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Message from the Executive Director

As I write this at the end of September, I am once again surprised at how fast time is passing. I’m sure you can relate, as it seems like everyone has been playing a game of catch-up for a long time now. Of course, this is to be expected as we adjust to whatever stage of the pandemic we are currently experiencing. After two-and-a-half years of constant change, many of us are trying to figure out what the “new normal” looks like—both now and in the years to come.

Here at the IPPF, that ever-changing “new normal” means that even though we’re hosting our own patient events virtually, there have been opportunities for staff and volunteers to return to in-person events focused on research, awareness, and advocacy. Over the past few months, we’ve attended the World Health Assembly, European Academy of Dermatology and Venereology Congress, Global Genes Patient Advocacy Summit, American Academy of Dermatology Association Legislative Conference and Hill Day, and Coalition of Skin Diseases Legislative Conference and Hill Day. At these events, the IPPF’s primary role is to make sure the patient voice is expressed and heard in ways that accurately reflect the needs of you—our community.

Looking forward, we’re excited for the final few months of 2022, as well as the beginning of 2023. As you may have seen, the IPPF Virtual Patient Education Conference is approaching on October 21-23. We have a packed agenda of disease experts, quality of life advocates, and partner organizations, all designed to bring you the most important P/P information. If you haven’t already, be sure to check out our agenda and register. If you can’t join us on October 21-23, you can still register now and access the recordings in the weeks after the conference.

In early 2023, the IPPF will host its first Externally-Led Patient-Focused Drug Development (EL-PFDD) meeting. This meeting is a chance for us to bring together patients, medical professionals, and international P/P advocacy groups to share their stories in front of the U.S. Food and Drug Administration about the burden of living with these diseases and the need for more research. More information about attending this virtual meeting will be available soon.

Finally, I want to take this opportunity to thank each and every one of you who has reached out during my first seven months as executive director. Though working remotely has many benefits, it makes personal interactions that much harder to come by. There are many days that it’s nearly impossible to keep up with work-related emails and tasks, but the personal notes from this community always stop me in my tracks. They remind me of how connected we all are and have always been. I’m happy to say that’s one aspect of the “old normal” that’s here to stay. For that, and for you, I am grateful.

Sincerely,

Patrick Dunn, IPPF Executive Director
patrick@pemphigus.org
After a two-year hiatus, the American Academy of Dermatology (AAD) returned live and in-person to hold their annual meeting in Boston, MA in March 2022. The meeting is attended by dermatologists all over the world and features sessions on many topics important to the dermatology community, including autoimmune blistering diseases.

For several years, IPPF Outreach Director Becky Strong has partnered with Dr. Donna Culton, MD, PhD, (University of North Carolina) to direct a session on pemphigus and pemphigoid (P/P). New this year, the session expanded to include several additional experts in autoimmune blistering diseases, including Annette Czernik, MD, FAAD, (DermMedical), Heather M. Holahan, MD, (University of North Carolina), and Brittney Schultz, MD, (University of Minnesota). The session provided updates for dermatologists treating these conditions in their pathogenesis and management as well as challenging cases, lessons learned, and practical clinical pearls.

The final portion of the session, led by Becky Strong, was unlike other sessions at the AAD meeting. It included a patient perspective on pemphigus vulgaris (PV), where Becky shared the journey surrounding her PV diagnosis, treatment, and everyday living tips. It was some of the most real-world education a dermatologist could receive on what it’s truly like to be diagnosed and live with an autoimmune blistering condition. Becky also provided an update on the IPPF and opportunities for physician engagement and patient advocacy.

As in years past, the 2022 session was highly rated and a wonderful opportunity that brought physicians and patients together to provide education for the international community on autoimmune blistering conditions. Excitedly, it was packed with standing room only. I was honored to be a part of it and hope to attend again next year!

Brittney Schultz, MD, is an Assistant Professor of Dermatology and Director of the Autoimmune Blistering Diseases Clinic at the University of Minnesota. She is also a Staff Dermatologist at the Minneapolis VA Medical Center.

Partnership with the IPPF and the American Academy of Dermatology
On May 25, I attended a side-event during the World Health Assembly in Geneva, Switzerland, titled, “Improving Health Outcomes for People Living with Dermatological Diseases Worldwide.” The event was co-hosted by the International Alliance of Dermatology Patient Organizations (known as GlobalSkin), the International League of Dermatological Societies (ILDS), and the International Alliance of Patients’ Organizations (IAPO).

