

2016 PATIENT CONFERENCE | PEMPHIGUS MANAGEMENT | BULLOUS DISEASE SYMPOSIUM

Quarterly

The Journal of the International Pemphigus & Pemphigoid Foundation

DIRECTOR'S LETTER:

Marc Yale, the IPPF's new Executive Director, welcomes the change

SUMMER 2016

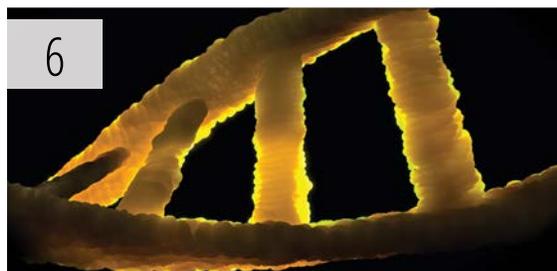


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Message from the Director



This summer seemed a little bittersweet as I watched my good friend of eight years, Will Zrnchik, leave the IPPF after making an indelible mark on the foundation. Will devoted countless hours towards our patients and community to make the IPPF what it is today. He will be sadly missed as he joins another nonprofit that helps military veterans. I also witnessed my two sweet daughters graduate. My oldest, Amethyst, from the University of Colorado, Colorado Springs and my youngest, Hannah, from middle school. Seasons, like life, signify graduation from one phase to another for us all.

I was diagnosed with mucous membrane pemphigoid in August, 2007, and was very lucky to find the IPPF. Like all patients at that time, I was welcomed into the fold by the IPPF's founder, Janet Segall, who nurtured the organization and all involved as if they were her own family. Janet convinced me to attend my first IPPF Patient Conference in Dallas, 2008. This was during the worst part of my disease, and the feeling that I would be in a room full of people who knew what I was going through was overwhelming. Ready to begin what would be two years of IV infusions and losing the sight in my left eye due to the disease, the Patient Conference was just what I needed to keep me filled with hope.

By the time I attended the conference, I had already been corresponding with other patients on the IPPF's forums and discussion group. After the conference, the IPPF asked me to help launch a new program and volunteer as a Peer Health Coach. I happily agreed, as I felt that helping others like me would help me heal in the process.

As the foundation grew, so did my understanding of pemphigus and pemphigoid. I worked with patients one-on-one, helping them learn to manage their condition. I also worked with the foundation to develop Comprehensive Disease Profiles for P/P. My role with the IPPF continued to expand as I built new programs to serve our community. I represented the organization at conferences and meetings. In the halls of Congress, I shared our stories, raising P/P awareness among senators, representatives, patients, caregivers, industry partners, and others.

When I was asked to step in as Interim Executive Director in June, I was honored. I never thought this disease would be such a blessing in disguise. Over the last eight years, I have been fortunate to be a part of such an extraordinary community. Our volunteers and staff work tirelessly to bring you this **Quarterly** journal and so many other programs designed to improve the quality of life of people affected by pemphigus and pemphigoid.

I look forward to working with each and every one of you in the future as we strive to find a cure for pemphigus and pemphigoid!

Marc Yale
IPPF Executive Director
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The Long Road:

A PEMPHIGOID PATIENT'S JOURNEY TO DIAGNOSIS

Patrick Dunn



This article first appeared on the Undiagnosed Diseases Network (UDN). The UDN is a research study that is funded by the National Institutes of Health Common Fund. Its purpose is to bring together clinical and research experts from across the United States to solve the most challenging medical mysteries using advanced technologies. The Coordinating Center of the UDN is based at the Department of Biomedical Informatics at Harvard Medical School. For more information on the Undiagnosed Disease Network, visit <https://undiagnosed.hms.harvard.edu>.

Pemphigus and pemphigoid (P/P) are rare, auto-immune blistering diseases that affect the skin and mucous membranes, causing lesions that do not heal. Like many suffering from a rare disorder, the diagnostic journey for a P/P patient is complicated and stressful. On average, this journey involves five different doctors over a period of ten months before a patient receives a correct diagnosis. (Source: pemphig.us/diagnostic-data) Once a correct diagnosis is received, P/P patients begin the long process of managing disease activity. There is no cure for P/P. Many patients are able to achieve a state of remission; however, insurance regulations make it difficult for some patients to receive the most effective treatments.

Sharon Williamson's journey to a pemphigoid diagnosis started in July of 2014 when her mouth began to bleed whenever she brushed her teeth. Like many undiagnosed P/P patients, Sharon first shared her symptoms with her dentist. This dentist did not recognize Sharon's symptoms and told her to floss more frequently.

