remains stable, and as long as it is not decreasing vision, can be left alone. If the vision is impaired the cataract is removed surgically by a procedure called phacoemulsification where the cataract is sucked out of the eye. Then a plastic intraocular lens is put in its place. Glaucoma can occur with oral Prednisone or Prednisone drops. The people whose eye pressure increases are called steroid re-sponders. The increased eye pressure usually develops within a few weeks to months. If the Prednisone has to be continued, treatment with glaucoma drops is necessary to prevent damage to the vision. Prednisone can bring out diabetic tendencies and diabetics need to be checked for eye complications. Also because Prednisone suppresses the immune system, people who have had herpes involving the eye can get recurrences and others may be more susceptible to new eye infection.

Fortunately, advances in medical care have helped to minimize the extent and impairment all of these problems can cause.
Pemphigus Questionnaire

Since little information is available on pemphigus from a patient’s point of view, we must obtain it for ourselves. This survey is the first step toward gaining that knowledge. Thank you for participating in this very important project.

Instructions: Please check the answer indicating your response and/or fill the blanks the best you can. Then photocopy or cut out this page and mail it to:

International Pemphigus Survey #1
1350 Washington St. #21
San Francisco, CA 94109

1. Sex: ___ Male  ___ Female

2. Age now: ____________________

3. What was your age at the onset? ____________________ years

4. What type of pemphigus?
___ Pemphigus vulgaris
___ Pemphigus foliaceus
___ Bullous pemphigoid
___ Cicatricial pemphigoid
___ Other ____________________

5. Where were you born?
___ USA
___ Canada
___ Central America
___ South America
___ Africa
___ Subcontinent (India, Pakistan)
___ Western Europe
___ Eastern Europe
___ Former Soviet Union
___ Asia:
___ Pacific Islands:
___ Australia/New Zealand
___ Other ____________________

6. What is your ethnicity? (Check all that apply)
___ American/Alaskan native: ____________________
___ Asian: ____________________
___ Black/nonhispanic: ____________________
___ East Indian (Subcontinent): ____________________
___ Hispanic: ____________________
___ Jewish
___ Occidentals (white/non-Jewish)
___ Persian

7. Who correctly diagnosed your disease?
___ General Practitioner
___ Dermatologist
___ Dentist/Oral Pathologist
___ Other: ____________________

8. Who, if anyone, misdiagnosed your illness? For how long? (In years) How long?
___ General Practitioner
___ Dermatologist
___ Dentist/Oral Path.
___ Other: ____________________
___ I was not misdiagnosed

9. Did you have an incorrect biopsy? ___ Yes ___ No

10. How long before the disease was controlled (i.e. no new lesions)? ____________________ months

11. Which part(s) of your body were involved during your first outbreak and how bad were they? (Please indicate the severity of each, using the guide below.)

| 0 | No symptoms at all |
| 1 | Slight, hardly noticeable |
| 2 | Occasional, very mild |
| 3 | Noticeable but mild |
| 4 | Moderate but could work |
| 5 | Moderate and could NOT work |
| 6 | Moderately severe, troublesome |
| 7 | Severe, but no hospitalization |
| 8 | Severe, required hospitalization |

I had lesions on my...
___ Scalp (If none mark 0, etc.)
___ Face (Not including eyes)
___ Trunk
___ Arms/Legs
___ Eyes
___ Nose (Inside)
___ Mouth/throat (Can swallow)
___ Mouth/throat (Difficulty swallowing)
___ Rectum
___ Vagina

11. Which symptoms associated with Prednisone did you incur? (Please use the same values as in question above. No involvement=0, an so on.)
___ Weight gain, (___ lbs ___ kilos)
___ Muscle weakness
___ Loss of muscle mass
___ Osteoporosis
___ Vertebral compression fractures
___ Stomach distress, ulcer, etc.
___ Impaired wound healing
___ Varicose veins
___ Increased sweating
___ Suppressed reaction to skin tests
___ Neurological difficulties (convulsions, etc.)
___ cushingoid syndrome (moon face)
___ Depression
___ Diabetes
___ Thin hair
___ Menstrual irregularities
___ Impotence
___ Increased body hair
___ Mood swings
___ Reduced ability to concentrate
___ Fatigue
___ Other: ____________________

13. Have you had eye problems before the course of your disease?
___ Skin around eyes and eyelids
___ Conjunctiva
___ Cornea
___ Cataract
___ Glaucoma
___ Other: ____________________

