What is Pemphigus Vulgaris?

Pemphigus Vulgaris is an autoimmune blistering disorder of the skin and mucous membranes. The immune system of an affected individual produces antibodies which erroneously perceive the skin as foreign and attack it, producing burn-like lesions that will not heal. In some cases, these sores can cover a significant area of skin. There is no cure. Pemphigus vulgaris is a rare but very serious, sometimes life-threatening disease.

Treatment

Although there is no cure, PV can be treated successfully with corticosteroid medications (i.e., prednisone). Chemotherapeutic (i.e., immunosuppressive) drugs, such as Imuran and Cytoxan, are also administered to offset the side effects of prednisone. These drugs can have serious side effects, but there is some evidence that treatment is easier in the early stages of the disease.

Symptoms

Very often, PV lesions first appear in the mouth and are frequently mis-diagnosed as simple mouth sores. The tissue on the gums may tear and heal and tear again. A simple ulcerated sore may persist and multiply. An itchy sore patch that is unaffected by application of conventional topical medication may appear on other parts of the body including the genitals. Another patch will appear, and then another, and another. More time than not, PV is the last disease considered during diagnosis.

The National PV Foundation’s Goals and Objectives

The purposes and goals of the Foundation are:

- to provide information on pemphigus to pemphigus sufferers, their families, friends and the general public;
- to provide PV sufferers and their families with general support and counseling;
- to inform those afflicted as to where the specialists are in their area;
- to provide information on how to handle the various side effects of drug therapy;
- to inform as to what and where the research on PV is being performed;
- to raise funds to promote and support research into the causes, diagnosis, treatment, and cure of PV;
- to provide the medical community with the knowledge it needs to properly diagnose before extreme measures are needed.
President's Message
by Janet Lehne

Welcome! This is the first issue of The National Pemphigus Vulgaris Newsletter.

Eleven years ago, when I first contracted pemphigus vulgaris, I asked my doctor if she knew of any support groups for PV. Because I was her only patient with PV, she did not know and suggested I call around. So, I did just that. I called various Foundations representing other autoimmune diseases. I called the American Academy of Dermatology. I was not successful. Eventually, after writing letters to various doctors around the country, I hooked up with a few people.

At that time, I thought I would try to start the Foundation. Unfortunately, I was not prepared for the enormity of the task, and found it all quite overwhelming; so I gave it up; though I continued to look for others and gather information. During that first outbreak, I remained on medication for three years. After a two year remission, a short two month relapse and another remission which lasted five years, I relapsed again last April.

When I could no longer ignore the signs, I went to the doctor and went back on prednisone. At that juncture, I decided again to attempt to start a Foundation. This time though, I was older, more familiar with the illness, and definitely more determined. One of my first objectives was to find a physician willing to support the Foundation. I wrote to Dr. Grant Anhalt from Johns Hopkins and he enthusiastically agreed to let the Foundation use his name and reputation. Thus, the Foundation was created; and, on October 14, 1994, the Foundation was recognized by the Internal Revenue Service as a non-profit, tax-exempt organization.

DONORS

Thanks go to those who gave their personal and financial support in order for the Foundation to go forward. Special thanks go to Dr. Grant Anhalt, Dr. Jean Claude-Bystyn, Jean Barish, and Arthur Rosenbaum for their generous contributions. Also thanks go to Sandra Feldstein, Justine Lehne, Helen Segall, and Omeed Memar for their tireless efforts on behalf of the Foundation.

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Board of Directors

The Foundation would like to introduce our Board of Directors.

President and Executive Director: Ms. Janet S. Lehne. Ms. Lehne is a 49 year old single mother. She has had 25 years of experience running offices for both non-profit and profit organizations. She has had pemphigus vulgaris for eleven years.

Vice-President: Ms. Kay Greisen. Ms. Greisen is a molecular biologist working for a pharmaceutical company in Alameda, California.

Vice-President in Charge of Scientific Affair: Dr. Grant Anhalt. Dr. Anhalt has been working with pemphigus patients and on finding better treatments for sufferers for the past twenty years. He is the Head of the Department of Dermatology at Johns Hopkins in Baltimore, Maryland.