The next doctor Sharon saw was an ear, nose, and throat (ENT) specialist. The ENT doctor thought her bleeding was a reaction to Plaquenil, which Sharon was taking to combat her rheumatoid arthritis. The ENT doctor put her on a mouthwash containing lidocaine. This numbed her mouth, but did not help the bleeding.

Sharon then saw her rheumatologist. She had been

wondering if her bleeding gums and cheeks could be symptoms of Sjogren's disease, which often affects arthritis patients. The test for Sjogren's came back negative.

By December of 2014, Sharon was spitting bloody tissue whenever she brushed her teeth. "It hurt so bad that all I could do was hang over the sink and cry... spitting out blood the entire time," she said. "No toothpaste was mild enough. No toothbrush was soft enough."

From January to May of 2015, Sharon had more appointments with other doctors: an optometrist, who saw nothing wrong with her eyes, though Sharon would soon experience ocular burning; a dermatologist, who didn't recognize her symptoms; and her primary care physician, who had no suggestions. "I felt as though my face was melting off," she said.

It's sad that you can lose your livelihood, your sight, and maybe even your life, because of delays in ... getting a diagnosis and treatment.

After researching gum disease specialists, Sharon saw Dr. Jarrett Manning in Smyrna, GA, who recognized her symptoms as pemphigoid. Dr. Manning referred Sharon to Dr. Ronald Feldman, a dermatologist specializing in autoimmune diseases at

Emory University. After biopsies and blood tests, Dr. Feldman diagnosed Sharon with mucous membrane pemphigoid (MMP)—a form of pemphigoid characterized by blistering lesions that primarily affects the various mucous membranes of the body, as well as the skin. Soon after, an eye specialist confirmed ocular involvement, which could lead to blindness. This was September of 2015, 14 months after Sharon’s initial symptoms.

Though Sharon found a doctor who was able to diagnose and treat her MMP, she has not yet achieved remission.

“I feel hopeless and fear the possibility of going blind,” Sharon said. “The pain from all of my conditions drains the energy from my body, making it hard to work. I go home, put warm compresses on my eyes, and lie in darkness for two hours. Eventually, I can get up and see my husband for a couple of hours before I go back to bed. Without the additional rest, I cannot do my job. I know that eventually, I will have to go on disability.”

Some emerging treatments, like intravenous administration of rituximab, have not been approved by the FDA for pemphigus and pemphigoid. This makes it difficult or impossible for patients to get approval for such treatments from medical insurance companies.

For Sharon, this has meant playing an anxious waiting game in hope that these treatments will be approved before her disease progresses even further. In fact, it was only during the writing of this post—in April of 2016—that Sharon’s infusions were finally approved by her insurance company after three previous denials and a call to her insurance commissioner.

“It’s sad that you can lose your livelihood, your sight, and maybe even your life, because of delays in... getting a diagnosis and treatment,” Sharon said. ^{ippf}

Patrick Dunn is the Communications Manager at the International Pemphigus & Pemphigoid Foundation.

Continued Funding from Sy Syms Foundation

The Sy Syms Foundation has awarded a \$75,000 grant to the International Pemphigus & Pemphigoid Foundation in support of the IPPF’s Awareness Campaign. This will be the fourth consecutive year the Sy Syms Foundation has supported the Campaign.

Since developing the Awareness Campaign in 2013, the IPPF has reached thousands of dental professionals and students. A particularly successful aspect of the Awareness Campaign is the innovative Patient Educator Program, which sends P/P patients to dental schools across the US to share their stories of delayed diagnosis. These emotional stories compliment traditional lectures, encouraging students to remember P/P should they encounter symptoms in future practice. Since March 2014, IPPF Patient Educators have given 28 presentations at 14 different dental schools, reaching approximately 2,800 students and faculty.

“The Awareness Campaign has made incredible inroads into the dental education universe, and if we keep the momentum going in 2016/17, we will save even more lives and erase more suffering,” said Marcy Syms, President of the Sy Syms Foundation.

The average patient with P/P sees five doctors over ten months in search of a diagnosis for their condition. Delays in diagnosis and appropriate treatment can lead to a number of complications, including significant functional impairment, resistance to treatment, psychological stress, and a lower likelihood of achieving remission.

The Sy Syms Foundation was established in 1985 by retail entrepreneur and humanitarian, Sy Syms. His mission was to support education, and through his generosity the non-profit Foundation has effected the continued growth of many institutions of higher learning, medical research, and civic and cultural bodies. For more information on the Sy Syms Foundation, visit syysymsfoundation.org or call (201) 849-4417.

