Treatment of pemphigus and linear IgA dermatosis with nicotinamide and tetracycline: A review of 13 cases
by Marsha L. Chaffins, M.D., Daniel Collison, M.D., and David P. Finenson, M.D.,
Detroit Michigan, and Hanover, New Hampshire

Pemphigus usually requires long-term therapy with oral corticosteroids, which can cause significant morbidity. Several immunomodulating drugs such as cyclosporinamide (Cytoxan), azathioprine (Imuran), and gold have proven beneficial as steroid-sparing agents. However, these agents also have limited long-term utility because of their potential to induce renal hepatic dysfunction and bone marrow suppression. Nicotinamide in combination with tetracycline has been reported to be effective for bullous pemphigoid (BP) and IgA bullous dermatosis (LABD). This regimen has the advantage of lower toxicity compared with corticosteroids and immunosuppressant regimens. We have treated 11 cases of pemphigus and two cases of LABD with nicotinamide and tetracycline and report our experience.

Material and Methods: Eleven patients with pemphigus (six with pemphigus vulgaris (PV), three with pemphigus foliaceus (PF), two with pemphigus erythematosus (PE) and two patients with LABD were treated with nicotinamide, 1.5 g/day, and tetracycline, 2 gm/day, with or without oral corticosteroids as summarized in Table I. The clinical diagnosis was confirmed in all cases by routine histopathology and immunofluorescence studies. Therapeutic responses were graded by the degree of clinical improvement after 8 weeks of treatment. Responses were recorded as follows: complete response (CR) - total clearing of lesions; partial response (PR) - clearing of more than 50% of lesions; and no response (NR) - clearing less than 50% of lesions or worsening of disease.

A complete blood work up was done on all the patients before beginning therapy, after 4 to 8 weeks of treatment, and periodically thereafter. In four patients pemphigus autoantibodies were determined by indirect immunofluorescence on monkey esophagus at the time of diagnosis and after at least 8 weeks of treatment.

Results: The therapeutic responses are summarized in Table I. When evaluated as a group, 7 of 13 patients experienced CR. Four patients had PR, and two patients failed to respond. Of the six patients with PV, three had CR, two had PR, and one had NR. Only two patients with PV were able to be treated with nicotinamide and tetracycline alone; the four other patients required a mean daily dose of 8 mg of prednisone to control disease activity. Patient 3 was also treated with 150 mg of azathioprine per day. The follow-up period for these patients ranged from 6 to 13 months. Of the 5 patients with superficial pemphigus, two had CR, two had PR, and one did not respond. Only one patient

Continued on Page 3
I would like to announce, that from now on, I will be Janet D. Segall. I have decided, due to personal reasons, to return to my maiden name.

I want to bring to you my thoughts on fund-raising and the Foundation’s efforts to procure the public’s and medical communities support I believe we need. With the development of our Medical Advisory Board, and our participation with the Coalition in Washington and at the Dermatology Convention, we are slowly becoming a recognized entity in some medical circles, but, we need more exposure. Many physicians are still not aware of our existence, and what we can do to help their patients who are living with pemphigus. We need public support. We need to find a path to the public, and with this recognition, perhaps more funding to promote our goals and objectives. Unfortunately, the bottom line is always, a financial one, even if it just means keeping the phone lines and newsletter in business, but I would certainly like to see us go beyond that. This means fund-raising, and finding a link to the media. This is a very difficult task. We have been writing letters to the media; so far with no response. Some very dedicated people are working very hard to try and raise money for our cause. This fall (in a special mailing) we will be putting out to you a fund-raising packet that I hope all of you will take the time to look at and, hopefully, participate in.

I know that asking people for money is probably the hardest thing for most of us to do. I know that when I decided to start the Foundation, I asked the help of my friends and relatives, many of who were reluctant to contribute, but I felt my life was at stake, so I had to at least try. I figured the worst they could do was say no, and some did. But, to my surprise, most did not, and the Foundation was born. I have met with representatives of some of the more financially successful foundations, most with budgets of over half a million dollars, and they tell me that they receive most of their funding from foundations, or companies, or individuals who either have the disease or are associated in some way with someone who has the disease. Some of these foundations have less or about the same number of afflicted individuals as we do. But what they have, and we do not, is public awareness.

