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For some people, the end of Summer means the start of school. Here at the office “Back to School” means helpful happy interns. Several interns from Sacramento State University and Cristo Rey High School (Sacramento) are working on website articles, informational materials, the Awareness Campaign, and more. Our Sac State interns are in health-related programs and our high school interns are part of a Work-Study program. The work done by them saves the IPPF thousands of dollars each year while giving them real-world experiences.

Volunteerism is on the rise! Daphna Smolka is working with us on a P/P friendly cookbook that will be also be a P/P Community effort. Tina Lehne is spearheading the Awareness Ambassador effort and preparing the program’s requirements and materials. Marketing guru Edie DeVine is helping with our public messaging and press releases. Dr. Maulik Dhandha is working on a paper reporting on the diagnostic delays of P/P we hope to publish in an academic journal.

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William Zrnchik, MBA, MNM
IPPF Chief Executive Officer
will@pemphigus.org

The Awareness Campaign has a new look, more help, and a catchy slogan. Kate Frantz keeps you up to date starting on page 6. Clinical psychologist and PV patient Terry Wolinsky McDonald explains the Nuts & Bolts of Depression (p. 7). The discovery of VH1-46 is the topic of two articles (pp. 8 and 9). Two P/P ex-

And speaking of journals, members of our Medical Advisory Board joined 30 other physicians in finalizing an MMP consensus statement providing clearer definitions and outcome measures for accurate and reproducible definitions for disease extent, activity, outcome measures, end points and therapeutic response. Thank you Prof. Dedee Murrell and Dr. Victoria Werth for leading this effort.

This issue of the Quarterly is another great one! PV patient Martha Cusick was so happy with the help she received from her Peer Health Coach, she set a goal to fundraise for research and awareness (p. 4). And what do you do when you need a cancer treatment, but getting it will cause a severe flare? Read Joan Blender Ominsky’s story on page 15.

Experts discuss the importance of measuring patient quality of life (p. 11). And we have another delicious Vicky Starr recipe on page 19!

Lastly, I hope to see you in New York for our 2015 Patient Conference. The Committee is busy finalizing the date (end of April) and venue (near Central Park). Keep an eye on your mailbox and inbox for more information in the coming months! I promise this event will be BIG!

“I really can’t find the words to once again, thank you, for your good work!”

No, thank you for supporting us and our important work.

If you have a question for the IPPF, want to comment on a previous article, or recognize someone in our community, contact us and we’ll get you an answer or response. . . and maybe use it in a future issue of the Quarterly!

Quarterly related: editor@pemphigus.org
Foundation related: info@pemphigus.org
Letters to the Editor: editor@pemphigus.org
Disease, Treatment, Lifestyle: phc@pemphigus.org

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I was looking forward to enjoying my four grand-children when I retired. However, in 2006 my life took a dramatic change. After the death of my ex-husband, I returned to my home town to care for my 90-year-old mother. It was the furthest place from where I wanted to be.

My health was deteriorating. I started experiencing new pain in my legs and muscles. I thought I had MS. I became depressed, tired, and I discovered sores on my face, mouth, and scalp that lasted six weeks. Of course, I went to my internal medical doctor who ran many tests and began treating me for depression.

He recommended modifying my diet. At first acidic foods were eliminated to see if that was the cause of the sores. I was given glucocorticoid spray that did nothing (and it was painful to administer). I saw a dermatologist who had no answers and put me on Lysine tablets. I went to my dentist to see if he knew what the cause was. Meanwhile, I waited six more weeks to get another appointment with my dermatologist.

All of these visits happened over six years and I was still undiagnosed — and could no longer eat. I returned once more to the dermatologist who ran a diagnostic test from a skin scraping and found PV. She gave me a small brochure and a prescription for niacinamide and the dreaded prednisone (10 mg twice daily, then 20...then 30 mg for nine months but I was slowly taken off due to complications). Nothing was explained to me about the disease. I went to the Internet and couldn’t believe what I read. I had always been healthy ... why me?

I saw a young rheumatologist who worked with medications suggested for PV: azathioprine, (Dapsone® 50 mg), mycophenolate (1000 mg) and prednisone (30 mg). The headaches from these medications were so bad for me that I spent most of my time in bed. The medicine was hard to keep down and I was constipated daily.

I thank God my daughter found the IPPF online. They knew what was happening to my body. They answered my questions. They helped with vitamins. They suggested specific foods to avoid. The Peer Health Coaches felt like my only source of help. They found me a doctor near my home at University of Kansas Medical Center.

I was put on doxycycline (200 mg daily) in December 2013 and still take it. The breakouts are now more controlled and I continue to work on my health with my support system of professionals in place.

