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2014 PATIENT CONFERENCE  
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RAISING AWARENESS  
RARE DISEASE DAY GIVES HOPE TO PATIENTS EVERYWHERE
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Houston Support Group, April 12, 2014
   www.pemphig.us/ippf-hsg-apr12-2014

   Chicago, Illinois www.pemphigus.org/2014conference

TELL US YOUR STORY!

The IPPF Awareness Campaign is looking for patient testimonials on
your ORAL diagnostic experience.

For more information, please contact Kate Frantz at kate@pemphigus.org
or 855-473-6744 x125.

Stories should be 300-800 words and emailed as a MS Word attachment or in the body.
Can you believe it’s already March? I can’t! Rare Disease Day came and went, Autoimmune Disease Awareness Month is upon us (see page 13 for a special offer), and the Patient Conference is nearly upon us! If you ever wanted to attend – this is the year! Chicago? Need I say more!

**New to the Office.** I’d like to welcome Noelle Madsen to the IPPF family as Patient Services Coordinator. Noelle has a background in health education, event coordination, and working with patients and physicians. She will join Marc Yale and the Peer Health Coaches helping new and existing patients, caregivers, and medical professionals every step of the way! If you will be in Chicago at the Patient Conference, stop by and welcome her aboard!

2013 Holiday Fundraiser. Wow! Let me say that again: **WOW!** Our 2013 goal was $50,000 and we not only surpassed it, we **crushed it!** This Holiday Season we raised over $64,000 that goes directly to supporting our patient services and education programs. Great job, everyone!!

**IPPF joins RareConnect.** RareConnect is a growing online network for rare disease communities, bringing together thousands of patients, families, and groups who might otherwise be isolated. Through RareConnect, patients and those who care for them can communicate, sharing experiences and in-
One Man’s Journey
DISCOVERING, BATTLING, AND OVERCOMING PEMPHIGUS VULGARIS

It is my first-year anniversary of being diagnosed with Pemphigus Vulgaris. The severe and painful blistering in my mouth and throat have subsided. My journey into discovering what was wrong with me wasn't as long as it is for most. My goal here is to help others take control over their health — especially when it is threatened by a disease that mystifies even the medical community. I didn’t know about the IPPF, so I was on my own when I was diagnosed, with no support from others who have suffered from this disease. I was able to self-diagnose through intense research and seek out the right type of treatment. Today, I’m in remission.

Stu Zirin, PV Patient

It all started with me – a true foodie – and a scrumptious kettle chip I sampled at a barbecue in late August 2012. As an executive chef and owner of two restaurants/bars, I’m one of those people whose world revolves around great food and friends, cooking and entertaining. I used to throw dinner parties for 14 people in my home and served up multiple courses every Wednesday night. I was even written up in Time Out Chicago...

Back to the barbecue. When I bit into that perfectly cooked kettle chip on that late summer day, I savored its salty thickness at first. But then, I said – ouch.

The sharp edge of the chip cut me on the area between my gums and my cheek. It didn’t really phase me. I thought to myself the mouth heals quickly and it would be gone in a couple of days.

Days passed and the cut remained. I went online to research homeopathic remedies. I rinsed with honey, gargled with a hydrogen peroxide/water mix, chewed basil leaves. If I came upon a remedy – I tried it. Weeks passed by, and there was no change. In fact, my condition grew worse. I instinctively knew something wasn’t right.

When my dentist examined me, she commented on how bad the sore in my mouth looked, but had no idea what was causing it. She surmised it might be Herpes and I completed a prescription meant to heal it. When I went back to see her there was no change – and no ideas what this might be.

I visited my uncle in New York who was a practicing dentist of forty years. He suggested I gargle with salt water and restrict my diet to soft foods. I was diligent in following his advice, but again, nothing changed.