I was recently made aware of a woman who, with a rare disease, was able to bring in substantial funding by a few minutes appearance on “Good Morning America”. So, I ask anyone out there with any kind of connections to media, to ask for their help. Some of us have been on dangerous drugs long enough. Some of us have been on these drugs too long. The research is out there to help us get off these drugs sooner rather than later, but we need to support it now not when it is too late for some of us. We need to do this for our lives. So, I ask for your support during our Fall Fund-raising Campaign, and hope that by the New Year we will be reporting success!

I would like to strongly recommend a new book on the shelves called “Coping With Prednisone” by Eugenia Zukerman and Julie R. Ingelfinger, M.D. I have read this book and feel that it has addressed issues that are important to all of us living with prednisone every day. Eugenia Zukerman was diagnosed with a rare lung disease and was placed on high doses of prednisone. Julie R. Ingelfinger, M.D. is a physician and Ms. Zukerman’s sister. Together they have managed to put together a very comprehensive look at prednisone. The promotional material describes the book very well.

“Coping with Prednisone explains exactly what glucocorticoids are, how they work, and why they are used. The book covers a wide range of possible side effects, both physical and psychiatric, and offers practical steps that prednisone users can take to make this course of treatment as trouble free as possible.”

Look for this book at your local bookstore and if it is not in stock, ask them to order it for you. It is published by St. Martin’s Press. ♦
UPDATE

Our e-mail support group is going well. We want to repeat the steps to take if one wants to join. To get started, you need an e-mail address. Then send a message to majordomo@lists.cyberwar.com. In the body of the message put: subscribe pemphigus_support.” The list will send you instructions about how to remove yourself should you want to do that. If you have any questions contact Miriam Schneidmill at zapot@cyberwar.com.

Milo Carreaga is setting up a support network for young adults (people in their 20’s) with pemphigus. If anyone fits into that category, contact Milo by e-mail at: Milo@csus.edu or by regular mail: 2220 River Trails Circle, Rancho Cordova, CA 95670, Phone: 916-362-3584.

The Foundation’s website should be up and running by the end of October thanks to the work of Sal Capo here in California. The website will provide basic information on pemphigus and other related issues. *

ATTENTION: DAPSONE STUDY RECRUITMENT

Patients with stable pemphigus vulgaris, but unable to taper Prednisone, may be eligible for a study looking at Dapsone as a steroid-sparing drug.

• Must be in maintenance phase of pemphigus vulgaris
• Must be taking 15-40mg of Prednisone daily, or 20-40mg every other day and unable to drop the dose without suffering a flare.
• Additional criteria apply.

Doctors may participate in this study by “piggybacking” onto the IRB at the University of Pennsylvania. Contact Dr. Victoria Werth for more information.

Tel #: 215-662-2399
Fax #: 215-349-8339

Table 1

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age/Sex</th>
<th>Diagnosis</th>
<th>Concurrent treatment</th>
<th>Response</th>
<th>Follow-Up period (mo)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>34/M</td>
<td>PV</td>
<td>Topical steroids</td>
<td>CR</td>
<td>8</td>
</tr>
<tr>
<td>2</td>
<td>47/F</td>
<td>PV</td>
<td>Prednisone 5 mg q.a.d - 30 mg, q.a.d.</td>
<td>PR</td>
<td>6</td>
</tr>
<tr>
<td>3</td>
<td>71/F</td>
<td>PV</td>
<td>Prednisone 5 mg/day - Azathioprine - 150 mg/day</td>
<td>PR</td>
<td>10</td>
</tr>
<tr>
<td>4</td>
<td>81/M</td>
<td>PV</td>
<td>Topical steroids</td>
<td>CR</td>
<td>9</td>
</tr>
<tr>
<td>5</td>
<td>57/F</td>
<td>PV</td>
<td>oral Prednisone 2.5 mg/day</td>
<td>CR</td>
<td>14</td>
</tr>
<tr>
<td>6</td>
<td>41/F</td>
<td>PV</td>
<td>oral Prednisone - 10 mg/day</td>
<td>NR</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>69/F</td>
<td>PF</td>
<td>None</td>
<td>CR</td>
<td>24</td>
</tr>
<tr>
<td>8</td>
<td>51/M</td>
<td>PF</td>
<td>Topical steroids</td>
<td>CR</td>
<td>41</td>
</tr>
<tr>
<td>9</td>
<td>50/M</td>
<td>PF</td>
<td>Topical steroids</td>
<td>PR</td>
<td>11</td>
</tr>
<tr>
<td>10</td>
<td>28/F</td>
<td>PE</td>
<td>Prednisone - 7.5-0 mg/day</td>
<td>PR</td>
<td>13</td>
</tr>
<tr>
<td>11</td>
<td>73/M</td>
<td>PE</td>
<td>None</td>
<td>NR</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>70/M</td>
<td>LABD</td>
<td>antihistamines - topical steroids</td>
<td>CR</td>
<td>2 mo w/medication - 11 mo. clear</td>
</tr>
<tr>
<td>13</td>
<td>69/F</td>
<td>LABD</td>
<td>Topical steroids</td>
<td>CR</td>
<td>19</td>
</tr>
</tbody>
</table>
The New York Area Support Group was pleased to have Dr. John E. Fantasia as its guest speaker at the September 11 meeting. Dr. Fantasia, Chief of Oral Pathology at Long Island Jewish Medical Center in New Hyde Park, NY, gave a presentation titled, "The Oral Manifestations and Treatments for Pemphigus."