The multi-stakeholder audience included patient leaders, dermatologists, industry, decision makers, and government representatives. The event was noteworthy as there were speakers from the World Health Organization (WHO) and Tanzanian Health Ministry, as well as a special preview of a new GlobalSkin documentary on the Burden of Atopic Eczema.

It was the first time that GlobalSkin hosted such an event, and its success is evident as it has already resulted in new connections with key representatives at WHO. GlobalSkin’s objective is to work to raise WHO’s awareness of dermatological conditions and the burden they impose on patients and caregivers. This could have a significant impact on all those affected by pemphigus and pemphigoid (P/P) around the globe.

At the event in Geneva, GlobalSkin publicly announced the formation and launch of the Global Dermatology Coalition. Becky Strong, IPPF Outreach Director, will represent the IPPF on the coalition. I had the honor of announcing the coalition, which is a patient-led multi-stakeholder group of 21 like-minded organizations. The formation of the coalition is a meaningful step and will help change perceptions about the impacts of dermatological diseases like P/P by elevating their prioritization in health policy at both global and national levels.

Marc Yale

Marc Yale was diagnosed in 2007 with cicatricial pemphigoid. In 2008 he joined the IPPF as a peer coach. He was the executive director from 2016-2020 and is now the research and advocacy coordinator. Marc currently resides in Ventura, CA, with his wife Beth and his daughter Hannah.
Join us virtually as we invite leading bullous disease experts to present on research and trends, educate about disease management, and answer tough questions regarding the care and treatment of pemphigus and pemphigoid.

The 2022 IPPF Patient Education Conference will be an exciting and educational event for any patient, caregiver, family member, practitioner, researcher, or student in the bullous disease field.

IPPF stakeholders are increasingly recognizing that the needs of the pemphigus and pemphigoid community are going unmet. The world needs a better understanding of these diseases and the treatments necessary to improve quality of life. The obstacles are substantial, but the opportunities are even greater.

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This past May, Staci White, IPPF Board of Directors Secretary, was asked by The Hill to participate in a virtual discussion on improving the rare disease diagnostic journey. We recently spoke with Staci about her experience and what she learned.

Can you tell us about your journey with pemphigus?

I was diagnosed with pemphigus vulgaris (PV) after being taken to the emergency room (ER) on Memorial Day weekend in 2011 with wounds and blisters over 85% of my body. My symptoms, though unrecognized as such by any physician who examined me prior to arriving at the ER, had begun in winter 2010. I was incorrectly diagnosed with a sexually transmitted disease and shingles.

I was hospitalized twice over a period of eight days, during which I received three biopsies. One of the biopsies was a deep tissue biopsy that left a scar. I was given fluids intravenously and treated with antibiotics, dilaudid, high doses of prednisone, and other medications. The extent of the damage to my skin was so severe that I received the same level of wound care as a burn victim. My treatment regimen included mycophenolate mofetil, prednisone, vitamins, and a laser focus on overall wellness. It also required weekly blood tests to monitor antibodies, which progressively stepped down to annual tests. My village of family and friends were an integral part of my treatment. They created a cocoon of compassion, love, care, and laughter in which I was able to build physical and mental strength.

I celebrated six years of remission on July 20, 2022. Along the way, I had a significant flare after my first-time tapering from prednisone. This required me to start my treatment over from the beginning. That moment cemented my understanding that PV was a chronic and
unpredictable condition. It also led me to incorporate counseling into my treatment program. During my journey to remission, my friend Cindy discovered the IPPF, which gave me a path to understand this disease and a community of those who understand life with PV. Despite the challenges of my journey, I remain grateful for PV. It forced me to be still. In that stillness my faith deepened, my creativity blossomed, and my core values were strengthened. All these were necessary for me to see the path to my purpose.

Can you tell us more about the interview and what it was for?