From there, I saw many doctors: my primary care physician, oral surgeons and oral pathologists, rheumatologists and allergists. The feedback was always the same – the inside of my mouth looked horrible, but the cause was unknown. All I knew was I couldn’t eat anything I used to love.

My diet was eventually restricted to egg whites, oatmeal and protein shakes. Any food harder was impossible for me to eat. Spices of any kind were out unless I wanted to be brought to my knees in pain. I began an investigation of my own.

I spent countless hours on the Internet looking up anything that could help me find the answer. . .what was wrong with me? I became so frustrated, I would leave doctors’ offices and picture them closing my file and moving on to the next patient – possibly shrugging their shoulders. Depression began to set in. This was my health, and I was grasping for answers.

I saw another oral surgeon who sent a biopsy sample to University of Chicago. I waited three long and excruciating weeks for the biopsy to come back inconclusive. More frustration.

One day I was having lunch with my brother and ordered a salad. Soon I felt a horrible burning sensation, one I likened to drinking a Molotov cocktail. I realized it was a red onion.

That onion eventually lead me to my answer...
A Long-Term Remission

Skip Van Lenten, PV Patient

I came down with my first symptoms of pemphigus in the early part of 1994. I have been in remission since 1998 – 16 years without medications or blisters. My life is so different now and I rarely think about it. But it wasn’t always this way.

After I bought my first computer I started searching for information about pemphigus. I came across the IPPF’s online discussion group. I cringed as I read each email and was reminded of how uncomfortable and unnerving this disease can be. I thought about how I managed to go into remission and remain blister-free for so many years, and what it was that might say to the group that could provide some hope of a similar outcome.

I kept journals during the time I had pemphigus, and wrote in them nearly every day. I recorded my medications and my physical state, thoughts and feelings. It was comforting to spell out my changing moods, from hope to despair, and vice versa.

My journals became a confidant of sorts, where I could bitterly complain about my condition, or exalt over the slightest sign of improvement. They also became a valuable source of insight when I looked back through them in my second year of pemphigus and found a pattern of ups and downs.

CONTINUED ON PAGE 9...
Introducing the IPPF’s NEW Awareness Campaign

Kate E. Frantz, MPH
IPPF Awareness Program Manager

“13 months, 6 doctors”
“4 months, 4 doctors”
“7 months, 4 doctors”

These statements are from three patients describing how long it took, and how many doctors they saw, before receiving a P/P diagnosis. I could put this data into a spreadsheet. I could tell you that, on average, it takes P/P patients 10 months and five doctors before receiving a proper diagnosis. But, I think we would be missing something.

Numbers are more than just data points: they tell a story. Between the digits we see frustration, anxiety, and confusion. We see seven months on the wrong medication, six physicians making misdiagnoses and 13 months in search of answers. For many P/P patients, the pathway to a diagnosis is quite grueling -- both physically and mentally. It doesn't have to be this way.

This is where the IPPF Awareness Campaign comes in!

Over the next three years, the IPPF will unleash a national Awareness Campaign focusing on early diagnosis and best treatments of P/P. The ultimate goal of the campaign is to achieve better patient outcomes by reducing the time it takes and the number of doctors seen along their pathway to diagnosis. This will be accomplished through five objectives:

1. **Media and Marketing.** Be on the lookout for a new website, patient and scientific videos, social media, ad placements and educational technology.

2. **Dental Student Education.** Patient educators will travel to 19 dental schools throughout the US to provide an emotional appeal to dental students. Topics include: pathway to diagnosis, symptom recognition, and patient care and understanding.

3. **Dental Provider Education.** The IPPF will provide tuition-free continuing education credits to train primary care dentists and oral specialists on P/P symptom recognition, diagnosis, and best treatments.

4. **Awareness Ambassadors.** The IPPF will develop an Awareness Ambassador program where IPPF community members provide outreach to dental providers in their region, build rapport with local media outlets, and plan and/or participate in community outreach and awareness activities. Stay tuned for information on how YOU can become an Awareness Ambassador!