The meeting - which attracted another impressive-sized crowd - was held at New York University Medical Center in Manhattan, thanks to Dr. Jean-Claude Bystryn, Professor of Dermatology at NYU. For many of the people and family members present who were unfamiliar with this medical discipline, Dr. Fantasia provided a brief overview of oral pathology and its role in diagnosing and treating pemphigus. This "obscure subset of the dental profession," as Fantasia calls it, specializes in diseases of the oral cavity. And since, according to Fantasia, in some 60 to 70 percent of all adults with pemphigus, initial symptoms first appear in the mouth and oral cavity, oral pathologists see a proportionally high number of pemphigus patients compared to other medical professions. In fact, Fantasia added, in children with pemphigus, the percentage of those with initial oral symptoms jumps to 90 percent. As a result, Fantasia said, the oral pathology profession - which numbers just a bit over 200 board certified doctors nationwide - works closely with the general dentistry community to help them recognize signs and symptoms of pemphigus so referrals to oral pathologists and dermatologists can be made in a timely fashion and before the disease worsens.

He said that oral pathologists can be found at many large teaching hospitals and medical university centers with dental schools. Dr. Fantasia provided a comprehensive slide-show presentation about pemphigus, complete with a history of the disease (did you know there are references to a pemphigus-sounding "blistering skin disease" in medieval writings?), the various manifestations of the disease, and treatment options. He very cordially took questions from the audience and stayed afterwards to discuss specific issues with those present.

One area of significant interest during the Q&A part of the meeting concerned oral hygiene and dentistry issues for people with pemphigus. While Fantasia cautioned against invasive dentistry work when pemphigus is active, he added that routine fillings and cleaning pose few problems for most. However, he cautioned that if there is any chance of abrasion to the gum or inner oral cavity, that we consult with our doctor about increasing our medication for a period of time before and after the work. Fantasia also cautioned against the use of alcohol-based mouthwashes, and suggested that people with pemphigus use one of the non-alcohol-based products on the market.

A notice will be sent to those on the New York Area Support Group list informing them of the next meeting date. If you are not on our mailing list and would like to attend future meetings, contact Joan Delucie, the group leader, via e-mail at jdelucie@aol.com or call her at (516) 586-6910.

**Philadelphia Support Group**

The next meeting of the Philadelphia Support Group will be October 24, 1997 at 5pm. Dr. Werth will speak on the dapsone study. For further information contact Miriam Schneidmiller at 201-471-2278 or e-mail: zapot@cyberwar.com.

**San Francisco Bay Area Support Group**

The SF group met on Sept. 14th, at UCSF. Our special guest speaker was Dr. M. Kari Connolly, Head of the Autoimmune Clinic at UCSF. We all want to thank Dr. Connolly for a very extensive and thorough talk on all aspects of pemphigus. We appreciate her support of our group. If anyone would like to join the group, please contact Janet Segall at The Foundation.

**Los Angeles Support Group**

*by Marcia Kassan*

Hi, Everyone! The last meeting was held on July 27, 1997. The volunteers for our Chapter Board are: Secretary - Carol Goren; Treasurer - Jag Patel; Printing and Mailing, Speaker Chairman, and Hospital Liaison - Marcia Kassan; and Fund-raising Chairman - Lee Heins. Lee Heins (through Janet Segall) presented a fund-raising proposal to the group. We will be discussing his proposal at the next meeting, which will be on Sunday, October 12, 1997. We will be holding our meeting approximately every three months. It was voted that we shall have yearly dues of $50.00. (Interested people in the Los Angeles Area, make your checks out to the Los Angeles Chapter of the NPF and send them to Jag Patel at 25073 Wintergreen Ct., Newhall, CA 91318)

We need your input for speakers to make our meetings interesting and informative. Please call Marcia Kassan at 818-340-7180 as soon as possible if you have someone in mind. Remember the date is October 12. Carol, our secretary, has proposed an agenda for the meetings of speaker, treasurer's Continued on next page
Support Group News, continued

report, fund-raiser business and at least 1/2 hour or more for us to share and support each other. A new roster is included. Lee’s proposal will be passed around at the next meeting.