The Molecular and Genetic Basis of Pemphigus

Mirella Bucci, PhD

At the physiological level, the mucocutaneous autoimmune blistering diseases pemphigus and pemphigoid (P/P) are characterized by blistering of the skin. At the molecular level, they are characterized by the presence of antibodies, generated by a patient's own immune system, that direct an immune attack of the connections that form between skin cells, so-called keratinocytes. While there is much evidence that the antibodies that cause P/P recognize keratinocyte proteins of the cadherin family, namely Desmoglein (Dsg) 3 and 1, these cannot explain all instances of these conditions. For example, a subset of pemphigus vulgaris (PV) patients do not generate any detectable antibodies that recognize Dsg proteins. As well, if PV patient serum antibodies (PVIgG) are depleted of antibodies against Dsg3, they can still cause blistering in an animal model of disease. Finally, healthy individuals who do not go on to display P/P symptoms have been found to produce autoantibodies against Dsg proteins.

The research group of Dr. Animesh A. Sinha at the State University of New York at Buffalo have been interested in understanding the molecular and genetic basis of P/P and have recently published their work exploring the question of whether there are other antibodies, beyond those against Dsgs, that can cause (or at least are associated with) P/P. Their recent study, with colleagues from Michigan State University, Stanford University School of Medicine, and Van Andel Institute was published in the Proceedings of the National Academy of Sciences (doi: [10.1073/pnas.1525448113](https://doi.org/10.1073/pnas.1525448113)).

The researchers first defined a set of candidate antibody targets, using knowledge gained from their own research and research from other labs published in the medical and scientific literature. These candidate “autoantigens” include muscarinic acetylcholine receptors mAChR3, mAChR4 and mAChR5 because another group had shown in 2004 that activators of these receptors can inhibit the formation of blisters caused by PVIgG.

They identified 15 candidate autoantigens and spotted samples of each of these onto glass slides to form a set of identical protein microarrays. To test which of the potential autoantigens could be recognized by autoantibodies in patients, they then incubated the arrays with the serum from 39 individual active PV patients one pemphigus foliaceus patient, 20 ‘healthy’ subjects without a family history of PV, and 20 healthy first- or second-degree relatives of PV patients. They found that five of the candidate autoantigens could bind to antibodies present in the serum of PV/PF patients but that did not bind to antibodies in the healthy unrelated patients. These were the three muscarinic acetylcholine receptors, Dsg3 (as expected) and a protein called TPO, which is involved in iodine metabolism. However, when the authors compared the autoantibodies that could bind to the array proteins from healthy related patients to those of the active pemphigus patients, only reactivity to Dsg3 was unique to the pemphigus patients: these healthy related patients also had autoantibodies against the muscarinic acetylcholine receptors and TPO.

The authors speculate as to the potential activity of antibodies against the nonDsg proteins, suggesting that since cholinergic signaling is known to be important in adhesion between cells, pemphigus autoantibodies that recognize the muscarinic acetylcholine receptors involved in these signaling processes may disrupt the adhesion of skin cells that they help mediate. As well, antibodies against TPO have been associated already with dysfunction of the epidermal skin layer in another autoimmune condition called urticaria.

The authors ultimately propose an interesting model for understanding these data from the microarray study. They suggest that at least in some patients, in order for pemphigus symptoms, such as attack of

keratinocyte contacts, to begin, there must necessarily be autoantibodies that recognize Dsg3, but this is not enough. To reach a pathogenic threshold where blisters actually form, attack of other autoantigens such as the muscarinic acetylcholine receptors and TPO may also be needed. This model helps explain why the only difference in autoantibody profiles that they detected between the P/P patients and the healthy relatives was in the reactivity towards Dsg3.

The authors perform one final analysis, this one more genetic and building on published observations made by Dr. Sinha’s group. They have previously shown that approximately 95% of Caucasian and Ashkenazi Jewish PV patients have one of two specific versions of specific genes within the Human Leukocyte Antigen (HLA) complex located on chromosome 6. There are many HLA types within the population, and these are inherited and encode for important parts of the immune system. So, it is not surprising that a subset of these types is connected with autoimmune disorders and other diseases. In PV, individuals carrying the DRB1*0402 or DQB1*0503 subtypes of HLA are genetically predisposed to PV. In the current study, the authors found that these HLA subtypes were correlated with the autoantibody profiles that included both anti-Dsg and anti-nonDsg specificities identified by the protein microarrays. Most interestingly, even in the healthy related patients, those that shared one of these HLA subtypes with their pemphigus relatives, but do not exhibit any signs or symptoms of disease, were more likely to have autoantibodies to the nonDsg proteins.

Understanding the genetic and molecular components involved in P/P diseases is an important step in development of more directed treatments that have with fewer side effects and risks than general immune suppression, which is the current standard of care for these conditions. [ippf](#)

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 in the Research Highlights column.



Anxiety

HOW MUCH IS TOO MUCH?