14. Have you had eye problems during the course of your disease? ___ Yes ___ No

15. What kind of eye problem did you have? (Mark all appropriate)
___ Skin around eyes and eyelids
___ Conjunctiva
___ Cornea
___ Cataract
___ Glaucoma
___ Other: ____________________

16. Did you receive any eye treatment?
___ Medications (List if known)
___ Surgery
___ Went away as disease was controlled
___ Other: ____________________
Physician@the_office.com

Doctors, patients and email

By Eileen Lucey

S
omeone in your office just mentioned a new health supplement, one which is recommended for exactly the symptoms you are experiencing. You know that information received in the coffee room can be less than reliable, but you are still interested. You think about checking the Internet, but you have sometimes found the sites for confusing or contradictory. You could call your doctor, but that’s often been a frustrating experience, and you dislike taking your time at work for personal business. So, send your doctor an email! Or can you?

The Internet is certainly being used by people who want to educate themselves and take an active part in their health care. When MEDLINE, a medical information service, offered free access to the public in 1997, usage jumped to a rate of 75 million searches annually, with 30% of those coming from the general public. In some cases, however, the sheer volume of information on the Net can be intimidating, and you still want to know what your doctor thinks. While using email to correspond to your doctor is a good idea, there are questions and concerns both you and your doctor need to address.

First, from the doctor’s point of view: there is a legitimate fear that handling a medical problem in a question and answer format would leave room for error, for overlooking issues that would be evident in an exam or for misunderstanding. In studies of medical facilities using email to answer queries, care was always taken to mention that a diagnosis could not be made using email.

The studies found email to be most useful as part of a planned course of treatment, where patients could report results of tests done at home (diabetes, blood pressure, etc.) or send questions that did not need an immediate reply. The scenario of hearing about a supplement is a perfect example: important enough to involve your doctor, but not critical enough to require intervention. You can certainly wait a day or two before buying the herbal tea or energy capsules; time enough to hear from your doctor about possible interaction with whatever other medications you are using.

Studies show that if the doctors are committed to reading and answering their email within a previously agreed upon time frame, they find that the time it takes is usually far less than would be used to return a phone call. There is the additional plus that one can look up all the information needed without keeping the patient on hold or having to call back with the answer after doing the research. It is certain that email would not be the correct way to handle all communications with your doctor, but there are many instances where it would be valuable for both parties.

The Journal of the American Medical Informatics Association has published a paper which outlines the guidelines for patient/doctor email (http://www.amia.org/position2.htm). The article makes the point that email allows confirmation and clarification of information received at the time of an exam. The stress of illness can easily cause any of us to forget oral instructions or realize later that there are questions we didn’t ask. “Examples include addresses and telephone numbers of other facilities to which the patient is referred; test results with interpretation and advice, instructions on how to take medications or apply dressings; pre- and postoperative instructions; and other forms of patient education.” Certainly we would not use email to report an adverse reaction to medication or some other possibly dangerous circumstance, but the potential for streamlining service in some areas is clear.

In summarizing the guidelines, the AMIA article makes clear that the most basic requirement is a clear agreement between doctor and patient. The parameters should be established from the start, leaving all concerned comfortable with the arrangement. Some of the points they make:

- Establish a turnaround time for messages.
- Discuss privacy issues. You should know who besides your doctor reads his email, who has access to the files and who handles checking during vacations or illness.
- Establish the types of transactions which can be handled by email. Prescription refills can be effectively handled this way, but you might not want an email with the subject “blood test”, especially if you are using email services at work.
- Make your email easy to sort by placing a clear statement in the subject line: “appointment,” “prescription” or “billing question.”
- Make sure your name and medical record number appears in the body of the message. As many email addresses are nicknames or office codes, you need to assure the doctor can identify you and correctly place your query.

Some doctors might also ask you to sign an agreement that they will not be held accountable for messages lost because of technical failures. Since nothing urgent should be handled by email, an agreement of this sort would seem to be reasonable and fair.

For many patients, one benefit in the arrangement is the time they can take to formulate their question, clarify the response they would like and assure that they have the doctor’s full attention in dealing with their issues. This is also valuable to the doctor, since he can be assured that he understands your question and can take the amount of time needed to give you the service you need. If you have found information on the Internet and want your doctor to see it, you can send him the URL (Uniform Resource Locator which is the Internet address) along with your question, assuring that he gets the complete picture. Some patients find the distance provided by putting their questions in writing allows them to raise topics that they might find embarrassing to discuss in full while sitting in an exam room.

