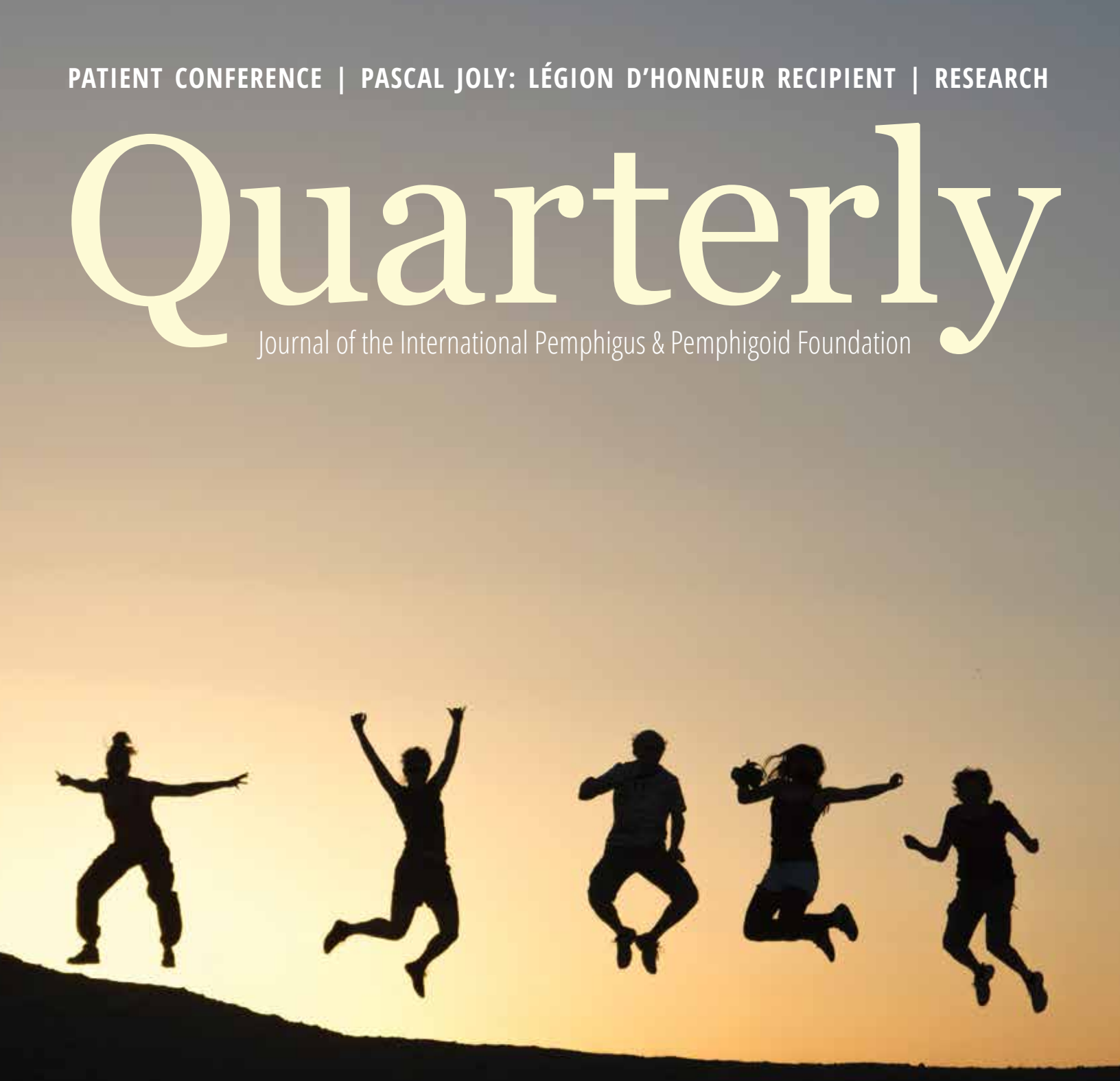
PATIENT CONFERENCE | PASCAL JOLY: LÉGION D'HONNEUR RECIPIENT | RESEARCH

Quarterly

Journal of the International Pemphigus & Pemphigoid Foundation



Forging ahead

OUR PEMPHIGUS & PEMPHIGOID COMMUNITY

FALL 2019 • ISSUE #98

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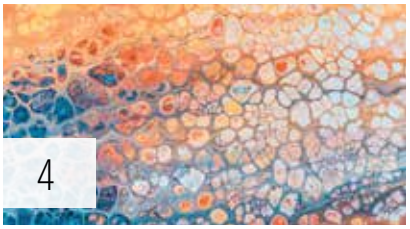
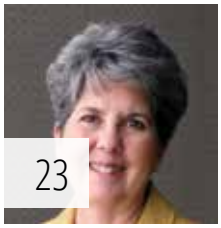
FALL 2019

SECTIONS

FEATURES





Quarterly

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ISSUE #98 | FALL 2019

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INTERNATIONAL PEMPHIGUS & PEMPHIGOID FOUNDATION

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Message from the Executive Director

As 2019 draws to a close, I can't help but think about all the progress the IPPF has made this year. Our small, but mighty, community has increased disease recognition and knowledge through public advocacy events, our "Biopsies Saves Lives" campaign, research and drug development partnerships, and our Patient Education Conference. The IPPF has strengthened existing collaborations and forged new relationships across the globe, providing hope for the future of our community.

Since the Foundation's humble beginning 25 years ago, we have sought to make a difference in the lives of those affected by pemphigus and pemphigoid. Through all our efforts and the commitment of so many, the IPPF community continues to grow stronger. However, we still have so much to accomplish. We can all agree that there is still more that we don't understand about theses diseases than what we do understand; we need to get people diagnosed faster; we need better treatments that don't wreak havoc on our overall health; we need therapies that don't severely impact our quality of life; we need to put pemphigus and pemphigoid "on the map;" and we need more medical research. Only together can we meet these challenges head on and take the actions needed to **find a cure for pemphigus and pemphigoid!**

I hope that this edition of the *Quarterly* shows you what being part of the IPPF community is all about. I encourage you to become more involved with the IPPF by becoming an advocate or ambassador, attending a support group, or by joining the IPPF Natural History Study so our community can learn more about these diseases. Our community is the thread that connects patients, caregivers, researchers, and physicians. With your help, we can build on what we have already accomplished. By working together, we can make our community even stronger!

Gratefully,

Marc 2 Yale

Marc Yale
IPPF Executive Director and MMP Patient
marc@pemphigus.org

IPPF Research Grant Update

Jens Wrammert and Ron Feldman

We are excited to share the results from our recent published report in *Cell Reports* entitled "Single-Cell Analysis Suggests that Ongoing Affinity Maturation Drives the Emergence of Pemphigus Vulgaris Autoimmune Disease." These data were generated partly from the generosity of an IPPF research grant to explore novel pathways in pemphigus.