Terry Wolinsky McDonald, PhD

Everyone is unique. How any given illness—whether chronic, acute, rare, ultra rare, or orphan—affects a person is unique as well. Surely some diseases and syndromes are more difficult to deal with than others and will depend on the severity of the illness; available and insurance-covered treatment modalities; a person's background, overall wellness, support system(s), relationships, family, friends, colleagues, and personal resilience; and many other factors. There is one certainty: getting a devastating medical diagnosis is a game changer.

So, where does the newly diagnosed P/P patient start? Because of the accessibility of IPPF support systems, social media, medical advisory board, and peer health coaches, no one has to feel alone going through the journey. As with the coping mechanisms that follow a loss, the first stage of coping with a medical diagnosis is often acceptance that the diagnosis is not a mistake. And that there really isn't a cure.

However, acceptance is much easier said than done, because denial is the most-used human defense

mechanism. This is the first, and most important, stage of the process. I have met people who carry diagnoses of pemphigus and pemphigoid—some for days or weeks and others for years—who remain in denial and have not allowed themselves to be educated about what the illness is or why they are being treated with specific treatments. They may have to rely on more active caregivers or family members. These are the people I refer to as "passengers" in their own healthcare journey. Some cases will be more or

When someone is diagnosed with a life-altering condition, it is normal to have strong emotions.

less severe and more or less resistant to treatment. Surely, not everyone's journey is the same. And, as most patients know, the treatments are not without their own problems, including severe side effects, especially over time. But, even though these diseases are somewhat difficult to understand and process, the

diagnosis of pemphigus or pemphigoid is no longer a death sentence, as it once was. Do not let older "doom and gloom" articles or textbooks lead you to believe otherwise.

As a patient, one must consider him- or herself to be a consumer. The saying that "an educated consumer is the best customer" can be repeated for patients. Most providers of healthcare prefer and expect patients to be aware of their diagnoses and up-to-date on understanding treatments and contraindications, and, hopefully, they will provide patients with the resources needed to understand. Personally, I would not even dream of going to an appointment without an index card or other printout of questions. My own preference is to err on the side of knowing too much when it comes to my healthcare.

When someone is diagnosed with a life-altering condition, it is normal to have strong emotions. Anxiety and depression are common. Not everyone who experiences difficulties or anxiety coping with their diagnosis will require psychotropic medications, professional counseling, or therapy. Positive support systems, resilience, and the other factors that are already in place before the diagnosis—or that can be obtained after it—become critical. Studies on anxiety have continually shown that while no anxiety can work against you, too much anxiety tends to be paralyzing; optimally, there is a healthy amount of anxiety that creates the best conditions in general.

How do you know if your anxiety level falls within a reasonable range? An evaluation by a trained professional may be helpful. If a patient (and/or caregiver)

with a serious, chronic, rare, or acute illness does seek professional therapeutic help or just an opinion, ask your insurance company, the local or state psychology association, or other associations for referrals to professionals in your area. Check websites for credentialing and training, areas of expertise, and therapeutic orientation. Just as all illnesses are not equal, neither are the physicians and other professionals. Ensure that the person who has been chosen has special training and/or experience in working with patients or clients who have serious, life-threatening illnesses. The provider does need more than a cursory understanding of working with people with underlying medical issues.

One surprise for me, having both professional training and personal experience with difficult chronic illnesses, has been the lack of training in this area for many, or even most, professionals. Writing articles and doing presentations on basic approaches for working with patients who present with chronic physical illnesses has been eye-opening for me. The more I have written or presented about this area, the more I have been asked to expand on the subtleties. If you are educating yourself, this can only help in your work with whoever you may see—or not see—as you move forward with your own personal and unique journey. There is not just one road. ^{ippf}

Terry Wolinsky McDonald, PhD, is a PV patient, clinical psychologist, and former IPPF Board member living in Pittsburgh, PA and Sarasota, FL. She is a regular contributor to the Quarterly newsletter in her "Psychologically Speaking" column.



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My Clinical Trial Experience

Marlis Lippow

My name is Marlis Lippow, and I reside in Northern California. I participated in a randomized, double-blind, double dummy study evaluating rituximab infusions vs. 2,000 mg of mycophenolate mofetil (Cellcept®). I had previously received rituximab infusions so I had a pretty good idea of what to expect if I was to again receive it. I had three previous rounds and my doctor said the effects should last about six months, after which I would probably need another round. I was lesion-free for about seven months before the lesions started to return. I also was on CellCept and prednisone, so I know how those affect me.



Learning About the Trial

My doctor mentioned a clinical trial, answered my many questions, and asked me to think about it. I returned a month later for a followup appointment and there was another doctor present. She was talking about the trial and it seemed she expected me to be a part of it. I was still unsure and had more questions. She did explain that before I could be accepted, I'd undergo a screening (ECG, chest x-ray, and blood work). That was great, I would find out how I am doing. If I passed the tests, I could decide if I wanted to participate. The trial included a stipend, \$50 for each session for gas and parking. That sounded good since I live about 45 miles away.