Secretary: Ms. Sharon Mortz. Ms. Mortz is a legal secretary working in San Francisco, California.

Treasurer: Mr. Jonathan W. Segall. Mr. Segall is a Regional Director of Marketing for a company in San Francisco, California.

On July 31, 1994, our Board of Directors had its first meeting in San Francisco, California. The Board voted on and adopted the Foundation’s Bylaws and set an agenda for the future.

We are also looking to expand our Board to include various members of the pemphigus community and/or unaffected people who would be interested and involved directly with all aspects of the disease; people who can give their time and dedication to the organization.

The Foundation’s First Project

To date, definitive statistics on the incidence and prevalence of pemphigus vulgaris are not available. The disease is known to affect people across racial and cultural lines. Although the onset usually occurs in middle-aged and older adults, pemphigus vulgaris has also been documented, although rare, in young adults and children. The Foundation is in the process of surveying Dermatologists across the USA to determine who gets the disease. We are asking the age, gender, and ethnicity of the patients.

We are sending out approximately 7600 survey letters. The first thousand have been disseminated across the country and answers are beginning to come in. We ask those physicians who would like our brochure to make a small donation to defray the cost of printing and postage.

We have begun receiving responses back and recording the data. When the results are in, we will publish them in the newsletter.

Future Projects

Future projects will focus on raising money for pertinent research, organizing fund raising events, establishing a physician referral service, creating support groups around the country, and making further contacts worldwide.
Research

The Target in Pemphigus Vulgaris

Pemphigus Vulgaris (PV) is an autoimmune disease, whereby antibodies target a component of keratinocytes, leading to blister formation on skin and mucous membranes (e.g., oral cavity). Recently, our knowledge about the target (autoantigen, pemphigus vulgaris antigen, PVA) in PV has been greatly expanded. Dr. Amagai, working with Dr. Stanley, at the National Cancer Institute in Bethesda, Maryland, has successfully identified the DNA sequence of PVA, and shown that it belongs to a family of adhesion molecules called cadherins. Generally, cadherins are protein molecules that are situated in the cell membrane, with a segment exposed on the exterior of the cell and a segment within the cell. Moreover, cadherins are commonly involved in maintaining the structure of organs, including the skin. The discovery that PVA is a cadherin gives tremendous insight into the pathology of PV, where keratinocytes come apart, leaving a blister.

Using recombinant DNA technology, Dr. Amagai produced the portion of PVA exposed on the exterior of keratinocytes and which is thought to mediate adhesiveness. This segment was specifically recognized by serum from all 35 PV patients tested. Next, the PVA segment was used to clear patient serum of antibodies that bind monkey esophagus (the source of PVA, and a diagnostic test commonly used). The PVA segment could clear to differing degrees these antibodies from 16/17 patient sera. More importantly, this segment could clear three patient serum of blister-causing antibodies, indicating that PVA is the target antigen in PV. We have independently confirmed

Research (con’t)

Dr. Amagai’s findings. Therefore, based on the few patients tested, antibodies directed against PVA are responsible for causing blisters in PV. The exact mechanism of blister formation subsequent to antibody binding is not known. We are currently characterizing the role of PVA in PV.

The availability of PVA opens doors to better diagnostic and therapeutic measures in the future. Possibly, PVA could be used to specifically remove blister-causing antibodies from patient serum (specific plasmapheresis). PVA could also be used to develop an animal model for PV, allowing the study of PV before, during, and after disease initiation. This would allow specific immunotherapies based on interfering with the different stages of an autoimmune reaction. Research in PV is active and has yielded precious information regarding the target in PV. Continued research is necessary to apply basic scientific findings to therapy and patient care.

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A Look Toward Our Future

The National Pemphigus Vulgaris Foundation is in its infancy. Those involved have worked very hard to see that the Foundation succeed. We are asking for your help in making sure that we can continue to bring you all the news on pemphigus vulgaris and its related subjects.

In the next issue we plan to include more research news from Israel and possibly India. We are also planning on including a section called Health Tips.

We are open to ideas and suggestions as to what we should include in future newsletters. Please contact us with any of your thought on this matter. Send your ideas to:

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