You may be aware that pemphigus is caused by destruction of the skin cells by proteins called antibodies. These antibodies are generated from a specific immune cell called a B cell. These B cells are generated in the bone marrow and further mature in other parts of what is termed the peripheral immune system, such as the spleen, lymph nodes, and, potentially, the skin. The B cells are an integral part of your normal immune response to infection, for they can rapidly generate high levels of protective antibodies against multiple pathogens, such as viruses. For example, when you are vaccinated for the flu, your immune system is stimulated to generate a protective response in case you ever encounter the real flu virus. It does this partly by generating long lived memory B cells. Upon exposure to the flu, these memory B cells expand quickly and produce large amounts of anti-flu antibodies to rapidly clear the infection. In essence, the B cells *learn* from exposure to various stimuli, and they can mature and selectively eliminate those various dangerous stimuli. Conversely in pemphigus, this same B-cell immune response is essentially hijacked, and instead of protecting you, the B cells are now primed to produce large amounts of antibodies against proteins that are critical for holding your skin cells together, including desmoglein 3. While we are familiar with the clinical symptoms of pemphigus, we are still at the early stages of understanding how these pemphigus-specific B cells develop, mature, produce antibodies, and ultimately where they reside in patients.

Our study provides evidence that memory B cells specific to desmoglein 3 (dsg3 mbcs) can be detected in the blood of patients prior to treatment and can expand in patients who undergo clinical relapse. In fact, we were fortunate enough to capture a blood sample on a patient 15 months prior to diagnosis of pemphigus. Surprisingly, we were able to detect dsg3 mbcs that generated low levels of antibodies that were not able to cause damage to skin cells. These dsg3 mbcs then began to learn and were able to mature enough to produce disease causing antibodies. These antibodies were

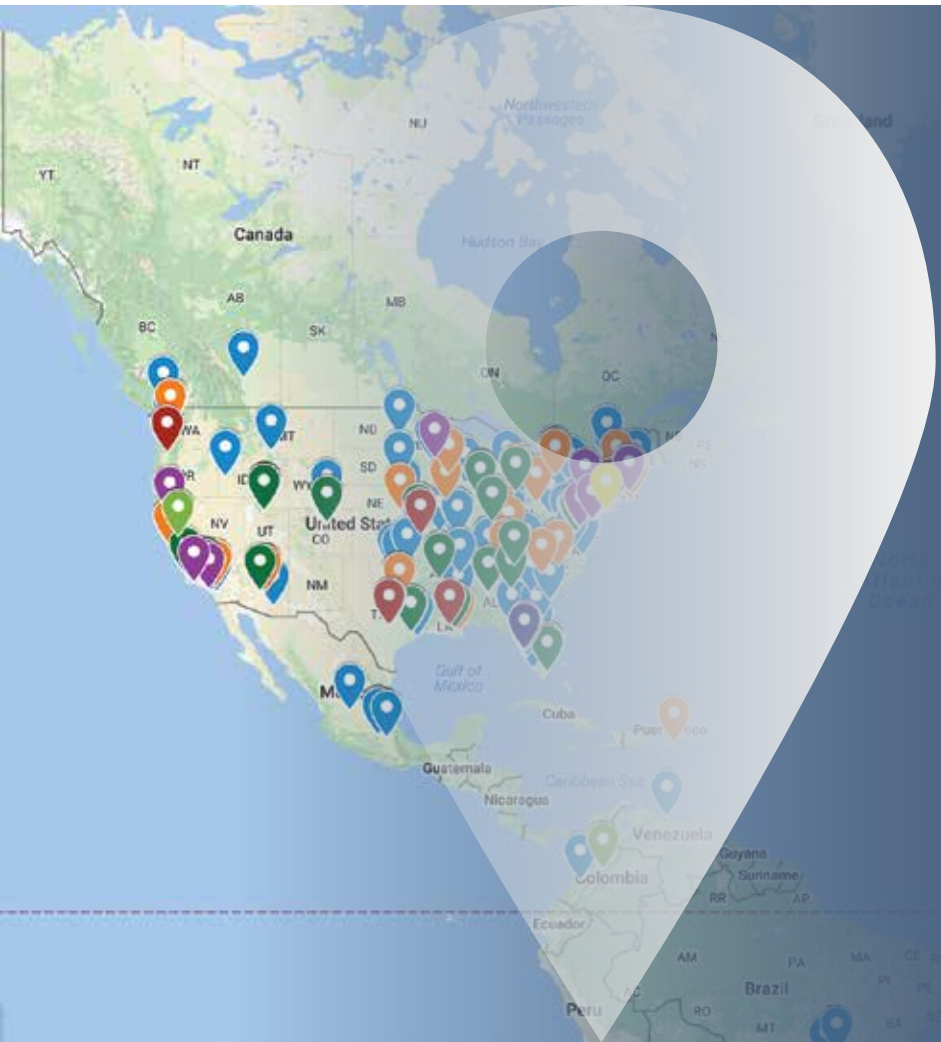
very efficient and were able to target multiple parts of the desmoglein, thereby resulting in full blown pemphigus 15 months later. In fact, the antibodies bound the previously described parts of desmoglein 3 protein in addition to a unique domain which has not been previously considered disease-causing.

This work provides unique insight into how these pemphigus antibodies are generated and eventually result in damage to the skin cells. Further understanding of the various B-cell subsets, where they normally reside, how they traffic in disease, and how they interact with the rest of the immune system is critical as new therapies emerge targeting B cells. This is especially relevant given the recent FDA approval of the B-cell depleting therapy, rituximab, and the positive results of the phase III PEMPHIX trial demonstrating the superior efficacy of rituximab therapy over conventional immune-suppressive therapies. We are hopeful that future studies will elucidate more of these B-cell directed pathways and are so appreciative of the ongoing support from the IPPF and blistering community to ensure that research continues to move forward in the quest for a cure for pemphigus.

The following doctors contributed to the article in *Cell Reports*: Alice Cho (Emory University), Amber L. Caldara (Emory University), Nina A. Ran (University of Pennsylvania), Zach Menne (Emory University), Robert C. Kauffman (Emory University), Maurizio Affer (Emory University), Alexandra Llovet (Emory University), Carson Norwood (Emory University), Aaron Scanlan (Emory University), Grace Mantus (Emory University), Bridget Bradley (Emory University), Stephanie Zimmer (Emory University), Thomas Schmidt (Philipps-University), Aimee S. Payne (University of Pennsylvania), Ron Feldman (Emory University), Andrew P. Kowalczyk (Emory University), Jens Wrammert (Emory University).

Jens Wrammert, PhD, is an Assistant Professor in the Department of Microbiology and Immunology at Emory University School of Medicine.