Making the Decision

My doctor and I discussed the pros and cons of the assorted medications and what I'd need to take if I did not participate in the trial but still needed rituximab. Either way, the side effects are not pleasant. Basically, we talked about the lesser of the two evils.

I learned my doctor is referred to as the Principal Investigator (PI) and the other doctor is the Sub-Investigator (SI). The SI would be seeing me every month. Since the SI is not my primary doctor, it is very important she have a complete grasp of my medical history. During my visits I would have blood work and urinalysis done. The PI would get the test results and be aware of my progress and any possible problems. If he felt there was a concern, he would end my participation in the study.

I also learned I could opt out of the study at any time if I became uncomfortable. In the end, I chose to be in the trial.

My Trial Experience

Throughout the study, I felt my doctor was most concerned about my well-being, as he should be. He even called me in between visits and that gave me a good, positive feeling.

I received two initial infusions two weeks apart. After about five months, I received two more, also two weeks apart. By the third round I no longer had any sores. I was told that I was "controlled" and after the last infusion, I'd be in remission! My doctor told me this remission should last anywhere from six months to three years. I am hoping it will be longer!

Don't be worried about the infusions. The infusion nurses are angels and take wonderful care of you! They are kind, let you know what to expect, and give you an idea of how you will feel. If you have any questions, you will have the phone numbers for the PI and SI and are encouraged to use them.

While the short-term benefits help us now, the long-term benefits may change the lives of patients for years to come.

Clinical trials are not for everyone. In fact, there are many qualifying and disqualifying criteria set by the drug manufacturer. I encourage everyone to consider participating in a trial to help advance research on new and emerging pemphigus and pemphigoid treatments. While the short-term benefits help us now, the long-term benefits may change the lives of patients for years to come. ^{ippf}

Marlis Lippow is a pemphigus vulgaris patient diagnosed in February 2014. She currently resides in Northern California.

Updates in the Management of Pemphigus

INTERNATIONAL GUIDELINES FOR THE DIAGNOSIS AND MANAGEMENT OF PEMPHIGUS ARE ON THE HORIZON

Sandra Peña, BA, Dedee Murrell, MD, & Victoria Werth, MD

According to the Institute of Medicine of the United States, clinical practice guidelines are systematically developed statements meant to assist both the physician and the patient in appropriate health-care decision-making in specific clinical situations⁽¹⁾. Clinical guidelines delineate the standards of care according to the most up-to-date evidence in an effort to improve patient care.

Until recently, there were no guidelines for the management of pemphigus that were accepted internationally, despite national efforts from several European countries⁽²⁻⁴⁾. In an attempt to establish international consensus, a group of European experts came together to define diagnostic and therapeutic guidelines for pemphigus. After a couple of rounds of voting, the final version was passed to the European Dermatology Forum (EDF) for a final acceptance⁽⁵⁾.

While the efforts of the European group made tremendous strides in standardizing the management of pemphigus, countries outside of Europe were not included; therefore, these guidelines may not ultimately be generalizable to

other countries. The International Bullous Diseases Consensus Group, convened by IPPF MAB members Dedee Murrell and Victoria Werth, met in March 2016 at the annual American Academy of Dermatology (AAD) conference in Washington D.C. to gather management and treatment opinions from the greater international community.

Prior to the meeting, participants were sent a survey based on the European guidelines and were asked to rate each statement from 0-100% based on their level of agreement. Statements that scored less than 30% consensus were automatically

removed, while the group automatically accepted statements that scored above 70%. For example, the statement regarding the initial evaluation, an experienced dermatologist is responsible for the treatment plan for patients with pemphigus, was accepted automatically after attaining 100% consensus amongst the group. Additionally, there was nearly unanimous agreement that the diagnosis of pemphigus depends on the clinical presentation, histopathology, direct immunofluorescence microscopy, or serological

Until recently, there were no guidelines for the management of pemphigus that were accepted internationally...

evidence of auto-antibodies and again these points were accepted consequentially.



Have a Support Group?

LET US KNOW!

It doesn't have to be formal to be a group. All you need is another person, a place to sit, and time to talk. The important thing is to share your experiences and get the support you need. To find others in your area, contact Noelle DeLaney: noelle@pemphigus.org.

Here's the Southern California Support Group at their April meeting in Santa Monica, CA!

During the consensus meeting, the points that scored between 30-70% were discussed and revised accordingly in an attempt to broaden the acceptance of all statements. Lastly, following the AAD conference, participants were once again asked to rate the revised statements to determine their level of agreement. The results of this process are being analyzed.