Midwest Chapter Support Network
by Arlene Strauss-Popper

Awareness Program

Unfortunately, the Foundation did not get enough support from the subscribers of the newsletter for our fund-raising “Awareness Program” – for T-shirts and/or bumper stickers. I am reaching out to everyone and making a personal plea to you to help us in our efforts. We need the support of everyone within our small network to group together to help us obtain our goals. Recognition is the key to our success! Our hope is to be recognized by the public and government. They are our only hope for the funding we are seeking to keep the Foundation alive, and to assist funding the terrific research that is being performed in order for us to have better treatments now. Without your support we cannot achieve the goals the Foundation has set, or find the public help to promote our cause. The public does not recognize us. Pemphigus is here to stay and will affect our future generations unless we do something to help the Foundation find the recognition it must have to be effective. It is our only hope to be taken seriously when seeking public and government support.

I encourage everyone to participate in our plight. Let us make a life-changing, lifesaving difference for all who are living with any form of pemphigus! Attached is an order form for the T-shirts and bumper stickers. Please take a moment of your time and place an order. All of the proceeds go to the Foundation.

Please send the completed form and make out a check or money order to The National Pemphigus Foundation and mail it to:

The National Pemphigus Foundation
Midwest Chapter
827 Saffield Square Lincolnshire, IL 60069

Please allow 2-4 weeks for delivery.
Thank you for your support!

Entertainment Book

It is that time of year... the 1998 Entertainment Books are almost here. The Foundation is selling Entertainment Books as part of our late summer, early fall fund-raiser. The Foundation brings in $7.00 per book and additional earnings for out of town orders. This is a terrific way to raise funds for the Foundation and offer great savings for you! They make a wonderful gift that keeps on giving all year long. Check on the order form to see if your town is listed. If you need additional order forms, you can make a copy of the enclosed order form, or send me all the information on a separate piece of paper.

Dana Silverman and Arlene Strauss-Popper

WIN-WIN!

Support a great cause and get great savings for yourself, too! The Entertainment Book is loaded with hundreds of two-for-one and up to 50% savings on dining, shopping, travel, movies, sports and more. Best of all, a portion of your purchase goes directly to our fund-raising efforts. What better way to support your community!

It’s never too soon to start saving! Get your books today!

Help support our fund-raiser! Contact:

THE NATIONAL
PEMPHIGUS FOUNDATION
827 SUFFIELD SQUARE
LINCOLNSHIRE, IL 60069
A big thank you to everybody who took the time to respond to the survey in the last edition of the newsletter. Fifty one responses have been received to date.

This sample size is not a sufficient quantity from which to extrapolate any meaningful data, so we'll focus instead on general observations and trends.

Half of the respondents were male, the other half female. Age at diagnosis ranged from a low of 18 to a high of 82, with many being in their 60's and 70's. Some are recently diagnosed, within the last few months, and some have been living with pemphigus for over 30 years.

Approximately 1/2 of us are blood type O, 1/4 type A, 1 was type B, 1 was type AB and 1/4 didn't know their blood type. In asking this question, I was hoping to correlate various types of pemphigus, response to drugs, and/or side effects to drugs with blood type. Due primarily to the small sample size, no direct correlation stands out. The distribution profile roughly matches the occurrence of the different blood types throughout society. If anybody would like to further analyze the data for possible correlations, please contact me.

Almost all respondents indicated pemphigus vulgaris, and a few have pemphigus foliaceus. One person has pemphigoid, and none reported vegetans. Several people reported thyroid conditions, diabetes and hypertension as other chronic illnesses. Roughly half of us are experiencing lesions on both the body and inside the mouth, and a quarter each are either in the mouth only or on the body only. Several respondents indicated body lesions on only one part of their body, the head area predominantly. Pemphigus is known to occur on internal organs, and one respondent indicated this (cervix).

Most people are initially started on Prednisone or a combination of Prednisone and either Imuran or Cytoxan. Initial results were mixed, some responding very well, and others going from drug to drug trying to find a controlling regime. Again, a larger sample size with additional factors may reveal connections not seen here.