Again, it is important to be clear on who will have access to your information, since most email systems are not currently guaranteed secure. There is legislation pending in Congress which will define the limits to which privacy will be protected during electronic transfer of information, but there is no consensus and some question about how privacy can be protected while also allowing the free flow of information so necessary to medical research. Until such time as these issues are settled, protect yourself. Talk to your doctor, ask him how he feels and see if you can simplify some of your health care by taking the Information Superhighway.
...new perspectives on pemphigus

Continued from page three

pemphigus antibodies were added to the culture [20]. In 1984, the Dr. Ahmed research group reported that similar results can be obtained using the corticosteroid hormone called hydrocortisone [21]. Since in either experiment, lymphocytes were not present in the skin cultures treated with corticosteroids, it was clear that the Prednisone was not working by way of acting upon lymphocytes and was instead directly affecting the keratinocytes themselves. This explains why antilymphocyte immunosuppressive drugs such as Imuran (azathioprine), Cytoxan (cyclophosphamide) or Sandimmune (cyclosporin) cannot control pemphigus on their own.

In our laboratory, we now find that corticosteroids stimulate keratinocytes to manufacture the increased amounts of the same protein that is destroyed upon binding by our novel primary pemphigus antibody [22]. Normally, this novel keratinocyte protein acts as a receptor binding to a signaling molecule, acetylcholine, which plays essential role in stimulating keratinocytes to maintain their shape and connection to each other. Similar proteins have been found in other tissues and shown to mediate therapeutic effects of corticosteroid by inhibiting the pathways of local inflammation that destabilize the plasma membranes. When a primary antibody parks at the receptor in acetylcholine’s place, it initially activates the receptor, but then causes it to stop working due to degradation of the improperly functioning receptor through a process termed receptor “desensitization.” What then occurs, the signal to stretch is broken, the desmosomes are shed, and the keratinocyte balls up (acantholysis). These observations led us to formulate a new hypothesis regarding the treatment of pemphigus.

Hypothesis two: Corticosteroids are effective in the treatment of pemphigus, at least in part, because they stimulate replacement of the acetylcholine receptor destroyed by the primary antibody, thus counterbalancing the effect of disease-causing antibody on keratinocytes. This stabilizes the keratinocyte plasma membrane, and, consequently, leads to regrowth of desmosomes. Since this response occurs directly at the keratinocyte level (that is, it is not mediated by way of lymphocytes), we believe that it may be possible to achieve that same effect from the use of non-corticosteroid drugs that can prevent primary antibody binding to keratinocytes.

Testing of this hypothesis is currently underway. We have already identified several nonhormonal drugs that can abolish pemphigus antibody-induced acantholysis in keratinocyte cultures [23]. One of them is an acetylcholine-like molecule called pilocarpine. Pilocarpine can mimic both acetylcholine and primary antibody in a sense that it can attach the same protein on the keratinocyte plasma membrane. In contrast to the primary antibody, however, pilocarpine binding should not lead to receptor destruction. The Institutional Review Board (IRB) of the University of California Davis Medical School has approved our request to evaluate the clinical effectiveness of a topical application gel containing pilocarpine. This medication, PILOPINE HSÓ GEL, is already approved by the Food and Drug Administration for placement in the eyes as part of the treatment of glaucoma.

We are now enrolling in this study patients with pemphigus. To be eligible, patients must have two or more active lesions. Patients who are already receiving other therapy will be allowed to continue with their current treatment. Patients who have not yet started treatment must have disease mild enough so that there is no problem delaying the initiation of other treatment for about two weeks.

Eligible patients participating in the study will need to attend the Dermatology Clinic at UC Davis Medical Center on two occasions. Lesions of pemphigus will be photographed and samples of blood and tissue (skin biopsy) will be taken. Patients will be given two un-labelled, identical appearing gels. One gel will be PILOPINE HSÓ GEL and the other will be a placebo (inactive) gel. Each gel will be applied to a separate lesion on an once daily basis for two weeks. During the two weeks application period, no changes will be allowed in other medications which are being taken for the treatment of pemphigus. To learn more about this study, you may contact me by letter, by telephone (916/734-6057), or by email (sagran@ucdavis.edu), or write:

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References:

The National Pemphigus Foundation

GOT SHIRTS?