References

1. Field MJ, Lohr KN, eds. Clinical practice guidelines: directions for a new program. Washington, DC: National Academy Press, 1990.
2. Joly P, Bernard P, Bedane C, Prost C, Ingen-Housz-Oro S. Pemphigus. Guidelines for the diagnosis and treatment. Centres

de reference des maladies bulleuses auto-immunes. Societe Francaise de Dermatologie. *Ann Dermatol Venereol* 2011; 138: 252-258.

3. Harman KE, Albert S, Black MM. Guidelines for the management of pemphigus vulgaris. *Br J Dermatol*. 2003; 149: 926-937.
4. Eming R, Sticherling M, Hofmann S et al. S2k guidelines for the treatment of pemphigus vulgaris/foiaceus and bullous pemphigoid. *JDDG: Journal der Deutschen Dermatologischen Gesellschaft*. 2015;13(8):833-844. doi:10.1111/ddg.12606.
5. Hertl M, Jedlickova H, Karpati S et al. Pemphigus. S2 Guideline for diagnosis and treatment - guided

by the European Dermatology Forum (EDF) in cooperation with the European Academy of Dermatology and Venereology (EADV). *Journal of the European Academy of Dermatology and Venereology*. 2014;29(3):405-414. doi:10.1111/jdv.12772.

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Pemphigus, Pemphigoid, & Social Security Disability

Social Security Disability Help

Pemphigus and pemphigoid are a group of rare autoimmune diseases that affect the skin and mucous membranes causing painful blistering and scarring. For some patients, the noticeable and sometimes life-threatening, symptoms are too much to work with. The Social Security Administration's (SSA) disability benefit programs are for patients like those who can't support themselves because of their disability.

Social Security Disability

If you cannot work enough to earn the SSA's monthly Substantial Gainful Activity (SGA) limit of \$1,130, then you may be eligible for Social Security Disability Insurance (SSDI). Generally, you need to have worked five of the last ten years in a job that pays into Social Security, and additional years depending on your age. Benefits average \$1,000-\$1,500 each month, but the SSA calculates your payment by averaging previously reported income, so it may be higher or lower.

After you are approved, you will not receive your first check until five months after the date your pemphigus or pemphigoid started, or your reported onset date. Be aware the SSA may take up to two years to approve the claim; however, you will be given any back pay in a lump sum check. Two years after your reported onset date, you will be eligible for Medicare.

Supplementary Security Insurance (SSI) is not paid from taxes, so it is best for those with little or no work history, or parents applying for children. In most states, you will also be automatically enrolled in Medicaid once approved for SSI. You are allowed to work while on SSI, but individuals cannot earn more than \$733 a month in countable income, and couples cannot earn more than \$1,100 (\$1,100 is the total countable income limit for a couple with SSI, if an SSI-eligible individual is married to someone who is not eligible, then the SSA does a process called "deeming" where they factor

in the ineligible spouse's income and resources to determine payment amounts.). Individuals and couples also cannot have more than \$2,000 and \$3,000 worth of assets. If a person/couple owns a home, that home is not counted toward their income or resources as long as it is their primary residence.

Medical Requirements for Pemphigus and Pemphigoid

The SSA evaluates all applications they receive first with the Blue Book, their official list of impairments and medical requirements. If you meet or equal a condition in the Blue Book, you will be automatically medically eligible for benefits. This means that an individual can qualify for benefits based off of their condition, but the decision is ultimately up to a disability examiner, so an individual may have to file an appeal if the examiner denies their application.

Pemphigus and pemphigoid can



be found in Section 8.00—Skin Disorders of the Blue Book. You need to provide medical evidence of the following:

- *Bullous disease (pemphigus, bullous pemphigoid), with extensive skin lesions that persist for at least three months despite continuing treatment as prescribed*
- *Chronic infections of the skin or mucous membranes, with extensive tissue death or extensive ulcerating skin lesions that persist for at least three months despite continuing treatment as prescribed*

If you do not meet a Blue Book listing but feel your conditions restricts you from performing SGA, you may still be approved with a medical-vocational allowance based on your limitations. Applicants do not need to do anything for a medical-vocational allowance. The medical-vocational allowance is a determination made by the SSA during the application

process after they determine that someone is not medically eligible by the Blue Book criteria. This determination takes into account a person's ability to do the work they have been doing, whether they would be able to be re-trained for another job, whether they can perform any type of work that they've done in the last 15 years among other factors.

Applying for Social Security Disability

Talk to your doctor before completing the SSI application. Because the claim can take so long to process, be reviewed, and a determination made, it may not be worth applying if your doctor does not think your chances are good enough. If you do decide to apply, a detailed statement from your doctor about the limitations caused by your pemphigus or pemphigoid is crucial to the claim.