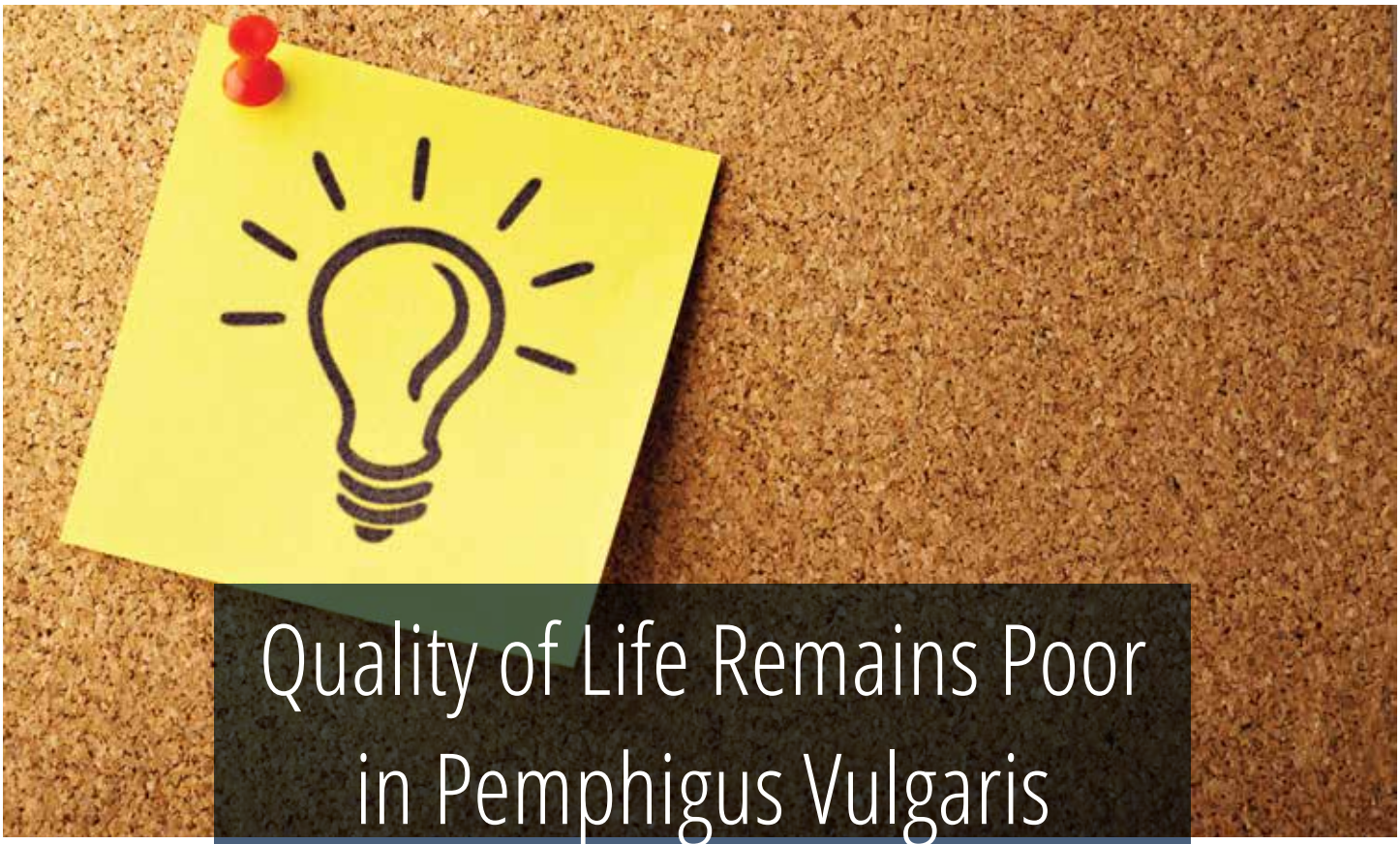
Ron Feldman, MD, PhD, is an Assistant Professor in the Department of Dermatology at Emory University School of Medicine and the Director of the Autoimmune Blistering Disease Clinic.



Find a Doctor: IPPF Physician Map

The IPPF's ***Find a Doctor*** P/P physician map provides patients with contact information for medical and dental professionals familiar with P/P.

Access the map online at:
www.pemphigus.org/find-a-doctor



Quality of Life Remains Poor in Pemphigus Vulgaris

Brittney Schultz and Nicole Fett

Last year, we partnered with the IPPF to study quality of life in patients with pemphigus vulgaris (PV). Given the targeted mechanism and efficacy of rituximab, which has emerged via expert opinion as first-line treatment for PV, we were specifically interested in how quality of life differed amongst patients who had received rituximab versus patients who had never received rituximab in treatment of PV.

In our study, we sent surveys to the IPPF community. Survey questions included basic demographic information, areas of pemphigus involvement, self-reported severity of disease, details of treatment history, and two validated questionnaires used to study quality of life in autoimmune blistering disease: The Autoimmune Bullous Disease Quality of Life (ABQOL) and the Treatment of Autoimmune Bullous Disease Quality of Life (TABQOL). The ABQOL asks questions about how the disease itself affects quality of life while the TABQOL asks questions about how the treatments for the disease affect quality of life. We then compared the responses of patients who had previously received rituximab to those who had never received rituximab to see if any differences were present. Over 200 responses were received.

> We also found that quality of life was associated with disease severity, regardless of rituximab use.

Overall, we did not find a difference in quality of life amongst patients treated with rituximab versus those who had never received rituximab. However, in all patients, quality of life was significantly affected. We also found that quality of life was associated with

disease severity, regardless of rituximab use. Patients with more severe disease had worse quality of life, but patients with less severe disease also had impaired quality of life. There were no differences in hospitalization or the need to stop medication between those who had been treated with rituximab versus those who had never received rituximab.

The main conclusions we drew from our study were: 1) Quality of life remains poor in patients with PV; and 2) While rituximab was as well-tolerated as other treatments, it did not definitively improve quality of life compared to other treatments despite its targeted nature. It may be that the questionnaires are not sensitive enough to detect differences between the patient groups, that our current treatments are not increasing quality of life, or that the effects of disease severity on quality of life overshadow the potential benefits of therapeutics such as rituximab. Going forward, we need additional studies to discover ways in which we can improve quality of life in patients with PV. Future studies that follow patients over time, rather than asking them to remember details in the past, would be helpful.

We would like to acknowledge the hard work and support of Becky Strong (IPPF Outreach Director) and the IPPF for their assistance with this project. We are also grateful to the Medical Dermatology Society, which awarded Brittney Schultz a mentorship with Nicole Fett, as well as to the authors of the ABQOL and TABQOL for sharing the questionnaires. Finally, a big thank you to the members of the IPPF community who completed this survey, as this research would not have been possible without you!

Brittney Schultz is an Assistant Professor of Dermatology at the University of Minnesota and a Staff Dermatologist at the Minneapolis VA Medical Center. She is the Director of the Autoimmune Blistering Diseases Clinic at the U of M and is passionate about her patients.

Nicole Fett is an Associate Professor of Dermatology at Oregon Health and Science University and Portland VA Medical Center. She is the Dermatology Residency Program Director and co-director of the Rheumatology-Dermatology Clinic at OHSU.

Chasing the Cure Together

Terry Wolinsky McDonald, PhD

There is a new television program called Chasing the Cure, and I may just be addicted at this point. It addresses real people with real medical conditions. Sometimes they have experienced decades-long debilitating symptoms that have been presented to numerous physicians—both generalists and specialists. (For P/P patients, we almost certainly wouldn't be alive if we had to wait that long for a diagnosis.)

This television show, anchored by Ann Curry, presents these people to the show's resident specialists, and then opens the case up to social media, both in the moment and for future follow-up. Patients often haven't received adequate medical answers or compassion before appearing on the show.

Recently, the show aired two cases in which the patients were both diagnosed with a disease that fewer than 10 people in the world are known to have. Then, amazingly, they found yet another person with the same symptoms and diagnosis through social media. When they found out that they weren't the only ones with their diseases, they discovered a whole new perspective. They knew they weren't alone, which is important psychologically and emotionally.