5. **Best Practices Conference.** The IPPF will hold a Best Practices Conference of clinicians and scientists to develop a clear message and consensus regarding the most effective P/P treatments.

**Dentists** are the primary focus of the Awareness Campaign because the majority of P/P patients develop oral symptoms first and see dentists early on, often well before other healthcare providers. Primary care dentists have the unique opportunity to shorten diagnosis times by recognizing P/P symptoms and making referrals to oral specialists. Dentists could lessen the uncertainty of a patient by educating them about P/P, providing clear information, and showing confidence in treating P/P.

Delayed diagnosis can increase patient pain and suffering, heighten risk for worsening illness and poor response to treatment, result in loss of productivity and reduced quality of life, cause anxiety and fear, and increase patient healthcare expenses. The Awareness Campaign aims to significantly reduce patient uncertainties and increase positive patient outcomes by shortening the pathway to diagnosis and providing patients with best treatments sooner rather than later.

We encourage our Community to get involved with the Awareness Campaign to help spread awareness. If you are interested in learning more about the campaign or getting involved, please contact Kate Frantz at Kate@pemphigus.org.
The most common complaint I, as a psychologist, hear from chronic pain or illness patients is not, “why me?” or “poor me” but rather “other people just don’t get it.” And, often the hardest part is that the people to whom the patient has felt closest to previously are often the very ones who ‘just don’t get it.’ This can include immediate or extended family members, co-workers/colleagues, or friends. It often includes primary care physicians, as well.

Additionally, being part of a not-well-known or rare chronic illness population has a lot of very real medical and life challenges. Being misunderstood can lead to even more frustration and feelings of loneliness, low self-esteem or depression.

Because of the groundbreaking work of the IPPF and its primary focus on patients, there is a pemphigus/pemphigoid (P/P) community, and no one has to feel alone again. But, because they are rare, P/P isolates patients from the people they spend most of their time with – and those people don’t fully understand. Simple comments or true answers to “how are you feeling?” can be misconstrued by some as incessant complaining. Others don’t want to hear about it. If someone looks okay, they must feel okay. Right? In a word, “No.”

How should people with chronic pain or illness present themselves in order to achieve an understanding from others around them? There is no “correct” answer that works for everyone. But there are choices. And different choices will work better in different situations. If someone is in a wheelchair, using a walker, limping, or having breathing problems, for instance, they will be seen as handicapped or sick.

Most people do not want to be seen as sick, but being seen as totally well, while suffering the debilitating symptoms of P/P, has expectations. If you are not sure where you stand on this, there is an easy way to determine your position.

Draw a horizontal line on a piece of paper: On the far left end write “human being”; in the middle write “human being with a (or several) medical problems”; on the far right, write “walking disease.” You can then think about 1) where you see yourself on this continuum, and 2) how you...
The main physical manifestation of the P/P diseases is the presence of blisters on the skin and mucous membranes. Underlying those blisters are numerous molecular processes including recognition of keratinocyte cells of the skin and cell death. But how these blisters actually form, that is, what is the order of events leading up to their formation, has not been clear. A recent study by scientists Parviz Deyhimi and Payam Tavakoli suggests that in pemphigus vulgaris (PV), cell death comes first, then the formation of blisters (Journal of Oral Pathology and Medicine, doi: 10.1111/jop.12022).

The blisters that form in PV are referred to as lesions, or suprabasal vesicles, because of where they are found within the layers of the epidermis (supra meaning above, so above the basal layer, see Figure 1a).

Because they are found so deep within the tissue, the blisters formed and PV disease itself is considered more severe than pemphigus foliaceus, where the blisters appear within the granular layer. The lesions formed during PV and in other mucocutaneous autoimmune blistering diseases are formed when the rogue antibodies formed during disease recognize proteins found at junctions formed by keratinocyte cells interacting with one another. The loss of these junctions that generates the tear in the skin is called acantholysis.