I am so thankful I went to the 2014 IPPF Patient Conference and was able to meet others living with P/P. There, and through the Quarterly, I’ve realized there needs to be more awareness for P/P. Just because I “look alright” externally doesn’t mean that I really am.

I have recently made a goal to raise money for research and awareness so I can give back to the IPPF and help others who have to go through what I did. I will always do what I can to help!

Martha Cusick lives in Wichita, KS, where she is retired after twenty-five years in Property Management.
A New Look

William Zrnchik, MBA, MNM

Fall is a time most clothing designers live for. They spend months creating new styles to be modeled on runways in New York, Milan, and Paris hoping to have the “must have” look of next Spring or Summer.

This Fall we present a new look from Sacramento tech-firm Uptown Studios (up-townstudios.net) for the IPPF that will be on computers, tablets, and phones around the world. After months of sketches, wire-frames, and coding, I am very excited to announce our new website: www.pemphigus.org.

The new site is easier to navigate and features resources and support at your fingertips. The “responsive” design keeps its look and feel no matter what screen size. We added accessibility features like variable text sizing and a high-contrast display. Our social media links, search box, and donation button are conveniently located at the top of each page. Our home page features a testimonial section. Read what others have to say about the IPPF. You can also submit your own story to be shared on our site!

A big part of our mission is support. In addition to the Email Discussion Group, RareConnect community, and social media sites, we added Desk.com support.

Our Patient Support team can now provide fast, awesome customer service on a modern, flexible platform over various channels. Patients and caregivers can find answers in the self-service portal using existing FAQs, email questions to our PHCs, or post to the general community. And when they are online, you can chat live with a PHC!

I am also excited about the Awareness Campaign portal currently being developed. It will contain resources and information for dental professionals and Awareness Ambassadors. Our goal is to keep P/P on the RADAR of dentists and provide them with tools and training needed to reduce P/P diagnosis time and increase your quality of life.
The average P/P patient sees five doctors over 10 months to obtain a diagnosis. The IPPF Awareness Campaign strives to change this statistic by reducing the amount of time it takes a patient to receive a pemphigus vulgaris (PV) or mucous membrane pemphigoid (MMP) diagnosis.

We are excited to share the Awareness Campaign’s new logo and slogan, Put it on Your RADAR. The IPPF will encourage dental professionals to put PV and MMP on their radar. To do this, we are using different methods to increase awareness, such as conference presentations, community outreach, and dental expert reviewed materials.

**The Dental Detective**

The IPPF partnered with four other autoimmune disease foundations and submitted a joint conference presentation proposal. We are happy to announce our proposal was accepted and will be presented at the American Dental Association’s Annual Meeting on October 9, 2014, in San Antonio, TX. This was a very competitive process and we are thrilled to use this opportunity to spread awareness.

Dr. Vidya Sankar, DMD, MHS, and Sjögren’s Syndrome Foundation Board Member, will present on “The Dental Detective: Investigating Autoimmune Diseases.” This is a wonderful way to get information on PV and MMP, as well as other autoimmune diseases, out to an entire dental team.

**Awareness Ambassadors**

Tina Lehne, Volunteer Awareness Ambassador Coordinator, is working hard to bring the Awareness Ambassador program to life. The first set of activities will include outreach to dental professionals. This may include one-on-one meetings or presentations to dental classes or societies.

**CONTINUED ON PAGE 14...**
One of the first things to remember about illnesses and the side effects of medications is the effects of illness are not just physical. There is an emotional component as well.

For example, the prednisone roller coaster is both physical and emotional. The ups and downs often have patterns and triggers, and these are not always predictable. The mere fact of having an illness can lead to depression, with or without side effects from medications.

Psychologists have been called “an angry bunch of shrinks” (Newsweek, December 2013) because of their collective response to new and unsettling upcoming changes in current diagnostic criteria and standards. The Diagnostic and Statistical Manual (DSM-IV) of the American Psychological Association has been the “bible” of the psychiatric profession for more than a decade, with the new version (DSM-V) going into effect in October 2015. The physicians’ ICD-10 (or International Statistical Classification of Diseases and Related Health Problems) will also be issued at that time.

In this article I will review some current diagnoses and criteria related to depression. With the aforementioned changes more than a year away, now is a good time to go over the diagnostic criteria for depression as outlined by the DSM and ICD standards. Lenore Sawyer Radloff’s Screening Test for Depression (see p. 17) can be used to monitor your own symptoms and patterns.