Those of us with P/P are familiar with the pain, frustration, isolation, and emotional distress associated with the road to diagnosis. And after we receive our diagnosis, we may have mixed feelings. There is often a feeling of relief to have a name for our frightening and painful symptoms and to know we're not infectious to our loved ones. We also may go online to learn more about these diseases, all while still learning how to pronounce their names. We know we will likely need to make permanent adjustments to our everyday lives.

The idea of not having to go through a rare orphan disease alone is powerful to experience (and to watch). I don't think that any of us who are part of the IPPF community, especially us old timers, take the idea and reality of community for granted. Some of us have had major problems getting diagnosed (though this has improved with education). However, once we know our diagnosis, we have knowledge, compassion, and help from the P/P community. None of us needs to feel alone anymore on our personal journeys, and providing support for others helps us all.

I was finally diagnosed with pemphigus vulgaris (PV) shortly before Thanksgiving in 2001, so this particular time of year is especially meaningful. I am grateful every day, but Thanksgiving remains an incredibly special time for me. I had trouble swallowing water that first Thanksgiving and had to put sweet potatoes into a blender. At the time, my hope came from another PV patient who eloquently wrote on a blog about his own first Thanksgiving, how he moved on, and even how he went into remission.

Yes, we are still chasing the cure, but research and quality-of-life progress has come a long way. None of us needs to face P/P alone. Our community is thriving. All of us have something to offer. Don't ever sell yourself or our community short, and be grateful for what has been accomplished and for what is yet to come.

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 in her "Psychologically Speaking" column.

DEALING WITH PEMPHIGUS?

THE PEGASUS TRIAL IN PEMPHIGUS VULGARIS (PV) AND PEMPHIGUS FOLIACEUS (PF) IS NOW ENROLLING

PEGASUS

Principia Biopharma is conducting a Phase 3 clinical trial of a new investigational drug (PRN1008, an oral pill). PRN1008 is a Bruton's tyrosine kinase (BTK) inhibitor which may improve pemphigus. This trial is being done to see if PRN1008 is safe and can achieve disease remission (clear skin with no blisters) with reduced corticosteroids.

PEGASUS TRIAL DETAILS

- Over 18 years of age
- Newly diagnosed or relapsing PV and PF patients with moderate to severe disease
- Length of the trial for each patient is about 17 months
- Reimbursement for travel may be available (talk to your Trial Doctor)
- Clinical Sites are open globally, including US & Canada
- ClinicalTrials.gov (NCT03762265)

FIND OUT ABOUT A CLINICAL TRIAL SITE IN YOUR AREA

EMAIL: clinicaltrials@principiabio.com
PHONE: 833-477-6700

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Looking Back at the 2019 Patient Education Conference

The 2019 Patient Education Conference is in the books! All of us at the IPPF extend our gratitude to the doctors, speakers, and sponsors for making this event possible.

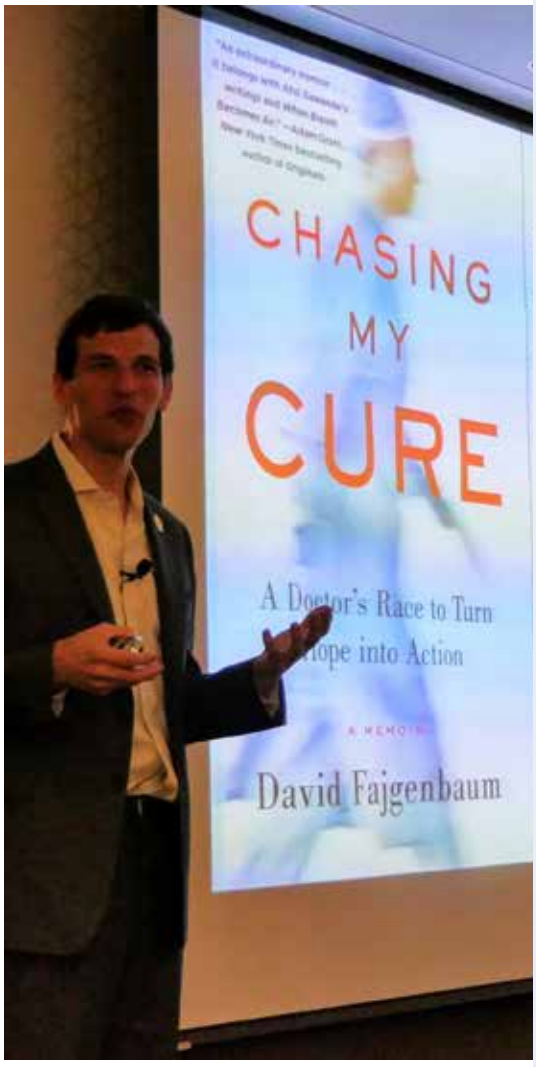
We would especially like to thank our hosts—Dr. Aimee Payne and Dr. Victoria Werth—as well as the patients and caregivers who attended. YOU are the reason that this year's conference was our best yet.

We hope to see as many of you as possible next year at our 2020 Patient Education Conference in Las Vegas.

FRIDAY: WELCOME RECEPTION & BOOK SIGNING








All Patient Education Conference photos by Jim Manley Photography.

SATURDAY: CONFERENCE SESSIONS

All Patient Education Conference photos by Jim Manley.

SATURDAY: AWARDS DINNER & CASINO NIGHT

All Patient Education Conference photos by Jim Manley.

SUNDAY: HISTORIC WALKING TOUR

IPPF AWARDS DINNER

The IPPF hosts an Awards Dinner to honor those whose service to our community has made a significant impact over the previous year. It's a great opportunity for all those affected by pemphigus and pemphigoid to come together in celebration of each other. Congratulations to our 2019 Award Winners:

LIFETIME ACHIEVEMENT AWARD
Dr. Grant Anhalt

FOUNDER'S AWARD
Marcy Syms

DOCTOR OF THE YEAR
Dr. Pascal Joly

DENTAL PROFESSIONALS OF THE YEAR
Jennifer Harmon
Dr. Ricardo Padilla

BRIGHT STAR AWARD
Nelly Filippov

STAR AWARD: EDUCATION
David Baron

STAR AWARD: OUTREACH
Amethyst Yale

STAR AWARD: PATIENT SUPPORT
Anna Lane

VOLUNTEER OF THE YEAR
Mary Lee Jackson

HEALING HERO OF THE YEAR
Dr. Aimee Payne

THANKS TO OUR 2019 CONFERENCE SPONSORS

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Patty's Story

Carola Pulvirenti

I've been a professional nurse in Italy since 2003, and until a few years ago I didn't have much knowledge of autoimmune bullous diseases. In 2016, I received a text message from my 45-year-old Aunt Patty. She asked me if I could refer someone that did wound care at home. I told her I could and asked her what kind of wound it was. When she replied "pemphigus," I asked her to send me a picture since I had never heard of it. As soon as I saw the photo, I realized that the situation was serious and that I needed to evaluate it in person.