Acantholysis is more than a tearing of the skin. There is also cell death (also called apoptosis) within the lesions. But it has been unclear when and where apoptosis occurs with relation to acantholysis and to recognition of the junctions by antibodies generated by the immune system of the patient.

Besides the ordering of events, it has been unclear which of the various types of apoptosis are at play. In the intrinsic pathway of apoptosis, a cell essentially commits suicide because of an internal trigger, perhaps as part of a genetic program as occurs during cell or tissue development. In the extrinsic pathway, the trigger to commit suicide is external. Perhaps this is where the antibodies of PV patients play a role, then?

At least two models, both with excellent experimental support, exist for the ordering of events. The first suggests that apoptosis is a late event in pemphigus and that it is not required for acantholysis and blister formation, while the second suggests that apoptosis occurs early, before significant acantholysis. A related viewpoint to the second is that the two occur simultaneously, though independently, though evidence exists for apoptosis actually causing acantholysis. For instance, chemical inhibitors of apoptosis have been shown to prevent lesion formation and a time-course study has shown that apoptotic cells were present before blisters in pemphigus foliaceus.

The current authors looked at tissue samples from 25 patients with oral lesions due to PV. They used immunohistochemistry, the same technique that is used to diagnose PV (Figure 1b). Looking closely for regions where normal lesion-free tissue was adjacent to lesions, so-called peri-lesional regions, they found that 100% of the cells within lesions had fragmented DNA, the hallmark of apoptosis. In the adjacent normal tissue (in the parabasal region) of most of the samples, 75% of the cells had the marker of apoptosis. Looking at the acantholytic cells within the lesion, the re-
and downs that seemed to correspond to those times when I had experimented with various vitamin and mineral supplements.

I could see after adding supplements for a few weeks my entries were more upbeat and optimistic; and when I quit taking them, I became discouraged and frustrated. I tried the vitamins several times to see if I continued to improve. Within 6 months I saw my blisters healing, I tapered off of prednisone, and entered remission. I can’t say for sure taking supplements led to my remission, but I think it’s safe to assume improving my overall health increased my chances.

One key element that could have helped is the mineral zinc. Zinc stimulates the adrenal glands to produce natural corticosteroids. Prednisone is a synthetic corticosteroid. It’s possible increasing zinc levels lowered my need for prednisone while restoring the natural secretion of corticosteroids helping the immune system distinguish between healthy tissues and foreign “invaders.” Zinc also plays a role in the Thymus gland, which produces regulatory T-cells (or T-regs), responsible for secreting immunosuppressants.

You should know I never considered supplements a substitute for prednisone or any other medication prescribed by my doctors. I never deviated from my doctor’s orders. I learned certain key vitamins and minerals are depleted by prednisone and other drugs, including Folic Acid, Magnesium, B-6, B-12, Calcium, and Vitamin D.

I chose a multi-vitamin supplement containing 23.5 mg. of zinc and the recommended levels of other nutrients to counter that effect. After several weeks I reached a relatively normal balance between the two. At least I was able to get through the disease and function without experiencing severe side effects drugs can produce.

Adding a supplement to your diet is something you should to discuss with your doctor. Finding the right multivitamin for you is easy. What was once unique to the health food market is now a common formula found in most pharmacies under a variety of brand names.

The farther away I get from my experience with pemphigus, the less I think about it. The years have gone by quickly. Now that I am retired, I find myself working quietly at home doing things I love, and feeling very grateful for having come through it without any long-term effects.

I no longer take the supplements I feel helped me to recover, or any medications at all. I believe I am living my life the way it was meant to be, but I still keep a journal of sorts. You never know when you might want to look back and remember the good days, as well as the bad.

Skip Van Lenten is a retired house painter, amateur artist, musician, and writer. He has been a member of the IPPF email discussion group for many years. After going into remission, he established the web site www.pemphinremission.com, publishing personal stories of remission as a source of hope and encouragement for pemphigus and pemphigoid patients. He can be reached through this site, or his personal site www.vanlenten.com, where he posts his own art and writings.