Almost everybody surveyed viewed their health prior to pemphigus as either good or excellent, with a few fair's and one lousy. Very few can look back and discern any sign that they were developing a serious illness.

Many people reported that they have never had a cavity, which I found surprising. Cavities, by many natural health barometers, are one of the first signs of a malfunctioning immune system. In future surveys, I may reword this question to indicate cavities over your entire life, whereas I believe many people answered this for a post-pemphigus time period. If not, and they indeed have had zero cavities, this poses an interesting anomaly.

Drug usage and history is extremely variable and very personal. Each person is different, and changes of drug and drug dosage over time are common. There do not appear to be any common denominators. Future surveys may address this and attempt to determine if and what a common thread is.

Several people reported that they are in remission. By way of clarification, it is important to understand the different types of remission that are used. Clinical remission means that a person is continuing to take a drug and does not have signs or symptoms of the illness (it is suppressed), versus remission, which means that the person is no longer taking medications and is disease free. Some respondents were in clinical remission, while others are in remission.

The most common side effects reported are: emotional distress, sleeplessness, weight gain, fatigue, and Cushingoid (pumpkin face). Next are muscle loss, diabetes, osteoporosis, potassium loss and hypertension. A few people reported problems with their eyes, frequent urination, and gastric distress.

Some people take no vitamins, minerals or supplements, while others take so many that a meal itself can probably be made from the pills. A similar trend was noticed concerning herbs and other nontraditional therapies. Some take none, others take from a wide variety, including herbs, acupuncture, homeopathy, and other products. Of those taking supplements and herbs, some appear to be taking them to help stop the pemphigus itself, while others are focusing on reducing the side effects of the medications.

Comments were of course open-ended. Each of us has experiences to share, from treatments, supplements or diet changes that we believe helped, to general comments about how we are doing. Perhaps you will consider writing a short article about your experiences for the newsletter.

If anybody would like to receive copies of the survey responses for their own analysis, please send me $6.00 to cover the cost of copying and postage. All survey responses are being kept confidential, I have crossed out the names and addresses of anybody who included them on their response.

Make checks payable to:
Steve Shapiro
PO Box 50353
Eugene, OR 97405-0977

P.S: I communicated with several people who did not respond to the survey. Procrastination was the overwhelming reason for non-response. I strongly believe that information is our strongest tool in understanding and healing pemphigus. I hope everyone will take the responsibility to be proactive in their own healing, and contribute to the open exchange of information.
RB Cameron
2112 Acacia Park Dr., Apt. 406,
Lyndhurst, OH 44124
216-473-1336

Bety Anne Cash
5816 E 64th St.
Tulsa, OK 74136-2137
(918) 492-1949

Gregory Davis
490 James St.
King of Prussia, PA 19406
(610) 337-8293

Joan Delucie
(216) 586-6910
E-mail: jdelucie@aol.com
(Lender, NY Support Group)

Elaine Guice
2009 Sallsbury Ave.
Baltimore, MD 21219

Alice Hammel (Has had pemphigus vulgaris for 30 years)
8196 Plumeria Ave.
Fair Oaks, CA 95628
(916) 961-5378

Sharon Hickey
RR #1, Box 1170
Little Meadow, PA 18830
717-623-2502

Elizabeth L. McLendon
4067 Mcrose Dr.
Martinez, GA 30907

Nicholas Moskowitz
2108 Providence Place,
Richmond, VA 23236-1856
804-276-0853
e-mail: sendervoice@firstsaga.com

Adene Strauss-Popper
827 Suffield Square
Lincolshire, IL 60069
(847) 940-1440
E-mail: MidwestPV@aol.com

Peggy Prober
5721 Hillcrest Lane
West Bend, WI 53095
414-629-9101

Venus Rastegar
P. O. Box 3653
Santa Monica, CA 90408-3653
E-mail: Venus_Rastegar@cox.Xerox.com

Miriam Schneidmill
340 Ayerigg Ave.
Passaic, NJ 07055
(201)471-2278 (evenings) fax #: 201-473-3408;
E-mail: zapot@cyberwar.com

Ann Sconyers
202 Maple St.
Enterprise, AL 36330
(w) (334) 347-8228
(h) (334) 347-0919

Steve Shapiro
PO Box 50335
Eugene, OR 97405-0977
(541) 686-5575 (7-9 pm pacific time)
E-Mail SS@shapiro@oregon.uoregon.edu
http://darkwing.uoregon.edu/~sshapiro/Pemphigus/
(focus on alternative therapies, natural healing & diet)