I visited Patty's home the next day, and her sisters answered the door. When I entered, I smelled a strong odor. They took me to her bedroom, and my aunt was almost unrecognizable. She had been lying in bed for 15 days, and she had wounds on 90 percent of her body. Her skin was sticking to her clothes and the sheets, she was thin, and she couldn't eat because of the wounds in her mouth. She was not able to perform her daily activities, and she was dehydrated. I had never seen anything like it.

First, I sat down to speak to Patty and her two sisters about the clinical situation. Patty developed her first symptoms in 2006. Skin spots appeared on her chest, then the skin peeled off and left wounds that would not heal. Despite living in a big city with many prominent hospitals, it took a year for her to receive a diagnosis of pemphigus vulgaris (PV).

After seven years of corticosteroid treatment, she could no longer tolerate the side effects. So she refused corticosteroids (both tablets and ointment) for her wounds. She made the decision to experiment with

alternative therapies based on dietary restrictions and medical herbalism. Patty considered drugs harmful to her health. However, she continued taking pain medication due to the strong pain she felt.

While visiting Patty, I provided directions on wound care and diet. I explained to her that taking herbs with a painkiller to replace corticosteroids would not give her any health benefits. I reminded her about the importance of including protein in her diet, which would help her body heal and exudate her wounds. I advised her to supplement her vegan diet at least with fish.

When I returned home, I researched pemphigus on PubMed. (PubMed comprises more than 30 million citations for biomedical literature from MEDLINE, life science journals, and online books.) I understood that wound care was not enough and medical therapy was necessary. That night, I could not sleep. I thought about Patty and her skinless body. She was left without any protection from infections and was in strong, constant pain. She wasn't able to go to work, do her daily chores, or even hug her loved ones.

When I searched the internet for other people's experiences with pemphigus, I found the National Pemphigus/Pemphigoid Association Italy (ANPPI). The next day, as I went to work and picked up my children at school, my thoughts were focused on Patty. I wondered how I could help her and felt powerless. I decided to contact ANPPI's vice president for advice on how to handle the difficult situation.

I bought Patty non-adherent clothes at the pharmacy and told her we had to go to the hospital for treatment. Patty refused to go, so I looked for a dermatologist that also specialized in homeopathy, and he agreed to visit her at home right away. The dermatologist was shocked by Patty's condition and explained that alternative medicine could offer her support, but not save her life.

The doctor and I counseled Patty for a long time about therapeutic adherence, alternative medicine, and PV. After many hours, Patty was persuaded to receive treatment, and the doctor organized the hospitalization in a specialized dermatological institute. Patty was completely bandaged and taken to the hospital by ambulance. As soon as she received the appropriate treatment, she immediately felt great pain relief. Later, she was treated with biological immunotherapy that put her in remission. Patty is still in remission today and does not take any medication.

My experience with Patty was shocking. *Why did it happen? Why does an informed young woman who lives in a big city decide to refuse the prescribed therapy?* I continued gathering information on autoimmune bullous diseases and learned that they are not well-known by many people. I learned that P/P patients have many unmet needs, and this has motivated me to dedicate myself to them. I signed up for the ANPPI and worked hard. I contacted patients, doctors, and researchers with very specific objectives—to increase P/P awareness, strengthen patient resources, engage them in research projects, and make disease and treatments more tolerable in everyday life.

When we are sick, we are defenseless and need someone to take care of us—someone like the ANPPI.

In December 2018, I was appointed as the vice president of the ANPPI. I am happy to work with a team of patients and professionals who share the same enthusiasm for supporting people with these rare diseases and their families. When we are sick, we are defenseless and need someone to take care of us—someone like the ANPPI. When we are healthy, we should take care of others. In the future I don't want any P/P patients to feel misunderstood or alone.

Carola Pulvirenti works at the National Institute for Infectious Diseases in Rome, Italy and was elected Vice President of the National Pemphigus/Pemphigoid Association Italy in 2018. She lives in Rome with her family. The ANPPI can be found online at ***www.pemfigo.it***

Pascal Joly

Légion d'Honneur Recipient

Association Pemphigus Pemphigoïde France

On July 14, 2019, at the *Elysée* Palace, Professor Pascal Joly was awarded France's highest distinction, the *Légion d'Honneur*, for his work on rituximab applied to moderate and severe pemphigus. Instituted on May 19, 1802, by Napoleon Bonaparte, the *Légion d'Honneur* was part of the total restructuring of French society following the French Revolution of 1789 in which both the royal administration and the privileges of the nobility had been abolished. This new distinction was put in place to reward a few exceptional citizens, scientists, and members of the military who provided outstanding services to the nation and whose actions contributed to the common good of society.

Professor Joly's nomination resulted from letters written to the Health Minister, Madame Agnés Buzyn, by his colleagues—foreign colleagues in particular—and by patient organizations such as the IPPF and the Association Pemphigus Pemphigoïde France (APPF). These letters recommended him for recognition of his work and research on rituximab and for the improvement this work had on the quality of life of patients suffering from pemphigus.

In light of his award, Josée de Felice and Laurence Gallu from the APPF met with Professor Joly and asked him a series of questions dealing with topics that don't usually come up in the patient-doctor relationship: questions about his journey as a doctor, the research progress on rituximab for pemphigus, and the results emerging from this new research.

Our conversation revolved around two areas: the path of Professor Joly's medical career and the "how and why" of his work with rituximab. The process of getting a new medication approved for a disease is quite long. In this case, it took 15 years of clinical trials, publications, and conferences to get rituximab approved for pemphigus by drug agencies. We also discussed the treatment of blistering diseases in France, including the particularity of our medical system whereby the treatment of related diseases has been regrouped in Expert Centers to accelerate diagnosis, as well as the contribution of the national study group—the "Blister Group"—and its collaboration in rituximab research.

Professor Joly is 60 years old, and both his parents were doctors. While an intern in rheumatology, he met Professor Lauret, head dermatologist at the Hospital Center University De Rouen, who told him he would give him a research lab at the Hôpital Henri-Mondor. Accordingly, at 26 years old, he became the head of this lab, researching HIV. There he met Professor Catherine Prost, and she oriented him toward autoimmune blistering skin diseases (AIBD) to see what links could be established between AIBD and immunology. Professor Joly says he owes his career to Professors Lauret and Prost. He has thus been involved with AIBD since his internship as a medical student.

Professor Joly then did his military service at the *Val-de-Grâce* Hospital (a military hospital in Paris) working in immunology. At 34, he was appointed to the Hospital Center University *De Rouen* as University Professor, both teaching and leading research while working as a practitioner caring for patients. He became head of the dermatology department at 42 years old and has remained in Rouen ever since. Of course, his career took him outside of Rouen as he often had to travel the world to give lectures on his research and breakthrough findings in AIBD, which have led to the approval of the biological medicine rituximab by both

the United States Food and Drug Administration and the European Medicine's Agency.

Rituximab was already in use for non-Hodgkin's lymphoma, but Professor Borradori of Bern, Switzerland, thought of using it to treat autoimmune diseases (such as lupus and polyarthritis). He was the first doctor to do so successfully in a child who had both B cell lymphoma and pemphigus. Rituximab had already been approved for the treatment of B cell lymphoma for some time and was working well. Since it was starting to be used for autoimmune diseases, Professor Joly and his collaborative study group (see below) thought to use it for pemphigus and set up the first clinical trial. A few more clinical trials would follow.