The SSA offers a convenient application on their website for SSDI. SSDI is Social Security

Disability Insurance – meaning that it is based off of your work history and income, and its premiums are paid with payroll taxes. SSI is a needs based program and because of that has income limits, restrictions on resources, and a lower maximum benefit amount. If you're not comfortable applying online or you are applying for SSI, you will need to make an appointment at your local SSA office. You can find a list of documents (such tax information and a birth certificate) you will need, in addition to important medical evidence (labs, imaging tests, and surgical reports) on the SSA's website. ^{ippf}

Social Security Disability Help is an independent organization that specializes in providing information about disability benefits for people affected by debilitating diseases and conditions like pemphigus and pemphigoid. Learn more: www.disability-benefits-help.org



It's Natural

Noelle DeLaney

It's no secret among the pemphigus and pemphigoid (P/P) community that treatments to help these rare diseases are not always ideal. It's also common for patients and caregivers to reach out to the IPPF seeking advice on holistic/natural/eastern medicines to treat P/P before the typically used western medicines recommended by physicians.

The IPPF understands that the potential side effects of steroids, immunosuppressants, and biologics can be very concerning. You already have a lot going on with your body and immune system. Why subject your immune system to these medications if you don't have to?

The IPPF is not against patients trying holistic or more natural remedies. We truly want P/P patients to get into remission by whatever means work for them. However, you need to know one very important fact before trying any natural treatments.

Many natural or holistic treatments work by “boosting” or “enhancing” your immune system.

When you have an autoimmune disease like pemphigus or pemphigoid, that means you have a compromised immune system. Therefore, if you “boost” or “enhance” your immune system with an active autoimmune disease, you may be putting yourself at risk of “boosting” or “enhancing” your disease activity.

The IPPF highly recommends that you do your research before you try any natural or holistic treatments. Make sure the treatment you are interested in won't boost your immune system. The IPPF also recommends that you consult your doctor before trying any treatments. While your intention of trying this treatment will be to make you healthier and improve your disease activity, you may be inadvertently making your disease worse. That is the last thing we want for you, and we know it's the last thing you would want for yourself.

The IPPF doesn't want to discourage you from trying something that could alleviate your disease symptoms or make you feel more

comfortable. We just want you to thoroughly research the product and consult your doctor before doing so.

Going off of prescribed medication in order to try a holistic treatment is not recommended. Patients who go off of treatments to try natural therapies without consulting their doctors can put themselves in a potentially dangerous situation. Let your physician be on this journey with you.

The IPPF does recommend you take your vitamins, exercise, and eat a balanced diet. We also recommend consulting your doctor about ALL treatments that could help treat your pemphigus or pemphigoid. [ippf](#)

Noelle DeLaney joined the IPPF in 2014 as the Patient Services Coordinator. Noelle facilitates the support and education programs within the foundation. She works directly with the Peer Health Coaches to help improve the quality of life for pemphigus & pemphigoid patients. Noelle lives in Dixon, CA with her husband and dog.

Bullous Disease Symposium at International Congress on Autoimmunity

Sergei Grando, MD, PhD, DSc

The International Congress on Autoimmunity held its 10th meeting in Leipzig, Germany from April 6-10, 2016. This Congress is entirely devoted to all autoimmune diseases affecting every organ system. It provides an opportunity for physicians and scientists to share their research and discuss the latest in their areas of expertise in autoimmunity. This is a unique time to network and develop international collaborations.

The session “Autoimmunity and the Skin” was chaired by Dr. A. Razzaque Ahmed, along with Dr. Enno Schmidt from Germany. The speakers included Dr. Ralf Ludwig (Lübeck), Dr. Sergei Grando (UCI), Dr. Thomas Luger (Muenster) and Dr. Miklós Sárdy (Munich) and Dr. A. Razzaque Ahmed (Boston).

Dr. Ahmed has chaired the sessions “Autoimmunity and the Skin” at the last four meetings in Ljubljana, Slovenia; Granada, Spain; Nice, France; and Leipzig, Germany. Speakers at these meetings included Drs. Janet Fairley, Sergei Grando, David Woodley, John Zone, and A. Razzaque Ahmed from the US. European speakers included Drs. Pascal Joly and Frédéric Caux from France, Dr. Jose Mascaro from Spain, and Drs. Detlef Zillikens, Enno Schmidt, Ralf Ludwig, and Thomas Rhoer from Germany.

At the Gala Dinner of the 10th Congress on Autoimmunity in Leipzig, Prof. Yehuda Shoenfeld, President of the Congress, presented on behalf of



Dr. A. Razzaque Ahmed (left) receiving a certificate of appreciation for his contributions to the Autoimmunity Congress Series. The award was presented by Prof. Yehuda Shoenfeld (right), President of the International Congress on Autoimmunity.

the Congress a “Certificate of Appreciation” to Dr. A. Razzaque Ahmed for his “Extraordinary Contributions to the Autoimmunity Congress Series.” There were 2,750 physicians and scientists who attended the Congress. Dr. Ahmed was the only physician who received such a Certificate of Appreciation.