The Hill is the largest independent news and digital media site in the US. They hosted a live, virtual event titled “Improving Rare Disease Diagnostics & Care” on May 13, 2022. I was invited to be one of the four headliner interviews. The event brought together policymakers, medical experts, patient advocates, and key stakeholders for a conversation focused on improving the diagnostic journey of individuals living with rare disease. Moderated by The Hill’s contributing editor, Steve Scully, panelists included:

- Dr. Joni Rutter, PhD, Acting Director, National Center for Advancing Translational Sciences, National Institutes of Health
- Rep. Rodney Davis (R-IL), Member, Rare Disease Congressional Caucus
- Dr. Bradford Wilson, Co-Founder & Chief Scientific Officer, IndyGeneUS AI
- Ashley Valentine, Co-Founder & President, Sickle Cells
- Dr. Marshall Summar, Director, Rare Disease Institute, Children’s National Hospital
- Juliet K. Choi, President & CEO, Asian & Pacific Islander American Health Forum
- Rep. Diana DeGette (D-CO), Member, Energy & Commerce Committee; Member, Congressional Research and Development Caucus; Sponsor, Cures 2.0 Act.

The event brought together policymakers, medical experts, patient advocates, and key stakeholders for a conversation focused on improving the diagnostic journey of individuals living with rare disease.

- Molly Murray, President & CEO, Autoimmune Association
- Dr. Anne Pariser, Vice President, Medical and Regulatory Affairs, Alltrna; Former Director, Office of Rare Diseases Research, National Center for Advancing Translational Sciences

My interview section lasted for about 10 minutes. We discussed the twisted road to diagnosis, the specifics of PV, PV’s impact on my life then and now, and my message to the healthcare community. My core messages for physicians were to listen to patients and dig deeper because their diagnosis might be new to you. Steve asked me a question no one has ever asked me: “How do you ensure the disease doesn’t come back?” I replied that “I can’t, so I don’t focus on that. I choose to focus on living the lessons the disease has taught me. I focus on advocacy work which will hopefully lead to a cure one day. I try to keep my body in as healthy a state as a I can.”

What was your first reaction when you were approached about this opportunity?

My first reaction was to say, “Yes.” The IPPF has been a part of my journey since the beginning. Prior to becoming a board member, the IPPF earned my trust. When asked to do something advocacy-related, I usually say yes, and then figure it out.

How did you prepare for this great interview opportunity?

I was invited to do the interview a few weeks before the event. I spent that time learning more about The Hill, gathering information about important legislative priorities for our community with the help of Marc Yale, IPPF Advocacy and Research Coordinator, and Patrick Dunn.

CONTINUED
I wanted to represent the rare disease community with an empowered vulnerability that would make humanity the focus of the conversation.

IPPF Executive Director, and by creating talking points. I understood the responsibility of being one of the faces of a community of over 30 million people living with rare disease in the United States. I wanted to honor them.

What did you hope to accomplish, and do you think you were successful?

I wanted to represent the rare disease community with an empowered vulnerability that would make humanity the focus of the conversation. I hoped it would send a message to legislators and physicians about the importance of taking decisive action in support of patients’ multi-faceted needs.

I knew my role was not to control the interview, but it was my responsibility to communicate core messages that would be of service to our community. I decided to focus on three key points in whatever order worked with the rhythm of the interview. I planned to share my journey in a way that communicated the long, twisted, and often painful road to diagnosis. Second, I planned to connect my story to at least one legislative priority. Third, I planned to issue a call to action for physicians to focus on listening to patients with the same curiosity that led them to a career in medicine.

I shared the interview with a few physicians after it was over. I accomplished two of the three based on their feedback. I did not have the opportunity to speak to legislative priorities, but hopefully my continued advocacy work will lead to that road another time.

Do you have any tips and tricks for those who want to share their stories too?

My first step was sharing my story with myself. I began journaling early in my diagnosis and still do. I wrote down any and everything about my journey. The beauty of journaling is in its imperfection. It is your space. You can write whatever you want, however you want, without judgment. It can be liberating. It is in that space that your story will come together. I started verbally sharing my story before I wrote it, but you must do whatever works for you. When you are ready to share, think about your why. What is your purpose for sharing? It does not have to be a lofty purpose. It can just be a release for you. The first time I shared my story, it was one hundred percent for me. Sharing it gave me a place to make sense of an experience that I struggled to understand. After having shared my story a few times, advocacy is now my primary drive. However, each time I share, I feel more empowered, and I understand my experience a bit more.

How can other members of the IPPF community share their stories?

Tell your family and friends. Reach out to Becky Strong, IPPF Outreach Director, and ask about advocacy opportunities for patients. Submit an article to the *Quarterly*. I submitted my first article to the IPPF in 2018 on an impulse. Since then, I have shared some, or all, of my story outside of the IPPF, including at work. Often, those who connect with it the most are those who do not have a rare disease. Stories create connection. You might be surprised by how much your story can change someone’s life in large and small ways.