As mentioned earlier, rituximab has totally changed the lives of patients suffering from pemphigus. Prior to its use, treatment of pemphigus consisted of corticosteroids, often in high doses, and non-targeted immunosuppressants, which could result in muscle loss; osteoporosis; diabetes; glaucoma; repeated infections; heart, liver, and kidney problems; and other conditions. The adverse effects of this kind of treatment could lead to severe disabilities in family, social, and work life. Rituximab infusions allow patients to quickly stop corticosteroids and to avoid taking non-targeted immunosuppressants; therefore, they don't have to suffer from the procession of secondary diseases and side effects from earlier treatment protocols.

We know that rituximab is also an immunosuppressant, but it targets the CD20 B lymphocytes that produce the antibodies that attack skin and mucous membranes, killing a large number of them. We also know that it is not without risk and can lead to serious infections and Progressive Multifocal Leukoencephalopathy (PML). But the precautions taken before every treatment, as well as the 15 years of experience using the drug for other diseases, are very reassuring. For us in France, the experience has been very positive, and the use of biosimilar rituximab drugs has not been a problem.

Professor Joly is not alone in France working on AIBD, researching new treatments and ways to improve patients' lives. All the AIBD dermatologists working in the public health sector belong to a research group, called the "*Groupe Bulle*"—literally translated as the "Blister Group." This group has been in existence since the 1980s and meets several times a year to discuss and compare research. The APPF's president and vice president are invited to its General Assembly. The "*Groupe Bulle*" was instrumental in helping orchestrate and design the clinical trials put together by Professor Joly, as well as helping him gather the necessary patients. It should be noted that Professor Catherine Prost (Avicenne Hospital) is a key component of Professor Joly's research and success. She participated in the design of the Ritux 3 study, the Pemphix study and the bullous pemphigoid and mucous membrane pemphigoid studies being conducted right now with Professors Joly and Caux. She has also referred the greatest number of patients to several successive rituximab clinical trials. Professor Caux is the active president of the study group previously presided over by Professor Joly.

[I]t was Professor Joly who . . . fought to get the rituximab breakthrough treatment finally approved for all who suffer from pemphigus in the world.

Another French structure that has acted as an effective tool in helping AIBD doctors communicate with each other across the country to share research results and refer patients for clinical trials is the combination of four "Expert Centers" and a network of "Competence Centers." These act to group the AIBD dermatologists working in the public sector. It is thanks to them that rituximab is made available to patients.

The *Légion d'Honneur* is therefore in recognition of Professor Joly's efforts, successes, and breakthroughs with rituximab—and indirectly of the collaborative work put in place by our AIBD dermatologists, by the "*Groupe Bulle*," and the "Expert Centers." But it was Professor Joly who orchestrated the work and relentlessly fought to get the rituximab breakthrough treatment finally approved for all who suffer from pemphigus in the world. *So, thank you, thank you, thank you, Professor Joly!*

The Association Pemphigus Pemphigoïde France provides information, contacts, and support for people living with pemphigus or pemphigoid, their relatives, friends, and people who help them to live better with the disease and to try to offset the side effects of medications. ***www.pemphigus.asso.fr***

The Gift of Growing Old

Becky Strong

Awhile ago, I downloaded a popular app that allows you to see what you might look like when you are older. Though it was in the spirit of fun, impatience set in as I anxiously waited for my phone to download this app. Once it downloaded, I quickly opened the app and snapped my selfie. Let the editing begin! (I did pause for a moment to ensure the accuracy of the photo, to be confident my current laugh lines weren't washed out by the brightness of the room, nor were they more pronounced due to the shadows.)

I started to edit, first looking at my smile. I added bangs and changed my hairstyle. Then I went to my original photo and clicked on the age button. First, I tried the "young" button. *Yes!* I looked like me in college, so I figured it must be fairly accurate. Then I clicked on the "old" button. *Yikes!*

I looked older, but I thought I still looked good. I had more wrinkles, more frown and laugh lines, and my teeth were a bit yellowed. I chuckled because I thought with the placement of the wrinkles, I still wouldn't be able to control my eyebrows and their movement in my old age. I enjoyed the chance to look into my future with every bit of light and happy heart I thought I would. Then, another thought went through my mind. *I never thought I would have the chance to get old. I look pretty good for a senior citizen, and I hope to heck I have the chance to get even more wrinkles!*

I was in my early thirties when my first symptoms of pemphigus appeared. Slowly, the lesions spread. The pain got worse, and I could feel the unknown disease taking over my body. As I was looking at the older version of myself on my phone, I was taken back to a time when I wasn't sure if I would ever know what was happening to me or if I would make it through alive.

That photo took me to a time when my husband, Tim, and I discussed what was happening to me and the toll it was taking on our very young marriage. We sat down and had a legitimate heart-to-heart. I made

it very clear that if what was happening ever got to be too much for him, he could leave me, and I would be okay with that. That photo then transported me to a time a few years later when Tim admitted to me that he thought he was watching his new wife die, and he loved me so much that he would never let me die alone. That photo was a blinding reminder of a time when I thought all of my hopes and dreams were going to be completely wiped away like a sandcastle on the beach with high tide approaching. What I thought was going to be a very funny and carefree moment filled with laughter suddenly turned to tears. Tears of loss, but also tears of gratitude.

I couldn't believe I lived with those feelings for 17 months before finding a doctor who was able to diagnose me and change my bleak future. At that moment, I was grateful for the doctors who treated me, who answered my never-ending lines of questions, answered their personal cell phones because I wasn't done asking questions, and would stay with me for hour-long doctor appointments in tiny exam rooms to explain it one more time. I couldn't look away from my older self. I examined every line and wrinkle, and I had an unsurmountable appreciation and hope that I will one day look like that lady in my phone. The lines and wrinkles will represent happy, angry, and sad times—each a line of memories I will hold dear or wish to forget.

Looking at the photo on my phone, I realized how fragile my life was and how I came close to not having the blessing of being old. I'm lucky to currently be in remission. I now live each moment loudly and look forward to the future. I appreciate that my hair has come in gray and my skin has scars. I love each of my current laugh lines and wrinkles. They are from moments and memories that I was afraid I wouldn't have.

So, yes, the app made me look funny. It made my skin sag and gave me age spots. But I cannot wait to compare that photo to my older self after my children are grown and even after a pemphigus flare or two. I'm sure I'll have another good laugh as I compare reality to that "old age" photo and share it with my children and grandchildren.

Becky Strong is the IPPF Outreach Director. She was diagnosed with PV in 2010 and is currently in remission. She lives in Michigan with her family.

Looking for a Support Group?

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 Becky Strong: **becky@pemphigus.org**.

Find more Patient Support Group locations and dates at **pemphigus.org**

A Reason for Hope in the IPPF Community

Carolyn Fota

Community is defined by Merriam-Webster as "a unified body of individuals who share common interests while living in a particular physical location or region." In previous centuries, people didn't often move far from home; generations of families attended the same school, place of worship, grocery store, and were even buried in the same cemetery. They connected to their local communities because they were born and raised in that location. It's also important to point out that people could only use local resources due to limited transportation, financial resources, and (especially) information. However, a new paradigm has shifted the traditional concept of community and made radical changes. I believe these changes present incredible opportunities to the IPPF community.