One mood disorder in the current DSM-IV is simply called “Mood Disorder Due to ____________.” The blank is filled in with a specific general medical condition, such as pemphigus vulgaris. The diagnosis may develop into a clinical depression over time, which has a different etiology. The diagnostic criteria for these generic mood disorders include:

- A prominent and persistent disturbance in mood predominates in the clinical picture and is characterized by either (or both) of the following: Depressed mood or markedly diminished interest or pleasure in all.
Our immune system is a killing machine. It consists of various types of specialized cells and proteins that function to destroy invaders and "non-self" or mutated "self" proteins, such as those that come from viruses, bacteria, and cancer cells. In the autoimmune diseases such as the P/P diseases, this mechanism has gone awry and the immune system actually attacks its own cells.

In P/P patients, antibodies generated by B cells of the immune system block the function of desmoglein proteins Dsg1 and Dsg3 known to be important in binding together keratinocytes of the skin and mucous membranes, but it is not known how the rogue antibodies are generated by the immune system, how they escape the quality control mechanisms in place that only allow B cells with non-"self" specificities to survive, and why P/P patients are so rare.

New research led by Dr. Aimee Payne in the Department of Dermatology at the University of Pennsylvania (Nature Communications, http://www.nature.com/ncomms/2014/140619/ncomms5167/abs/ncomms5167.html) helps us begin to understand why.

In previous work, Dr. Payne and colleagues have identified antibodies that recognize Dsg1 and Dsg3 (so-called anti-Dsg1 and anti-Dsg3 antibodies) and have also identified regions of those antibodies that are important for the ability of those antibodies to be pathogenic – that is, to recognize their Dsg targets in pemphigus vulgaris (PV) and pemphigus foliaceus (PF) and to disrupt their function. To extend this work and to better understand how PV autoantibodies arise, Dr. Payne and colleagues have performed a similar analysis of PV patients.

PV patients can present as either mucosal-dominant, where only the mucous membranes are affected or as mucocutaneous, affecting both the mucous membranes and the skin. Almost all mucosal-dominant PV patients have anti-Dsg3 autoantibodies, while the mucocutaneous patients have anti-Dsg3 autoantibodies as well as anti-Dsg1 autoantibodies. Since it is thought that Dsg1 and Dsg3 can compensate for each other's function, the presence of functional Dsg1 in the skin in the presence of anti-Dsg3 autoantibodies can explain why mucosal-dominant patients do not have skin lesions.

The authors first isolated the full antibody repertoire from four different untreated PV patients, all with mucocutaneous disease. They isolated and characterized these in a multistep process that ultimately allowed them to determine the amino acid compositions (by cloning and DNA sequencing methods) of the patient's PV antibodies. This led to the assignment of six unique antibodies from Patient 1 and five additional
The discovery of the VH1-46 autoantibody linkage to pemphigus vulgaris (PV) is an exciting one for P/P patients around the world. Its discovery is described in a recent research paper published in *Nature Communications*, and is also the subject of this issue's Research Highlight article (see p. 8).

The author’s discovery shows PV patients have VH1-46 autoantibodies. Although this was the most common autoantibody variant, the authors found other autoantibodies appeared in some PV patients and not in other patients.

**What this means for P/P patients**

This discovery is a great stepping stone for future research. The key takeaway is researchers are closer to understanding PV.

However, because the existence of the autoantibodies is correlative (not yet shown to be causative), we still do not know what specifically causes P/P – or what cures it. Even if researchers can mutate this gene, they still may not find a cure for PV. But the work they do could help future researchers accomplish that.

The next step is to determine if VH1-46 appears in people without PV as well. If it does not appear in non-PV patients, then they will study why it is a common denominator in PV patients. This research focuses only on PV and not other P/P diseases such as MMP or PF.

**What this means for the IPPF**

Like most P/P research, new findings give our community members a perfect opportunity to get involved.

We encourage patients, caregivers, family members, and friends to write their elected officials. Tell them P/P needs more research funding. The National Institutes of Health (NIH) is one source for some of our researcher’s funding. With increased NIH funding, P/P researchers could be one step closer to our goal: a cure for P/P.

You can find your congressional representatives’ contact information at [www.govtrack.us/congress/members](http://www.govtrack.us/congress/members). If you have questions about this study, or about how to contact your elected officials, contact me at noelle@pemphigus.org.

Noelle Madsen is the IPPF Patient Services Coordinator and lives in Sacramento, CA. She is dedicated to providing support and education to those affected by P/P. She is a new contributor to the *Quarterly* and can be reached at noelle@pemphigus.org.
Quality of life (QOL) has been defined as individuals’ “perception of their position in life, in the context of the cultural and value systems in which they live and in relation to their goals, expectations, standards, and concerns.” Quality of life impacts everything we do, so it’s important that physician evaluate the QOL of patients.