The IPPF and its supporters congratulate Dr. Razzaque Ahmed on receiving this unique certificate given by the most respected international organization entirely devoted to the field of autoimmunity. He has made several important contributions to the pathogenesis and treatment of autoimmune blistering diseases.



PEMPHIGUS & PEMPHIGOID

2016 Patient Conference in Austin, TX

September 22-24

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HOSTED BY DR. TERRY REES

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- World-renowned speakers on plenary panels
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- Patient & caregiver workshops

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- Book by phone: (512) 457-7961
(Ask Gisselle for the IPPF room block.)

Don't Miss the
IPPF CASINO NIGHT
CONFERENCE KICKOFF!

THURSDAY, SEPTEMBER 22

Cocktail hour
Awards Dinner
Casino Night

FRIDAY, SEPTEMBER 23

Conference Sessions:
8:00am to 5:00pm
Lunch at Eighteenth over Austin
Cocktail reception

SATURDAY, SEPTEMBER 24

Conference Sessions:
8:00am to 12:30pm

2016 Patient Conference Registration Form September 22-24 Austin, TX

Print Name (as you want it to appear on your name badge)	Organization/Employer Name (Corporate Sponsors Only)
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I am a Patient _____ (PV, BP, etc) Caregiver Friend Other _____

Mailing Address: Street Home Work _____

City	State/Province	Zip/Postal Code	Country (other than US)
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Daytime Phone: Home Work Cell () _____

Email: Home Work _____

Patient Conference Registration Fees <small>A portion of your registration may be tax-deductible. Please consult with your tax professional.</small>	Number of People	Total
I WANT IT ALL! Complete Conference Package: \$199.00 per person Join the IPPF at our Annual Awards Celebration! Cocktail hour starts at 5:00pm followed by dinner at 6:00pm. Cards fly from 8:00-10:00pm at Casino Night! Guests receive \$1,000 in chips for the Blackjack, Craps, Texas Hold'em Poker, and Roulette tables. Rebuys available. Prizes awarded throughout the evening -- and a special Grand Prize for the person with the biggest chip stack at the end of the evening.	x \$199	
MEETING ONLY! Conference Registration Only: \$150.00 per person You will be registered for all plenary sessions and workshops on Friday and Saturday at The Hilton Garden Inn Downtown Austin. Also includes light breakfast, breaks, and conference materials.	x \$150	
AWARDS DINNER ONLY! \$75.00 per person Cocktails, Dinner, Awards, and Casino Night!	\$75	
Scholarship Donation. Please help someone else attend this year's conference.	\$	
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Total Enclosed	\$	

Check/Money Order made payable in U.S. funds to IPPF

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AMEX Exp. Date: ____ / ____ Security Code: _____

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Signature: _____ **Indicates you agree to have your card charged.**

WAYS TO REGISTER	OTHER ATTENDEES (as indicated above)
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<ul style="list-style-type: none"> Online: www.pemphigus.org/2016pc Phone: 855-4PEMPHIGUS (855-473-6744) Contact your Peer Health Coach Mail this completed form with payment to: IPPF 2015 Patient Conference 1331 Garden Highway #100 Sacramento CA 95833 	Enter information as it should appear on badge:												
	<table border="1" style="width: 100%; border-collapse: collapse;"> <thead> <tr> <th style="width: 33%;">NAME</th> <th style="width: 17%;">CITY & STATE</th> <th style="width: 50%;">DISEASE OR ROLE*</th> </tr> </thead> <tbody> <tr><td> </td><td> </td><td> </td></tr> <tr><td> </td><td> </td><td> </td></tr> <tr><td> </td><td> </td><td> </td></tr> </tbody> </table>	NAME	CITY & STATE	DISEASE OR ROLE*									
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* PV, PF, PNP, BP, OCP, MMP, Caregiver, Family, Friend, Physician, etc.

Registration cannot be processed without payment. Payment must be in US funds. All cancellations must be received in writing or by email to info@pemphigus.org. For cancellations postmarked on or prior to **August 15, 2016**, we will refund registration costs less \$25 administrative fee. Due to the contractual agreements for food, beverage and material costs, cancellations postmarked between August 16 and September 12, 2015 may be refunded up to 50% of the registration fees. **We may be unable to make refunds after September 12, 2016.** We will gladly transfer your registration to another person (scholarship) or credit it as a donation.

International Pemphigus & Pemphigoid Foundation
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