The 21st century brought a wave of information technology growth and development that resulted in easy-to-download personal software (apps), hardware (laptops and tablets), and mobile phones (I literally tracked a NASA liftoff this spring). We can now transport ourselves through Uber, visit friends around the world through online groups, and attend virtual meetings. We can check our medical records online and track our health through smart watches, all while ordering food, clothes, and books. We can talk to people halfway around the world by saying, "Alexa, call my friends in Ireland."

Modern communication has also made it possible for people to select their own communities. We no longer need to passively accept the idea that community is limited to the location in which we were raised. Technology, along with societal shifts that support

independence, allows us to explore communities we are interested in based on where and with whom we identify.

It can be difficult to find a balance between traditional and modern concepts of community. For pemphigus and pemphigoid patients who are already facing challenges due to their rare conditions, the IPPF offers outstanding resources that can be accessed in both traditional and modern ways. For those of us (myself included) who tend to stick more to our location-based communities, we can engage with the IPPF in more traditional ways, such as by attending local support groups, talking with a peer health coach over the phone, and attending a Patient Education Conference. Additionally, you may want to consider the wealth of information available on the IPPF website and social media platforms, as well as Patient Education Series webinars.

For the modern communicator who actively identifies with various communities and is more comfortable with technology, you may already have connected with the IPPF through social media, have an ongoing email conversation with a peer health coach, and follow PemPress on your phone's news app. Remember to still connect with people in real life along the way, even people outside of your usual zone, as making and keeping relationships is important.

We are drawn to the IPPF to build a community that shares the common experience of living with a rare and serious illness. Even though there is not currently a cure, you don't have to feel alone, isolated, or scared. We are all part of a community, and that community is what you make of it. Whether you connect through traditional or modern ways—or a combination of the two—it's up to you.

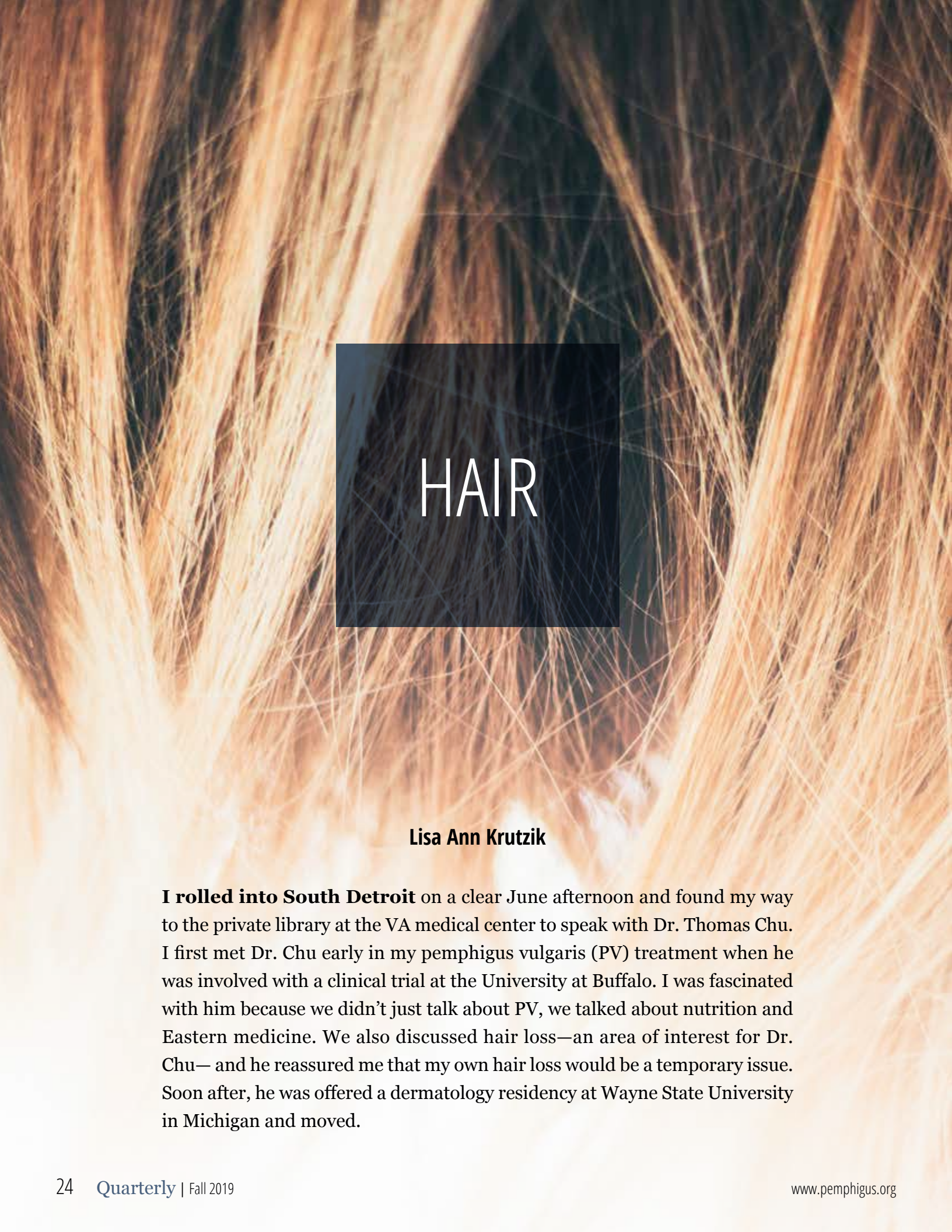
In February of 2016, I called Marc Yale, who was an IPPF Peer Health Coach at the time. I shared my story of delayed diagnosis, hospitalizations due to bullous pemphigoid, and the isolation that I felt. The first thing that Marc said was, "You're not alone anymore, and you have a community of people who care about you." That made all the difference.

Carolyn Fota lives in Stafford, VA, with her husband, Frank, and their three crazy cats. Carolyn enjoys walking, yoga, church and writing.

The IPPF Welcomes its Newest Peer Health Coach: Carolyn Fota

Carolyn Fota joins the IPPF Peer Health Coach team with a wealth of experience that includes almost 30 years in the United States Army serving as a medical service officer, retiring at the rank of lieutenant colonel. Her favorite experiences serving in the Army were being an airborne soldier dropping out of the sky, supporting various medical missions, flying in C-130s, transporting patients, and serving in the United States Army Surgeon General's Office.

Carolyn is currently employed by the Department of Defense working in software systems, communications, and training at Fort Belvoir, Virginia. Carolyn holds a bachelor's degree in government from Evangel University and a master's degree in human relations from the University of Oklahoma. Most of the time you'll find Carolyn either writing, reading, playing with one of her three crazy cats, or enjoying time with her husband of over twenty-nine years, Frank.