For many dermatological conditions, measuring QOL provides a way to evaluate the impact of a patient’s disease on their well-being. Treatment can also affect patients’ QOL, and impaired QOL can have important implications for treatment adherence and perceived quality of care.

Quantifying the QOL impact of skin conditions is a relatively new effort, stemming from a movement in medical science to capture the impact of a disease on the patient. Questionnaires help measure the burden of the disease and the treatments.

Generic QOL measures have been used by the medical community to measure the QOL impact of general medical problems. For skin conditions, investigators have used ge-
or almost all activities. Elevated, expansive, or irritable mood.

- There is evidence from the history, physical examination, or laboratory findings that the disturbance is the direct physiological consequence of a general medical condition.
- The disturbance is not better accounted for by another mental disorder so as to distinguish this general mood disorder from “Adjustment Disorder With Depressed Mood” in response to the stress of having a general medical condition.
- The disturbance does not occur exclusively during the course of a delirium.
- The symptoms cause clinically significant distress or impairment in social, occupational, or important areas of functioning.

Common symptoms of depression look different in each circumstance and with each individual. A diagnosis may be given if there is a prominent and persistent disturbance in mood that predominates in the clinical picture, and it is further characterized by five or more of the following:

- Persistent feelings of sadness
- Difficulty sleeping or excessive sleeping
- Poor or increased appetite
- Weight loss or weight gain
- Anxiety, restlessness and agitation
- Inertia: feeling “slowed down” or low in energy
- Tearfulness or an inability to cry
- Difficulty concentrating, remembering, or making decisions
- Loss of interest in sex and other normal activities
- Social withdrawal
- Difficulty functioning at work, at home and/or in social situations
- Irritability
- Suicidal thoughts or passive thoughts of death.

Ill people will often try to hide their symptoms until they lose the energy necessary to keep up the act. After all, the last thing most people want is more prescription medications or treatments. This is more so when their bodies have already “betrayed” them and medications are necessary just to not get sicker. It is important to understand what is happening emotionally and to get a proper diagnosis. With a diagnosis can come appropriate treatment.

The simple 20-question screening test for depression can be self-administered. I often recommend that anyone who is concerned or has symptoms they do not understand make copies and re-test themselves roughly every two weeks. This particular screen looks at the feelings and thoughts for the previous seven days, so you could use it weekly if you wanted to.

I often use this tool as a handout at presentations. Patients (and caregivers) usually come up to me and express surprise at how many statements they have endorsed. Many have no idea these particular feelings and thoughts were actually signs of depression. As I noted above, with diagnosis there is treatment.

CONTINUED ON PAGE 17...
...continued from QUALITY OF LIFE, page 11

Generic QOL measures such as the Short Form (SF)-36. These generic tools use questions about physical functioning (limitations in performing physical activities related to work and other daily activities as a result of physical health); bodily pain (severity and limitations due to pain); vitality (feeling tired vs energetic); social functioning (interference with normal social activities caused by physical or emotional problems); and emotional functioning (limitations in work and other daily activities as a result of emotional problems). Generic measures have been tested rigorously for psychometric properties. However, they do not necessarily capture issues specific to patients with skin disease.

Many QOL measures specifically evaluating the relation between skin and QOL have been developed. One of the first was the Dermatology Life Quality Index (DLQI). Among the 11 skin-specific QOL measures, the Skindex-29 is one of the more recent. It is a valid and reliable tool designed to measure health-related QOL in patients with skin conditions. It has three scales that assess symptoms, functioning, and emotional state.

Even more sensitive than skin-specific QOL tools are disease-specific measures. These are designed to capture issues related to the disease that even skin-specific instruments cannot. These disease-specific measures, such as the Autoimmune Bullous Disease QOL (ABQOL) measure, are generally even more sensitive to changes in disease status. The Ro-saQOL (Nicholson et al., 2007), a published rosacea-specific quality of life instrument, captures such topics as avoiding certain foods or drinks and frequency of flushing. These issues are not captured in skin-specific quality of life measures because they do not pertain to all skin conditions. As a result, disease-specific measures are generally even more sensitive to changes in disease status.

Autoimmune Bullous Diseases (ABD) are rare but potentially serious immunobullous diseases of the skin and mucous membranes. They often require long-term immunosuppressive therapy. Physical symptoms such as bullae formation (blisters), pain, itching, and functional limitations can significantly affect a patient’s QOL.