Images from Rare Disease Day

From top to bottom: Janet Segall and Will Zrnchik at the California State Capitol Event; Marc Yale outside the office of US Senator Dianne Feinstein (D-CA); Kate Frantz, Janet Segall, Monique Rivera & Will Zrnchik; Janet Segall talking about P/P; Marc Yale ‘enjoying’ the DC. weather.
I searched online for ‘mouth sores’ and ‘onion.’ The words ‘Pemphigus Vulgaris’ popped up, front and center. As I read more about this disease, I discovered five documented symptoms and I suffered from all of them. I learned a number of foods, including onions, aggravate the disease and its symptoms. Though PV can occur in individuals of all ethnicities, I discovered people of Mediterranean ancestry, Ashkenazi Jews and people from Columbia and Brazil are more likely to have it.

The most damning information I came upon demonstrated what was believed to be the severity of this condition in a 2007 publication.

“Unfortunately, if you have been diagnosed with the autoimmune blistering disorder known as Pemphigus Vulgaris, it is very likely that your life expectancy will be significantly reduced, often to just within a few years of the onset of symptoms. Most early deaths associated with autoimmune blistering disease are the result of secondary infections, such as that of the genitourinary system.”

That was one of the first published articles I read about the disease and I was horrified. Though I could not give in to the idea it would kill me in a few years, I did believe I was in for a miserable life ahead.

Despite the doom and gloom, I finally had an answer after three months of living in the dark. Finally. Now I had to do something about it.

My primary doctor suggested I see an ear, nose and throat specialist. I reached out to a dear friend who is a Harvard educated physician, Dr. Scott Smith, who was practicing dermatology at Northwestern. He said he had attended a seminar years ago about this disease, and agreed to help me. I learned about a blood test for Desmoglein, which are proteins that join human cells together. People who test at levels of 0–9 don’t have PV, or express any symptoms; levels of 9–20 are classified as ‘undetermined’; levels of 20+ indicate a person has the disease. When I was tested, my level was at 75.

The very next day I couldn’t even talk; I was certain this disease had already begun to take a terrifying toll on me.

Dr. Joquin Brieva, a specialist in Chicago, explained how different parts of the mouth, throat and eyes could be attacked. Basically, it is the good cells attacking other good cells in the body’s mucous membranes. The glue that held my skin together was actually being attacked by my own body.

What I also learned some people respond positively to Rituxan® (rituximab), a type of chemotherapy. The good cells doing harm to the body are tagged by this treatment and stopped from doing more damage. Monoclonal antibodies (clones of original cells) help to strengthen the immune system.

I told people the most dangerous thing I did every day was to brush my teeth; it took one slip of the toothbrush from the front to back and I ripped the lining of my cheek like stripping wet drywall of the wall with a putty knife.

When I began Rituxan® in January 2013, my Desmoglein levels had jumped from 75 to 128. Every day after I finished my treatment I would go shopping. There really is something to be said for retail therapy to get your mind off of things. I would push myself to go to the gym for at least ten minutes and ended up working out longer. It made me feel more positive. I never missed a day of work. I would listen to motivational tapes on my drive. I got dressed up and wore a suit and tie every day. I even got dressed up for my treatments.

Besides the exhaustion, and severe depression at times, I became very moody because of the steroids. I got infections in my nails, sties in my eyes, and rashes. It was just awful.

I couldn’t get a good night’s sleep and was in constant pain. Then I met Dr. Judy Paice. She walked into my life at just the right time, and was so compassionate and caring. I was concerned about being addicted...
want others to perceive you. This is an exercise that can be done daily, weekly or any way that helps you. Don’t be surprised if it changes over time. It will affect your mood. It will also affect the way you both “think” AND “feel” about yourself and others. Also, be aware of how YOU perceive yourself; this will directly affect how others perceive you – unless you are a very good actor.