+ T-SHIRTS $15.00 each, adult sizes only.
Back: Please Support the National Pemphigus Foundation
S ___ M ___ L ___ XL ___ XXL ___ Quantity ___

+ BUMPER STICKERS $2.50 each, white on blue Quantity ___

+ BUTTONS $2.00 each, 2" round, blue metal-flake color "Support the National Pemphigus Foundation" Quantity ___

Total amount enclosed $ ___

NAME: ____________________________
ADDRESS: _________________________
CITY: ___________________________
STATE/COUNTRY ___________________ POSTAL CODE ____________

Return with check or money order (U.S. funds only) to: The NPF, Midwest Chapter, 514 Inverary Lane, Deerfield, IL 60015. All prices include shipping.

Coping with Prednisone
by Zukerman & Ingelfinger

If you live overseas and previously could not get this important book, now you can. Just send $30.00 (U.S. funds only) to the NPF and a copy will be sent to you. See page two for the Foundation address.

The National Pemphigus Foundation

If we don't tell you, who will?

Please detach the form below and return to:
The National Pemphigus Foundation
P.O. Box 9606, Berkeley, CA 94709-0606. Please print clearly.

☐ I would like to receive the quarterly newsletter. I enclose my annual donation of $50.00 ($65.00 outside the United States) to continue the Foundation’s efforts on behalf of those living with pemphigus. These funds help offset the costs of the NPF website, the online newsgroup and the quarterly newsletter, among other things. Please make contributions in U.S. funds.

☐ I would like to make an additional donation of $ _________ to further the work of the Foundation.

☐ I would like to receive the newsletter but I am under a financial hardship. I enclose a donation of $ _________ to cover the costs of printing and postage.

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Address: ___________________________
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New Medical Board member tells how it works in Australia

By Dedee F. Murrell, MA, BMBCh, FAAD
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NPF Medical Advisory Board

Anyone who has the right of permanent residence in Australia or is an Australian citizen is entitled to receive health care benefits in the form of Medicare from the Australian Government. The patient receives a card with their own Medicare number which means that visits to family physicians and dermatologists are paid for in part by the government. The out-of-pocket expenses for a patient seeing a dermatologist in private practice would range from zero to $40.00 depending on the generosity of the specialist or whether the patient had received a government pension.

Private health insurance in Australia does not cover any outpatient care, so it is not necessary for ongoing outpatient treatment. Patients with pemphigus quite often attend a University Hospital outpatient clinic, in which case there is no out-of-pocket expense involved, but there is a long waiting list for the low numbers of these clinics. This only applies to private practice offices outside the hospital system. Currently there is no central specialty clinic for patients with autoimmune bullous diseases. There are a number of dermatologists interested in this area, as well as immunologists. In the majority of instances, the patients are managed individually by their private practice dermatologists.

Patients may be admitted to either the public hospital system which involves no out-of-pocket expenses or to a private hospital. When patients are admitted in Australia to a private hospital the health insurance that they have covers the hospital room, drugs and hospital fees, but the patient still has to meet all the doctor’s fees. Whilst a portion of the doctor’s fees can be claimed from Medicare, there is still an out-of-pocket cost. Health insurance is therefore generally much cheaper than in the United States and is paid for by the individual and not by employers. The overall tax rates in Australia are much higher than in the U.S. to pay for this generous Medicare system.

In the public hospitals, the cost of expensive drugs for treating pemphigus outpatients are met by the hospital pharmacy. If the patient is seen in the outpatient clinic of the public hospital, they only pay $3.20 per prescription. If prescribed by a private practice dermatologist and collected at an outside chemist, some of the drugs will be covered by the public benefit system or “PBS”, such as prednisolone, methotrexate and azathioprine. In this case patients would pay about $20.00 per prescription. After they have spent $612.00 per year on prescriptions, any prescribed medications are at no cost.

Paradoxically, inpatients in a private hospital have to pay for their PBS medications, but their insurance covers non-PBS items. Cyclosporin is also on the PBS for limited indications including transplant patients and severe psoriasis but would normally have to be obtained through the hospital outpatient pharmacy with special arrangements on a case-by-case basis from the hospital’s drug committee to be used for other indications.

In summary I think that patients with severe chronic diseases such as pemphigus do not have to worry excessively about the cost of their treatment in Australia. Improvements however could be made in terms of having specialist clinics for patients with pemphigus depending upon changing referral patterns of dermatologists here. In general, the public hospitals prefer to fund general outpatient dermatology clinics rather than specialty clinics. Those specialty clinics which exist have been set up in the private sector, which does not have access to the public hospital pharmacy.

...dangerous trends

Continued from page two

The second trend does not concern the present, but rather the near future, and that is the potentially devastating possibility that specialized physicians and researchers may not be available at all.