The few studies that have investigated the quality of life of P/P patients report a strong impact on physical and emotional status. In 2013 the ABQOL was introduced by Dr. Sebaratnam and Dr. Dedee Murrell (Br J Dermatol 2013, doi: 10.1111/bjd.12623). This ABD tool is a valid and reliable patient-based measure. It is composed of 17 items: 9 related to the psychosocial subscale (embarrassment, depression, anxiety, family/friends, sexual activity, relationships, social life, work and study, discrimination), 5 on the symptom subscale (pain, itching, healing, clothing changes and bathing/showering) and 3 for mucosal involvement (mouth pain, gingival bleeding, food avoidance).

Treatments for ABDs have a significant risk of medical complications and impact on QOL due to treatment. It is hard to differentiate the impact from disease burden or the effects of treatment. Treatment of autoimmune blistering disease frequently requires long-term therapy. After many weeks, or even months, the QOL depends on the disease and the treatment. For this reason, experts in bullous disease refined the ABQOL, selecting only those questions pertaining to treatment effects. They recently introduced a pilot Treatment of Autoimmune Bullous Disease Quality of Life (TABQOL) questionnaire that measures treatment burden in ABD separately from QOL related to the disease itself (Br J Dermatol. 2013, doi: 10.1111/bjd.12623.). Three of the 17 questions in the TABQOL are (1) do you take many medications for your blisters disease? (2) does the treatment for your blisters disease result in you feeling bloated? (3) does the treatment for your blisters disease make it difficult to walk?

Dr. Finlay recently reported yet a third dimension of skin disease burden, the long-term impact, including impact on friends and family. Examining the long-term and family impact of ABD, in addition to the patient’s QOL, also needs evaluation.

Many QOL measures exist, both generic and specific. More scientific studies are needed to better understand which tools are the best for evaluating QOL in ABD. The impact of blistering diseases on QOL is multifaceted, and may create significant burden. Integrating formal QOL evaluation into clinical evaluation of patients helps assess disease severity and mapping of disease trajectory, and can capture outcomes of therapeutic intervention relevant to the patient. It is important to also evaluate the QOL of ABD patients to globally evaluate the impact of the disease and treatment on patients. The questionnaires are completed by patients and thus provide feedback directly from the patients.
BANDING TOGETHER

Ambassadors sign up for a one-year commitment and receive training prior to engaging in awareness activities. If you are interested in becoming an Ambassador, contact awareness@pemphigus.org.

Dental Advisory Council

We are pleased to introduce the formation of the IPPF Dental Advisory Council (DAC). The DAC provides critical review of dental materials related to the Awareness Campaign. It is comprised of both dental professionals and students. We are very lucky to have such an expert panel devoted to PV and MMP awareness.

And the Oscar goes to...

Becky Strong, IPPF Patient Educator (pictured below), is the first recipient of the IPPF Awareness Award, appropriately shaped like an Oscar for her starring role in the Patient Awareness Video (release date coming soon). Becky has devoted countless hours to P/P awareness by presenting to dental schools and sharing her diagnosis story to help reduce diagnostic delays. Thank you, Becky!

Funding Announcement

The IPPF is happy to announce continued funding from the Sy Syms Foundation (www.sysymsfoundation.org). The Sy Syms Foundation awarded the IPPF’s Awareness Campaign with a check for $75,000. These funds will go a long way in our efforts to spread awareness and reduce diagnostic delays for all pemphigus and pemphigoid patients.

Thank you to the Sy Syms Foundation!

“As a person with PV since I was 15 years old, I’m so pleased to be part of this campaign.”

Marcy Syms
President of the Sy Syms Foundation

The IPPF encourages our Community to get involved with the Awareness Campaign. If you are interested in learning more about the campaign or getting involved, contact awareness@pemphigus.org.

Kate E. Frantz, MPH, CTTS, is the Awareness Program Manager at the IPPF living in Dixon, CA. She is a contributor to the Quarterly newsletter in her “Awareness and You” column. Kate can be reached at awareness@pemphigus.org.

I would like to thank the Academy and each one of you for giving me the strength and courage to get out there and tell my story to wide-eyed dental students across this country. If it weren’t for you, your stories that you shared with me, and your support, I know none of this would be possible. Also, a loud shout out to the video crew for making me look good and for all of their patience with me and my family.

—Becky Strong
Cancer & Pemphigus: Is Radiation Possible?

Joan Blender Ominsky

As a pemphigus patient for more than 12 years, I kept my disease under control with medications. It was a chronic annoyance, but I thought the worst was over.