As someone with multiple autoimmune diseases, I sometimes hear disconcerting comments from colleagues and professionals who I suspect actually know better. One colleague I respect, scratched his bearded chin a few years ago and said to me, “Do you know what your problem is? You don’t look as sick as you are.” How does one respond? Whose ‘problem’ is it exactly? It was more than a comment; it was a total misunderstanding of my world.

All I said at the time was “Thank you.” It seemed clear to me that a true response/explanation would take more energy than I wanted to expend. I also didn’t think this person would understand or that he really wanted to at that time. I had to let it go and move on. This caused some more chin scratching on his part. The comment all of us hear is, “You look good; you must be feeling better.” Where is there a direct link between how chronic illness patients look and how they feel?

These people mean well, which makes the situation difficult. I usually don’t bother to explain things; I do when there is a sincere concern or interest on their part. I don’t want to waste any energy; it is a personal choice. For me, carrying around cards with the IPPF website has been the right choice. I can easily hand it to someone. The ball is now in their court, and they can decide whether or not to take the next step.

Because of my particular profession, more people I know tend to look it up; however, most people will never bother. I am actually fine with it since I have no expectations they will take that next step.

And, I absolutely do not want to look sick. Being understood is always better than being misunderstood, but why would anyone actually want to look sick? Well, believe it or not, there are people who thrive on being ill and helpless. Sometimes it is because of a “Dependent Personality Disorder”, and sometimes it can go back to having had “special” treatment when ill as a child. What are the criteria anyway?

I can have a bit of a “Far Side” sense of humor, and I love to observe other people’s reactions. I tend to listen more than talk. Since I lose my voice easily, this is probably a positive thing for me! I often assign people the task of being a “participant observer” in uncomfortable situations. This can be an eye-opening experience.

Letters and comments that I receive from people and many patients I meet at meetings and presentations say that this is the hardest part for them on an emotional and psychological level. I often ask how they themselves feel when they see someone “who looks totally healthy” using a handicapped space; this is something most people can relate to at some level. This is how others view a chronically ill person with invisible symptoms.

An excellent story and innovative (copyrighted) rationale can be found at: www.butyoudon'tlooksick.com/the_spoon_theory. It is about one woman’s spur of the moment way to explain to a good friend who wanted to understand her illness, but could not.

At our upcoming Annual Patient Conference in Chicago (last weekend of April 2014), you will have the opportunity to see and talk with both expert professionals and others going through physical and emotional changes. You will learn how others have not only coped, but also flourished.

And don’t forget, you can always ask your Peer Health Coach, the Email Discussion Group, or the IPPF Forums and someone will help find an answer for you.

Terry Wolinsky McDonald, PhD, is a former IPPF Board member, PV patient, and a licensed clinical psychologist living in Pittsburgh, Pennsylvania. She is a regular contributor to the Quarterly newsletter in her Psychologically Speaking column.
Thank You!

The IPPF’s Medical Advisory Board (MAB) includes leading names in autoimmune and blistering disease research and treatment. As the IPPF matures, objectives change, and careers wind down, we see turnover and rebirth in the organization and MAB.

The MAB have co-authored papers regarding standardized definitions and outcome measures for pemphigus vulgaris and bullous pemphigoid, and the use of standardized scoring systems like the Pemphigus Disease Area Index (PDAI) and Autoimmune Bullous Skin Disorder Intensity Score (ABSIS). Several of our European members also worked on the European Guidelines for Autoimmune Bullous Diseases for pemphigus diagnosis and treatment, published in 2013.

The IPPF would like to thank those stepping down after years of service. We wish you continued success and added thanks for those who continue serving the IPPF community on various projects.

**DR. RUSSEL J. HALL III** United States
Dr. Hall joined the MAB in 2005 and was a speaker at the 2010 IPPF-NIH Science Meeting.