Gene Shuman
2487 Brentwood Rd.
Columbus, Ohio 43209

Charles A. Smith
15039 Chemise Creek Rd.
Ramona, CA 92065-5138
760-788-1625

Diane Warinner
220 River Bend Dr.
Newcastle, CA 91646
970-984-2885

David Zarett
E-Mail: zarett@aplemp.pap.jhu.edu

In Canada
Stuart Grossman
3219 Yonge St., Ste 316
Toronto, Ontario, M4N 2L3
Fax (416) 482-0821
E-mail: stuart@user.rose.com

In Australia
Julian Chan (09) 31 617 87
18 Stanbury Way
Booragoon WA 6154 Australia
E-mail: juchan@ca.com.au

In England
The Pemphigus Network
Flat C 26 St. Germans Rd.
London SE 23 1RJ

Liz Johnston
3 Hardwick Dr.
Copthorne, Shrewsbury SY 3 8UZ, England, U.K.
01743-344013.

In Israel
Victor Leikeman
972-4-822-3439
E-mail: Victorl@sctcom.co.il
http://www.geocities.com/hotsprings/7445/

In New Zealand
William Eaton
1/6 Ngaire Ave.
Auckland 1003 New Zealand
E-mail: William_Eaton@compuserve.com

Gloria Romano
P. O. Box 1051
Nelson, New Zealand

It is the policy of the Pemphigus Foundation to keep the names and addresses of our members confidential. However, some have expressed an interest in networking and making their names public. If you would like to be on this list, please contact the Foundation.
Dear Janet,

I feel as though I should tell my story of living with PV in order to encourage others. In August, 1983, at the age of 63, my family doctor put me in the hospital. The whites of my eyes were blood shot, my nose was dripping with blood, my mouth was all ulcerated and the genital area was covered with white spots. After 2 biopsies, the diagnosis was PV. Up until that time, I had never had any illnesses to speak of. I had taught school for 38 years and had 2 years of sick leave coming when I retired. The diagnostician in the hospital put me on 100 mg. of prednisone daily and gave me 5 mg. tablets of valium to take 3 times a day as needed. I only took one every night and that was to make me sleep for the 100 mg. made me hyper. The prednisone was gradually lowered to 60 mg. In 1986, I was told I would be in a wheelchair if the medicine wasn’t lowered. My next step was to go to Johns Hopkins Hospital here in Baltimore and be under the care of Dr. Grant Anhalt. Dr. Anhalt lowered the prednisone gradually while starting me on Cytoxan. Since cytoxan is a cancer drug, I lost my hair and wore a wig until my body adjusted to the new drug which Dr. Anhalt gradually reduced. (and the wig looked better than my own hair). At the present time, I am on 5 mg. of prednisone every other day and no Cytoxan. I have my fingers crossed for I have been in remission since August, of 1992 and have no signs of PV. I still see Dr. Anhalt every 6 months to have a blood test and urinalysis. I take Calcium and I am a big “skim milk drinker”. My last bone scan showed my bones to be in better shape than they were a few years back. I have had all the side effects of prednisone—cataracts removed from both eyes plus glaucoma (but I don’t need glasses except to drive); diabetes - but it is under control with 2 1/2 mg. of Glipizide a day, and naturally a gain in weight. Lowering the prednisone put me in a terrible state of depression. For 77 years old, I feel as though I’m in great shape today. My husband and I have a motor home and spend many months of the year on the road. I still ride my outdoor bike and walk a great deal. Of course, I give most of the credit for my well-being to Dr. Grant Anhalt who has been there for me for 14 years. I can talk to him personally on the phone, and can contact him immediately should I ever have any problems. To all PV “victims” - take a positive attitude. Believe me, having a good mental attitude helps. Smile!

Respectfully, Elaine Guice

Elaine Guice has been living with PV for over 12 years.

---

Special Note

In order to fit more information into the same amount of space, we have slightly shrunk the size of the type in this edition of the newsletter.

We acknowledge that some of our readers may have difficulty reading the smaller type size.

If you are experiencing difficulty reading the newsletter due to the smaller type size, please let us know by contacting the Foundation.
De Sauvages (1706-1767), in 1760, created the word pemphigus; but the bullous eruption which he described as pemphigus maior probably represented erythema multiforme since it was accompanied by a high continuous fever and only lasted about two weeks. The definition given by de Sauvages to pemphigus maior, that of a bullous eruption of short duration, was widely accepted and most conditions described as pemphigus in the following years were of that type.