Imagine my horror when a small, cancerous lump on my breast was discovered during a routine mammogram. I was assured the tumor was small and I had nothing to worry about. The surgeon performed a lumpectomy. Lab analysis found the tumor to be very fast growing, and further steps were needed. My oncologist suggested chemotherapy and radiation.

Then came the hitch. The radiation oncologist refused to administer the radiation fearing severe blistering would occur under the breast due to the pemphigus. I was devastated. I knew long-term survival depended in large part on having the radiation treatments.

I asked my dermatologist, Dr. Timothy Berger, for his opinion. His very words were: “You don’t fool around with cancer. You need the radiation. Find another doctor who will perform it.” Dr. Berger agreed to work with a radiation oncologist to address any skin problems that might arise.

Relieved, I found a new radiation oncologist, Dr. Barbara Fowble, who agreed to proceed, citing the 70% 5-year cure rate for patients undergoing radiation. I heaved a long sigh of relief. Events proceeded. First I underwent three months of chemo with cytoxan and Taxotere®, followed by five weeks of daily radiation treatments.

These treatments went swimmingly during the first four weeks. Then, during week five, signs of trouble began. Blistering occurred and none of the regular creams would clear it. Dr. Fowble continued with the treatments.

She experimented with various creams and cures. At the end of five weeks we found the perfect treatment for me: Aquacel® Ag. My wounds completely healed. In a later visit, she told me that Mepilex® Ag also works in other blistering diseases.

The message I took from this is not a new one: Don’t easily take no for an answer when confronted with cancer and the need for radiation therapy. Pemphigus does not exclude radiation. However, a patient needs to find a radiation oncologist willing to take some risk and to work with a dermatologist should trouble arise. For physicians, it is important to know radiation and pemphigus don’t have to be mutually exclusive, and tools are available should blistering occur during treatment.

A happy side effect of this story is two years after this saga, my pemphigus titers are much lower than before. They dropped after chemo treatments with cytoxan. According to my dermatologist, Cytoxan is known to lower titers in many patients and used in India and Pakistan to treat pemphigus, but considered by American doctors as too toxic for pemphigus.

Dr. Fowble and Dr. Berger at UCSF School of Medicine told me they would be happy to consult with other physicians about their experiences with cancer, pemphigus, and radiation.

Joan Blender Ominsky’s pemphigus began after Sept. 11, 2001, when her daughter and nephew were working in New York City. It was a difficult case to control and was life-threatening, only controlled after two years of IVIG treatments. Joan worked as a publicist and wire service reporter on music, dance, and opera in the San Francisco Bay area. She is married to Dr. Steven Ominsky, a radiologist, and is the mother of three children and has three grandchildren.

www.pemphigus.org
Both houses of the United States Congress have introduced legislation to establish a fund to provide expanded and sustained national investment in bio-medical research. Rep. Anna G. Eshoo (D-Calif.) Introduced The America Helping Encourage Advancements in Lifesaving Science or the America HEALS Act (H.R. 4384) in April 2014 to the United States House of Representatives. This is companion legislation to the American Cures Act (S.2115) introduced in March 2014 to the United States Senate by Assistant Majority Leader Dick Durbin (D-Ill.).

In 2011, 53% of basic research funding in the United States came from the federal government. However, the federal government spends two-thirds less on research and development today than it did in 1965. The National Institutes of Health (NIH) is the foremost biomedical research institute in the world and has funded fewer research grants every year over the past 10 years.

The American HEALS Act would reverse that trend. This legislation would require amounts be transferred from the Fund each fiscal year to certain programs and agencies. These include NIH, Centers for Disease Control and Prevention (CDC), Department of Defense (DOD) health program, and the medical and prosthetics research program of the Department of Veterans Affairs (VA).

The bills would ensure funding for these programs and agencies does not fall below 105% of the level of funding provided for the preceding fiscal year. It would also provide an additional amount to account for any increases in the Gross Domestic Product for the year involved.

The legislation would require amounts appropriated for each program and agency not be less than the amounts appropriated for FY2014. It would authorize and appropriate to the Fund necessary amounts for each fiscal year to enable such transfers.

It also amends the Balanced Budget and Emergency Deficit Control Act to exempt the Fund from any sequestration order issued under such Act.

Please join the IPPF in supporting this legislation. Contact your Congressional Members and ask them to support the American HEALS Act (HR 4384 / R.2115). You can find your congressional representatives’ contact information at www.govtrack.us/congress/members.