The amount of money available to physicians and researchers in the academic community has been cut back to alarming levels. There are two main reasons for this. First, many academic hospitals are being required to compete with the HMOs and secondly, research grants formerly available to patient-oriented researchers are being shifted to more generalized research. Patient-oriented research (POR) is that which is done with humans as opposed to “core research” such as molecular biology on animals).

New ideas come from POR because the researchers see trends and they try new drugs and techniques. In fact, in the recent past, 50% to 75% of all the articles in medical journals have come from patient-oriented research.

To compete with HMOs, doctors who previously spent 50% of the time with patients and 20% to 30% of their time on research are now required to see patients 70% or 80% of the time leaving the remaining time for patient support and teaching. Little or no time is left for research. POR needs to be funded and the best way is through grants, which leads to the second half of this problem.

While all civilian research has seen government funding cuts, the National Institutes of Health’s research budget has nearly doubled in the last 10 years. This means that the NIH is now responsible for more than 25% of all the research done in America. New priorities at NIH have moved strongly toward core research. This means that those working so hard to study individual diseases are not getting a fair share of funding.

It is time for our elected officials to get back to the people’s work and consider health care. Presently the foxes are watching the hen house, and this can only bring further complication and delay to an already difficult situation.

Be well.

Janet Segall
President
The Foundation expands 1999 agenda

The NPF will add at least one new event to the calendar this year. For the first time, the Foundation will attend the Dermatology Nurses’ Association meeting in March. This should be extremely productive, as these nurses, along with the doctors at the American Academy of Dermatologists convention, are actually the audience most in need of education concerning pemphigus.

NIAMS Day—On March 9 & 10, representatives of the NPF will go to Washington, D.C. again this year for “NIAMS Day.” The National Institute for Arthritis, Musculoskeletal and Skin Diseases (NIAMS) is a division of the National Institutes of Health (NIH). Pemphigus is associated with this division. Our representatives will meet with Dr. Steven Katz, Director of NIAMS, to discuss what the NIH in general, and NIAMS specifically, are doing to help fund research for pemphigus.

The next day, Foundation officials will visit several of our Congressional representatives to discuss pemphigus and inform them of NPF support for a substantial increase in funding for the NIH.

Dermatology Nurses’ Association—March 18-19, the Dermatology Nurses’ Association meets in New Orleans, where the NPF will join the Coalition for Patient Advocacy for Skin Disease Research (CPA-SDR) to work the information booth. This is the first time the Foundation will have an opportunity to present information at this important meeting of the dermatology nurses.

The AAD annual conference—Beginning March 19 also in New Orleans, The American Academy of Dermatology (AAD) will hold its annual meeting. We will present our information booth as we have in the past. Here we provide brochures to physicians and interested visitors. Traditionally this has been a very productive event for the NPF, as dermatologist generally provide care for pemphigus patients.

CPA-SDR Conference—Congruent with the AAD event, the Coalition for Patient Advocacy for Skin Disease Research (CPA-SDR) holds its annual meeting. As a member, the Foundation will take part in discussions on topics of mutual interest and coordinate efforts toward common goals.

SID Conference—Although the National Pemphigus Foundation will not have a booth of its own, the Coalition for Patient Advocacy for Skin Disease Research will have a table at the Society of Investigative Dermatology’s (SID) Annual Meeting in Chicago, IL May 5 to May 9. The Foundation is inviting anyone in the Chicago area to help out at CPA-SDR information table. The hours of the event are not yet available. If you are interested, please contact Janet Segall. Contact information is available in the staff box on page two.

Second NPF Conference takes aim at the big apple

The National Pemphigus Foundation’s second annual conference is scheduled to take place July 31 and August 1, in New York City. This is designed to coincide with the AAD Summer Convention.

“We want to thank Dr. Jean-Claude Bystryn for securing a meeting room for the Foundation at New York University’s Medical Center for both days,” said NPF President Janet Segall.

The guest speakers, program and hours are yet to be finalized. “At this time we expect the program will include physicians, nutritionists and an expert on health care.” Conference attendees will enjoy dinner on Saturday night at a nearby restaurant, not yet determined. The Foundation hopes to be able to offer conference attendees hotel rooms in the NYU area at discount rates.

In order to provide a probable headcount (required to secure hotel rooms and restaurant reservations), the Foundation needs to hear from those interested in attending one or both days of the conference. “The first conference last year in Chicago was extremely successful and everyone is encouraged to attend this year’s conference. We want to make this event even bigger and better,” Segall said.

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

We’re online: www.pemphigus.org