**DR. TAKASHI HASHIMOTO** Japan
Dr. Hashimoto joined the MAB in 2008 and was a speaker at the 2001 and 2005 IPPF-NIH Science Meetings. He presented at an IPPF Town Hall in 2013 and is active in the Japanese Pemphigus Pemphigoid Foundation.

**PROF. DR. MICHAEL HERTL** Germany
Dr. Hertl joined the MAB in 2008 and was a speaker at the 2001 and 2010 IPPF-NIH Science Meetings.

**DR. PASCAL JOLY** France
Dr. Joly joined the MAB in 2008 and was a speaker at the 2010 IPPF-NIH Science Meeting.

**PROF. DR. MARCEL JONKMAN** Netherlands
Dr. Jonkman joined the MAB in 2005 and was a speaker at the 2005 and 2010 IPPF-NIH Science Meetings, and the 2012 Patient Conference in Boston, MA.

**DR. ROBERT SWERLICK** United States
Dr. Swerlick joined the MAB in 2006.
to pain medication after reading blogs about the disease. Dr. Paice worked with me to form a strategy to reduce the pain to help me not become addicted to pain medication.

During my chemotherapy treatments my levels of the disease fluctuated. I first dropped from 128 to 98, then from 78 to 58, but then spiked during my third month of treatment to over 100. I became discouraged, but I knew I had to stay positive. Within the year, my levels went down to 12 before my last results were classified as undeterminable.

I learned through my initial research that many people dealing with rare diseases which are a mystery to the medical community. It normally takes about a year and 12 doctors to determine what ails them.

It took me three months, ten doctors, and ultimately my diagnosing myself. It also took tremendous faith, determination, hope and belief that I would find my answer. I didn't give up on finding a way to heal myself once I learned what I was facing.

Along the way, I wasn't able to connect with anyone who was going through what I was. I know that could have been healing on its own. My hope is that my story can be an inspiration for others, and I am open to anyone who is going through a similar circumstance to reach out to me.

The larger message I'd like for others to take away is that you must be a true and unwavering advocate for your own health. I told myself at the start of this life-changing journey that I was going to overcome this. And I did, with a steadfast determination and the support and knowledge from a fantastic team of doctors who I was fortunate enough to encounter.

Today, I know there is no playbook for this disease and that it can reappear at any time. I also know I will stay positive and continue my treatment. As Dr. Mehta, my chemo doctor says, “we kill big cancer and we will kill this.” This, and my entire medical team at Northwestern were simply amazing and key in helping me to remain positive.

I never cried while going through this trial of not knowing. I learned what I was up against and did what was necessary to bring about my own healing. However, now when I tell my story to others it is extremely emotional. I hope I can be a beacon of hope for others who might otherwise not know where to turn.

Stu Zirin is an entrepreneur, businessman, and restaurateur foodie living in Chicago, Illinois who has been aggressive in his fight against PV. He does not consider himself a PV patient, but rather a PV survivor. His goal in writing this article is to increase awareness of PV and to give others who are suffering from it the motivation to take control of their health, and the hope that there can be brighter days ahead. Stu can be reached by email at stu@solsyst.com.

REGISTER TODAY AND SAVE!
ONLINE: www.pemphigus.org/2014conference
PHONE: 855-4PEMPHIGUS
MAIL: the form is on page 15

"Everyone should attend the IPPF Patient Conference."
"I never knew there were this many others like me!"
"I keep asking myself, 'Why did I wait so long to attend?'"
"I learned more in a weekend than 8 years of doctor's visits."

This is what past attendees had to say about the Patient Conference.

What will YOU say?
result was strikingly close to 75%, at 76% and at the roof of the vesicle, it was even higher, at 80%. Given the presence of apoptotic cells in the lesion-free patient tissue, the authors concluded that apoptosis is not a late event, but an early one that may cause acantholysis.