Marc Yale is a pemphigoid patient living in Ventura, California. He has been a Certified Peer Health Coach with the IPPF since 2008. Marc advocates for our patient community both on the State and Federal levels. Marc has contributed regularly to the Quarterly newsletter in his column “Coaches Corner”. Marc can be reached at marc@pemphigus.org
My philosophy is to refer patients to a knowledgeable psychiatrist for evaluation for possible psychotropic medication. The psychotherapy component may be a fairly short-term cognitive-behavioral model, or a more lengthy psychodynamic approach. The bottom line is that everyone is unique, and no one needs to feel worse than absolutely necessary. For some people this means medication, especially in the beginning, or more frequent therapy appointments. The doctor will monitor and make changes as necessary. Having said that, the sooner the emotional diagnosis and the sooner treatment begins, the better and faster the positive effects will be in stopping any potential downward spirals.

It is often easier to speak with a professional than with someone in your personal network. The key is to identify any problem areas and to address them, not just put on a band-aid when emotional surgery is necessary. 

You can download a copy of this screening test at pemphigus.us/ippf-cesd.

**ABSTRACT:** The CES-D scale is a short self-report scale designed to measure depressive symptomatology in the general population. The items of the scale are symptoms associated with depression which have been used in previously validated longer scales. The new scale was tested in household interview surveys and in psychiatric settings. It was found to have very high internal consistency and adequate test-retest repeatability. Validity was established by patterns of correlations with other self-report measures, by correlations with clinical ratings of depression, and by relationships with other variables which support its construct validity. Reliability, validity, and factor structure were similar across a wide variety of demographic characteristics in the general population samples tested. The scale should be a useful tool for epidemiological studies of depression. (Radloff, 1977, [http://apm.sagepub.com/content/1/3/385.short](http://apm.sagepub.com/content/1/3/385.short))

**This test is not intended to replace the advice of a trained mental health professional.**

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**A SCREENING TEST FOR DEPRESSION**

Circle ONE number in for each statement best describing how often you felt or behaved during the past week. DO NOT SKIP any items.

<table>
<thead>
<tr>
<th>DURING THE PAST WEEK:</th>
<th>Rarely or none of the time (less than 1 day)</th>
<th>Some or a little of the time (1 – 2 days)</th>
<th>Occasionally or a moderate amount of the time (3 – 4 days)</th>
<th>Most or all of the time (5 – 7 days)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. I was bothered by things that usually don’t bother me.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>2. I did not feel like eating; my appetite was poor.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>3. I felt that I could not shake off the blues, even with the help from family or friends.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>4. I felt that I was just as good as other people.</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>5. I had trouble keeping my mind on what I was doing.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>6. I felt depressed.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>7. I felt that everything I did was an effort.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>8. I felt hopeful about the future.</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>9. I thought my life had been a failure.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>10. I felt fearful.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>11. My sleep was restless.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>12. I was happy.</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>13. I talked less than usual.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>14. I felt lonely.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>15. People were unfriendly.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>16. I enjoyed life.</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>17. I had crying spells.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>18. I felt sad.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>19. I felt that people dislike me.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>20. I could not get “going”.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

**ADD the total for your circled responses:** ____________

0-15, No indication of depression.
15-21, Mild to Moderate Depression. You should consider speaking with your physician or a mental health professional. Over 21, Possibility of Major Depression. You should contact your physician or a mental health professional. 

This test is based on Laurie Radloff’s 1977 Center for Epidemiologic Studies Depression Scale (CES-D).
unique antibodies from the three other PV patients.

In total, the sequencing efforts identified 21 unique heavy chains among the four patients.

All 11 antibodies could bind to Dsg3 and this was mediated via a domain (called EC1) in Dsg3 that is known to be important for its adhesive interactions, suggesting that anti-Dsg3 autoantibody binding to Dsg3 leads to a direct block in Dsg3 function in keratinocytes (and subsequent skin blistering).

Curiously, not all of the antibodies that the authors identified that bound Dsg3 could cause blistering when added to human skin tissue samples; the VH1-46-containing antibodies did. They determined that these differences in functional effects were due to the inability of the nonpathogenic antibodies to bind to the functional domains of Dsg3.

Even more curiously, the authors found that all four patients had at least one PV antibody that consisted of an identical variable region termed VH1-46. They also found very little change in the VH1-46 amino acid sequence in the patient antibodies compared to the known sequence of VH1-46 that also exists in unaffected patients (considered the “wild-type” or germline sequence).

As noted by the authors, this is a pattern typical of a somatically mutated antibody sequence, meaning that very few changes were generated during the development of the B cells (each with its own single antibody that it makes, see Figure).

They did some additional experiments to define the ability of those amino acid changes to affect the binding to Dsg3. They conclude that VH1-46 autoantibodies in PV are generated during B cell development and require very little mutation to become pathogenic. This suggests that they appear early during the development of the disease and explains their prevalence in all of the patients tested here.