It was Wichmann (1740-1802) who in a lecture delivered and published in 1791 in Erfurt, Germany gave the word pemphigus its present meaning, that of a chronic bullous disease. For bullous eruptions of short duration he suggested the term febris bullosa. He was the first to describe a case of pemphigus vulgaris with the diagnosis of pemphigus.

In subsequent years the concept of pemphigus underwent repeated changes and the term became at times almost synonymous with vesicular or bullous disease. Thus, von Martius arrived at the formidable number of ninety-seven types of pemphigus. However, Hebra, in 1860, re-established Wichmann's concept of pemphigus by stating that pemphigus always was a chronic disease and that there was no acute form of this disease. This point of view has prevailed up to the present time.

Pemphigus Vulgaris. The first case in medical literature to which the diagnosis of pemphigus vulgaris may possibly be applied is that described in 1681 by Koenig of Berne, Switzerland. It is more likely, however, that the disease was bullous pemphigoid since the patient survived. No diagnosis was given by Koenig to this case. He merely referred to the disease as being strange and surprising.

"A woman, twenty-five years of age, became afflicted with a very painful disease in 1678. Blisters the size of a hand and filled with a clear and burning water began to appear on different parts of the patient's body. Whenever the blisters disappeared in one area they reappeared in another. After eight months, the patient was able to leave the hospital. For nine months she remained well. Then blisters reappeared, but for only ten days."

The first author to report cases definitely identifiable as pemphigus vulgaris was Machride. In the second edition of his book, published in 1777, four instances of a bullous disease are described. He called the disease morbus vesicularis and stated that this disease had "hitherto entirely escaped the notice of observers." All four patients were males and were sixty years of age or older. The description was as follows:

"The blisters and vesicles were perpetually coming and going... Two patients died... The other two patients survived and recovered perfectly; one of them in ten months, while the other did not become entirely free of lesions until three years had elapsed... In the two patients who died the vesicles were filled with a bloody ichor, and the putrid ulcers that ensued were intolerably painful and livid, threatening mortification; but in the two patients who recovered the fluid contained in the blisters did not appear to be of so malignant a nature; being either a clear lymph or yellow serum and the sores which followed were more tractable."

It is likely that the two patients who died had pemphigus vulgaris, as suggested by the refusal of the denuded areas to heal; whereas the two patients who survived probably had bullous pemphigoid.

The second author to report a case of true pemphigus vulgaris was Wichmann in 1791. He applied to his case the diagnosis of pemphigus and thus was, as already stated, the...
first to report a case of pemphigus with the diagnosis of pemphigus. The characteristic features of the case, as described by Wichmann in remarkable detail, were: flaccid bullae in some areas and detachment of the epidermis without the formation of bullae in others, resulting in large denuded areas of the skin; severe involvement of the oral mucosa with extension to the vermilion border of the lips; and a fatal outcome.

“The patient, sixty years of age, when first seen had already been ill for nine months. Large areas of the skin were deprived of their epithelium. The whole right leg from the knee down to the dorsum of the feet and large parts of the back and chest were denuded, like a scald. The mucous membrane of the mouth exhibited many wounds so that the patient could take only liquids. The lower lip was covered with crusts, which made the lip unshapely thick. The blisters were not raised but flat, the height and size of an almond and always broke soon. Even when the epidermis was not detached by the underlying moisture, it was loose and wrinkled, so that it could be moved; and so gradually it separated itself and left denuded areas. The patient died after having had the disease for fifteen months.”

Pemphigus Foliaceus. Pemphigus foliaceus first was recognized as a special type of pemphigus by Cazenave in 1844. He stated in his textbook:

“The bullae follow each other so that there are no intervals. The bullae flow together, break as soon as they have formed and cover large areas with small scales, resembling the flakes of fine pastry. Attached to the skin at one side only, they are movable and have a peculiar foliaceus appearance, so remarkable that I believe that I should establish this type as a peculiar form of pemphigus.”

In the literature prior to Cazenave’s publication only two cases were found which probably were instances of pemphigus foliaceus. The first case in which a diagnosis of pemphigus foliaceus may be made was published by de la Motte in 1772 under the heading: “Sur une maladie singulière de l’épiderme.” At the time of de la Motte’s report the disease had persisted for four years.