Recognizing that the structural damage (acantholysis) and death (apoptosis) of keratinocytes are mediated by the same molecular players – the caspase enzymes. Research led by Sergei Grando has proposed a novel theory of “apoptolysis”, combining the two terms. The work of Deyhimi and Tavakoli supports this model and suggests that once a threshold level of apoptotic cells exist in the basal cell layer, somewhere north of 80%, then a lesion will form. According to the authors, conventional therapy of PV consisting of high-dose corticosteroids is based on the hypothesis that acantholysis leads to apoptosis, so it will be critical to unravel the current results and to determine if treatments might be tailored differently in the future.

How apoptosis leads to formation of blisters and how antibodies to desmogleins may promote apoptosis is still under investigation, but one additional piece of information from the current work is that based on the absence of another cell death marker, Bax, the authors suspect the extrinsic cell death pathway.

The pieces to the pemphigus puzzle are beginning to be unraveled. Driven by the fact that the more we learn about the molecular events leading up to blisters, the more chances there will be to intervene before debilitating blisters can occur.

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.

Figure 1. The layers of the epidermis that are affected in pemphigus vulgaris. a) In normal tissue (left), the keratinocytes are joined by desmoglein and other junction proteins. In pemphigus vulgaris (right), suprabasal vesicles are formed, resembling a tear in the skin. b) Ultrastructural techniques help to visualize human tissue from normal (left) and PV (right) tissue. PV is characterized by suprabasal acantholysis, where rows of tombstone basal keratinocytes and half-desmosomes are visibly carved out by the vesicle. The image in a is adapted from Jens Waschke Histochemistry and Cell Biology 2008, Volume 130, pp 21-54. The normal tissue shown on the right is from http://commons.wikimedia.org/wiki/File:Normal_Epidermis_and_Dermis_with_Intradermal_Nevus_10x-cropped.JPG and the PV patient tissue shown in b is from Dr. Sergei Grando. Note that the coloration represents different stains and is not reflective of true differences between the tissues. The differences of note are structural only.

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A portion of your registration may be tax-deductible. Please consult with your tax professional.

**CASINO NIGHT GAMES ARE FOR ENTERTAINMENT PURPOSES ONLY.**

### Patient Conference Registration Fees  *Discounted pricing valid during March 2014 only*

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**I WANT IT ALL! Complete Conference Package:** $200.00  **$160.00** per person

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**Conference Registration Only:** $125.00  **$100.00** per person

You will be registered for all plenary sessions and workshops, Friday’s Welcome Reception, Saturday’s sessions, Sunday’s sessions and workshops. Also includes light breakfast on Saturday and Sunday, all breaks, and any conference materials.

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**20th Anniversary & Awards Dinner Only:** $125.00 per person

Can’t make the meeting but still want to enjoy great food, friends, and fun? Join us at our 20th Anniversary and Awards Dinner (Casino Night with entertainment and prizes). Roll the dice, spin the wheel, or double down with proceeds benefiting the IPPF’s Patient Support programs.

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### HOW TO REGISTER

1. **Online:** [www.pemphigus.org/2014conference](http://www.pemphigus.org/2014conference)

   Use discount code ADAMI4 during March!

2. **Phone:** (855) 4Pemphigus (855-473-6744)

3. Contact your Peer Health Coach

4. Mail this completed form with payment to:

   IPPF 2014 Patient Conference
   1331 Garden Highway #100
   Sacramento CA 95833

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### OTHER ATTENDEES

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* PV, PF, PNP, BP, OCP, MMP, Caregiver, Family, Friend, Physician, etc.

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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 **March 14, 2014**, we will refund registration costs less $25 administrative fee. Due to the contractual agreements such as food, beverage and material costs, cancellations postmarked between April 15 and April 10, 2014 may be refunded up to 50% of the registration fees. **We may be unable to make refunds after April 11, 2014. We will gladly transfer your registration to another person (scholarship) or credit it as a donation.**
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