These autoantibodies may not be the most common later on (during full-blown disease), but they may provide a clue to why and how pemphigus arises. The ability of these autoantibodies to escape the quality control machinery at play during B cell development is likely due to the low levels of Dsg3 antigen available that would distinguish these antibodies as “self” antibodies and therefore the ability of the machinery to mark the cells and their rogue autoantibodies for destruction.

These data led the authors to speculate that the five pathogenic (disease-causing) VH1-46 anti-Dsg3 mAbs that they’ve identified in this study are among the earliest autoantibodies formed in PV patients, caused only by how simple they are to generate from germline sequences. They also define a mechanism for how these autoantibodies are made and most importantly, how they are missed by the quality control machinery – all low probability events that likely account for the rarity of PV.

Cartoon illustrating the structure of PV autoantibodies (left) that recognize Dsg3, leading to loss of Dsg3 function in “gluing” cells together. Antibodies are proteins made up of long chains of amino acids, where four chains (two “heavy” in yellow and two “light” in blue) come together to form a single Y-shape that is found on the surface of B cells of the immune system (right). The B cells serve as vehicles through the bloodstream to reach their targets; for PV that is the keratinocytes of the skin and mucosal membranes. Every B cell clone is different based on the antibodies on its surface that allow them to recognize different antigens. The light colored regions of the heavy and light chains are the variable regions and are specific to each antibody clone, dictating which antigens the antibody will bind to. In the case of the VH1-46 antibodies characterized in this study, the antigens are Dsg3. In the variable regions of the antibodies, the tips (beyond the white stripes) contain twelve total domains called the complementarity determining regions (CDRs). In this study, the authors found antibodies containing up to six different CDR sequences from a single PV patient. All four PV patients they studied shared a single antibody clone, the VH1-46 clone.

Mirella Bucci, PhD, is Secretary of the IPPF Board of Directors and a scientific journal editor living in San Mateo, California. She is a regular contributor to the Quarterly newsletter in the “Research Highlights” column.

Donate online at www.pemphigus.org/donate
GRILLED SPIEDINI OF CHICKEN & ZUCCHINI

Vicky Starr vstarr@medprorx.com

SPIEDINI

- 1 ½ pounds skinless, boneless chicken breasts, cut into 1-inch pieces
- 6 small zucchini, cut into 1-inch slices (about 1 ¼ pounds)
- Cooking spray
- ¼ teaspoon kosher salt
- ⅛ teaspoon freshly ground black pepper

SALSA

- 1 tablespoon extra virgin olive oil
- ½ teaspoon chopped fresh thyme
- ½ teaspoon chopped fresh oregano
- ¼ teaspoon kosher salt
- ⅛ teaspoon freshly ground black pepper
- 1 cup chopped fresh parsley
- 3 tablespoons capers, chopped
- ¼ cup of white grape juice or peach juice

PREPARATION

1. Soak 12 10-inch wooden skewers in water for 30 minutes. This will help prevent the skewers from burning.
2. Prepare grill to medium-high heat.
3. Prepare salsa and set aside.
4. Thread chicken and zucchini alternately onto each skewer.
5. Coat spiedini with cooking spray.
6. Sprinkle with ¼ teaspoon salt and ⅛ teaspoon pepper.
7. Place on grill rack 6 minutes or until done, turning once.
8. Brush salsa on the chicken and zucchini to taste.
9. Serve and enjoy!

Spiedini is Italian for “little skewers.” This recipe comes from Vicky Starr who offered up several mouth-watering ideas during her workshops at the 2014 Patient Conference in Chicago.

These recipes are sometimes variations of popular recipes. For some P/P patients, different ingredients might be irritating or cause a flare. And for other P/P patients there may be no side effects at all.

As you learn how to live with your P/P you will develop an eye for what does and does not aggravate your condition. Adapt recipes as you need and as always, talk with your physician if you have any questions.
PEMPHIGUS & PEMPHIGOID
18th Annual Patient Conference

Mount Sinai Department of Dermatology
April 25-26, 2015
with a Welcome Reception on Friday 4/24.15!

Featuring Bullous Disease expert talks on skin, dental, and psychological issues.

The IPPF Patient Conference highlights the power of community featuring the collective voices of P/P professionals. Join us as we gather, engage and contribute to improving the quality of life for P/P patients everywhere. At this conference, attendees will identify strategies and discover solutions to help start your journey, or move closer towards remission. Come prepared to expand your personal support network, create professional relationships, and leave empowered and inspired.

REGISTRATION OPENS IN DECEMBER
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