“Blisters filled with serum arose... The epidermis thus lifted detached itself in large pieces. New fluid lifted another layer without forming more bullae. This layer fell off and a third layer rose. The unfortunate sufferer changed his epidermis from head to foot every twenty-four to forty-eight hours. The whole body had the redness of a lobster. Every morning one removed from the bed sheet handfuls of pieces of epidermis. One may well believe that one could have filled two barrels with scales had one collected them since the beginning of the malady.”

The Nikolsky sign was first described by Nikolsky as a characteristic sign of pemphigus foliaceus in a thesis on that disease published in 1895. In his textbook published in 1927 Nikolsky stated:

“The skin in pemphigus foliaceus shows a weakened coherence among its layers (especially between the horny and granular layers), even in places between lesions on the seemingly unaffected skin. This characteristic behavior of the skin may be demonstrated by two methods: First, by pulling the ruptured wall of a blister one can detach the horny layer for a long distance, even on a seemingly healthy skin; and second, by slightly rubbing the epidermis between blisters one exposes the moist surface of the granular layer.”

Pemphigus Vegetans. This form of pemphigus was first described by Neumann in 1886. He stated in his original communication:

“When the wall of the bullae bursts their base becomes raised in the course of a few days and shows warty, densely set granulations. These areas of granulation increase in size and spread seriginously. The seriginous patches are limited by a shaggy undermined epidermis.”

The literature before Neumann’s report seems to contain descriptions of only two cases of pemphigus vegetans. The first case was described by Hebra in 1860 under the name of pemphigus diphthericus.

“By the bursting of the bullae the dermis forming the base was exposed, and on its surface was found a whitish yellow granulating substance so firmly attached that it could not be detached.”

All current textbooks recognize the three forms of pemphigus just mentioned: pemphigus vulgaris, pemphigus foliaceus and pemphigus vegetans. Two additional variants of pemphigus were described in the early part of this century: benign mucosal pemphigus by Thost in 1911, and pemphigus erythematosus by Senear and Usher in 1926.
THE NATIONAL PEMPHIGUS FOUNDATION
Awareness Program
T-SHIRT AND BUMPER STICKER
ORDER FORM

NAME ____________________________________________

ADDRESS _______________________________________

CITY __________________________ STATE _______ ZIP CODE_____

PHONE ________________________________

T-SHIRTS - QTY. ________ SIZE: Adult S M L XL XXL
$15.00 ea. includes shipping (circle)

Child S M L (circle)

BUMPER STICKERS - QTY. ________
$2.50 ea. includes shipping

PAYMENT - CHECK OR MONEY ORDER (circle)

TOTAL AMOUNT ENCLOSED $__________

THANK YOU FOR YOUR SUPPORT!
**Answers & Questions**

Before trying any therapies or using or not using any medications, please consult with your personal physician first for his/her recommendations and/or opinion.

**What is the best regimen to use to combat yeast?**

Dr. Alex J. Sheehan, DC, DACBN, CCN from Staten Island, recommends L. acidophilus and b. bifidus - 1/2 teaspoon twice per day away from medication, and nystatin USP - 1/8 teaspoon in water twice per day. Nystatin is not absorbed into the systemic circulation. Dr. Sheehan writes, “therefore, it is used to kill intestinal yeast (yeast in the intestines can leak toxins into the blood and cause worsening of symptoms in many different conditions). Yeast (Candida) have been isolated in numerous studies as a factor in initiation and progression of autoimmune diseases. Meaning that you may receive a double benefit from taking nystatin.” Dr. Sheehan also recommends the following supplements for any autoimmune disease: fish oil - 3 caps with each meal, vitamin E - 400 IU two times per day, multivitamin/mineral vitamin, C - 1000 mg twice per day, selenium 200 mcg.

My stomach often gets upset with my medications. Do you have any natural suggestions?

Some of our patients have found ginger helpful in relieving stomach discomfort. Most supermarkets will carry crystallized ginger which is good, and health food stores will carry ginger tea.

**What side effects can we get from drinking coffee and eating chocolate?**

We suggest you indulge in very small amounts of caffeine and chocolate. Chocolate contains caffeine. Caffeine is a stimulant. Since prednisone can cause similar stimulating side effects, the addition of caffeine and chocolate can possibly make you very uncomfortable. In addition, caffeine and chocolate stress the adrenal glands and contribute to the development of osteoporosis by enhancing bone demineralization.

**What are good ways to replenish potassium in the body?**

Of course there are supplements, but from one study, taking potassium bicarbonate 1/4 teaspoon twice a day, might not only give you good added potassium, but might also help reduce high blood pressure.

---

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