HAIR

Lisa Ann Krutzik

I rolled into South Detroit on a clear June afternoon and found my way to the private library at the VA medical center to speak with Dr. Thomas Chu. I first met Dr. Chu early in my pemphigus vulgaris (PV) treatment when he was involved with a clinical trial at the University at Buffalo. I was fascinated with him because we didn't just talk about PV, we talked about nutrition and Eastern medicine. We also discussed hair loss—an area of interest for Dr. Chu— and he reassured me that my own hair loss would be a temporary issue. Soon after, he was offered a dermatology residency at Wayne State University in Michigan and moved.

When I caught up with Dr. Chu in Detroit, we discussed the reasons people with autoimmune disease may have thinning hair and hair loss. According to Dr. Chu, hair loss starts two to three months before a follicle falls out. When the body experiences massive stress or illness, it focuses on healing the body and disrupts the anagen (growth) hair phase which kicks it into the telogen (dying) phase.

Human hair is fascinating: unlike animals who shed their undercoat at once, we constantly shed. Random follicles die at different times, so most people never see a pile of hair in the tub, but instead have some stuck to their comb. The growth period is three to five years on average and keeps the bulb of the hair full, thus tight within the skin surface. When we get sick, or we do something like a fad diet, our bodies focus on fixing whatever is wrong and doesn't feed the hair. So, any follicles that were already near the final phase will fall out quickly, followed by others. Once a follicle has reached the telogen phase, there isn't a way to reverse it. Because of the timing, patients may blame medications for hair thinning or loss instead of realizing that this process likely started before any treatment plan was put in place.

Can anything be done? Unfortunately, no. "Until the underlying problem is solved or managed … hair will not grow and will continue to fall out," Dr. Chu said. It's unlikely that lotions or shampoos will help with hair loss. I believe that it's important for patients to follow their doctors' advice, get their autoimmune diseases under control, eat highly nutritious food, stay away from harmful chemicals, and learn how to deal with stress in a constructive way. Then, their hair should eventually grow back.

Dr. Chu and I extended our conversation into his background and interests. "I come from a family of physicians and surgeons: my grandfather, father, uncle and cousin," Dr. Chu said. "I learned at a young age that medicine was not just a job or a career, but a way of life." His parents are from Taiwan and met in a painting class while studying medicine at the University of Texas. Dr. Chu's father is a well-known cardiac surgeon who performed the first transplant, coronary artery bypass, and artificial heart transplant surgeries in Taiwan. His mother is a pathologist who made headlines after identifying a gene that makes people susceptible to infection by the SARS virus. Dr. Chu's grandfather was a general surgeon as well. Dr. Chu shared, "He was a man of few words who told me that to truly live, one has to enjoy one's work while improving the lives of others."

We should always be changing—even if the changes are not what we planned—and learning how to view those changes positively in our daily lives

Dr. Chu also shares the interest in art that brought his parents together; however, he prefers poetry. He quoted T.S. Elliot before I got back on my bike to head west: *We must not cease from exploration and the end of all our exploring will be to arrive where we began and to know the place for the first time.*

Dr. Chu also introduced me to *The Hero's Journey*, a biography of the mythologist Joseph Campbell. I think the book could be useful for anyone dealing with a disease (caretakers included). *The Hero's Journey* taught me to be content with the reality that life is a journey. We should always be changing—even if the changes are not what we planned—and learning how to view those changes positively in our daily lives.

Personally, my hair was important to me—it was long and full. I grew up naturally blond, and during the summer it was almost a shiny yellow. Now, it has grown back dark brown, thick, and curly. I've been cutting it short and coloring it pink because I haven't yet accepted it. Maybe this winter I will allow it to grow and see who the new, natural me is. My heart has been on a journey through this disease—and so has my hair. Thanks to doctors like Dr. Chu, I haven't been on this journey alone.

Lisa Ann is a photographer, writer, entrepreneur, religious education director, and student of the world. PV shifted her perspective onto health and wellness. Find her blog at LARoxLife.wordpress.com.

Trust and Support

Kelly Calabrese

Being part of a community, especially one that knows how you feel and what you are going through, is very important. I didn't know what to think when I started experiencing blisters on my scalp in March of 2015. Was it an allergic reaction to something? Could it be cancer? I made an appointment with my physician, and he thought I had a bacterial infection. He gave me a topical antibiotic, but it did not work. Then I went to an ear, nose, and throat specialist because I was having trouble swallowing. He was unable to diagnose me, and I later learned that I had blisters in my throat and mouth as well as on my gums and lips.

Since the blisters were rapidly getting worse on my scalp and also on my collar bone, I decided to see a dermatologist. They performed a scalp biopsy. When the results came back, I was told to come in for another biopsy. The doctors did not want to believe that I had pemphigus vulgaris (PV). But in fact, I did have PV and was diagnosed within three months. The doctor put me on a daily dose of 40mg of prednisone. My blisters progressively worsened, so the prednisone dosage was increased to 90mg per day with no end in sight. I felt so alone and isolated without a support system. I kept asking my doctors if they knew anyone who had PV, but they were unable to tell me due to privacy reasons. I went online, and at the time I wasn't able to find the IPPF or any other help.

My PV progressed so much that I had to shut down my entire life by October 2015. The disease took over, and I was headed for death. I even updated my will. By December 2015, I ended up in the hospital. I had lost 35 pounds, and I had blistering on my entire face that was attacking my eye, ear, scalp, full chest, and back. They tested me for hepatitis B. I tested negative, so they began administering infusions of Rituxan® every two weeks. After five months and 11 rounds of Rituxan®, I stopped infusion treatment, though I

stayed on prednisone. My symptoms subsided, and in August 2016, I stopped all medications to give my body a rest.

In January 2017, I started experiencing blisters again. I did not want to go through the same pain. I visited my doctor, and he unfortunately wasn't able to continue treating me due to my insurance coverage changes. I lost my doctor who knew me very well. I had to start over and find new doctors to treat me. It took three months to get to the right sources. By May 2017, the blistering was so painful on my scalp that I started Rituxan® again. I was prescribed 1000mg infusions every two weeks, but the blistering became progressively worse. I kept asking to either increase the dosage or the duration, but my doctors would not change their protocol. I was in so much pain. By June 2017, I lost all my hair and my entire scalp was filled with bloody blisters.

That June, I visited my family, and my brother got on the computer and found the IPPF. I remember it like it was yesterday, crying as I spoke to Mei Ling Moore, an IPPF peer health coach. I cried tears of relief. I remember how supportive she was and how she sent me research on PV. Marc Yale then became my permanent peer health coach, and he suggested that I talk to my doctors about receiving the lymphoma protocol. However, the doctors were not interested in changing my current plan. I was between a rock and hard place and was getting worse by the minute.

In July 2017, I was able to see the doctor that first treated me for PV. However, I had to drive 60 miles to Denver to receive Rituxan®. My doctor prescribed weekly infusions of Rituxan®, and after 22 weekly infusions, I was clear of PV by December 2017.

It has been almost two years that I've been free of blisters and pain. I am very grateful to the IPPF community, Marc Yale, and the dedicated and loving IPPF staff. Thank you so very much.

Kelly J. Calabrese MS, CCN, is a Certified Clinical Nutritionist residing in Colorado Springs, CO. She is a PV patient and has been in remission since December 2017. She is a contributing writer for the Quarterly *and writes on health, wellness, fitness, and nutrition.*

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