Staci White is the Operations Manager for Pediatric Physical Medicine and Rehabilitation and the Institute for Child Development at Hackensack Meridian Health. She is also a certified life coach who focuses on mindset and leadership coaching. Staci is a member of the Board of Directors for the IPPF where she serves as the Board Secretary. She was diagnosed with pemphigus vulgaris in 2011 and celebrated six years in remission on July 20, 2022.
Diagnosed with Bullous Pemphigoid? (BP)

Learn about a study of a potential new treatment for BP

With your participation, you can help researchers investigate a potential treatment for BP

How Do I Participate?

The first step is to talk with your doctor and share the study listing:
https://clinicaltrials.gov/ct2/show/NCT05267600

Speak to a Know Rare Patient Advocate who can help you connect to a study center to discuss the details of the study and if you are eligible to participate.

Go to balladstudybp.com

Who Can Take Part?

• 18 years old+
• Confirmed diagnosis of moderate to severe BP
• Active blisters
• There are additional study requirements you must meet to take part in this study. A study representative will discuss these with you.

What Will The Study Involve?

• Screening period: 2-3 weekly visits
• 43 weekly visits – most at the study center, some at home
• 1-2 minute subcutaneous injection (under the skin)

Are There Any Costs?

• No costs to participant
• Travel, accommodations, food and drink expenses reimbursed by study sponsor

never would have imagined 15 years ago after being diagnosed with pemphigoid that there would be as much interest and research in pemphigus and pemphigoid (P/P) as there are today. Even more impressive is the shift in attitudes regarding the importance of the patient perspective and its role in drug development. Clinical studies are now patient-centric, and these studies are being designed to include the things that patients feel are most important to them.

According to https://clinicaltrials.gov/ there are currently 16 clinical trials recruiting for pemphigus vulgaris (PV), six clinical trials recruiting for pemphigus foliaceus (PF), nine clinical trials recruiting for bullous pemphigoid (BP), and four clinical trials recruiting for mucous membrane pemphigoid (MMP). That’s 35 clinical trials—which doesn’t include the four trials that haven’t started recruiting yet for pemphigoid!

Many of the sponsors of these clinical trials have been working with the IPPF to understand the impact on people living with our diseases. The IPPF is actively assisting with setting up patient advisory boards, patient interviews, surveys, and reviews of patient-facing materials for clinical studies. The IPPF has also co-authored the abstract, Quality of Life in Patients with Bullous Pemphigoid, with the hope of presenting a poster at the 2022 Rare Diseases + Orphan Products Breakthrough Summit, taking place October 17-18 in Washington, DC. Patient data for the project was extracted and analyzed for the IPPF Natural History Study to help illustrate the burden of disease BP patients face every day with their disease.

Our research efforts haven’t stopped there, and working with academia is critical in helping us learn about different aspects of P/P. In recent months, with the help of our community and researchers at the University of Southern California, we have conducted multiple surveys and published the results on the effects of COVID-19 and P/P patients. This has sparked interest by other academic researchers to explore how P/P can negatively affect a patient’s quality of life.

Perhaps the most ambitious research initiative that the IPPF is working on is the upcoming Externally-Led Patient-Focused Drug Development (EL-PFDD) Meeting. The IPPF is collaborating with our international partners The Association Pemphigus/Pemphigoid France (APPF), PemFriends (UK), and the Friends of Pemphigus/Pemphigoid Japan to host the meeting. The U.S. Food and Drug Administration (FDA) established the Patient-Focused Drug Development initiative to collect patient insights on specific diseases and their impact on daily life, as well as treatment options. These meetings are designed to connect patients and families directly with the FDA as well as drug researchers and developers, thereby elevating the patient voice in disease-related research, treatment development, and evaluation. Meetings are either organized and held by the FDA (referred to as FDA-led) or patient advocacy organizations (referred to as externally-led) to gather public input on gaps in disease management for specific diseases. Please watch for a survey prior to the meeting as we need your feedback on how P/P has affected you.

This is a very exciting time for the IPPF community, as research is moving forward at a fast pace. Your engagement and support in all of these projects will help the IPPF continue improving the lives of all those affected by pemphigus and pemphigoid.

Marc Yale was diagnosed in 2007 with cicatricial pemphigoid. In 2008 he joined the IPPF as a peer coach. He was the executive director from 2016-2020 and is now the research and advocacy coordinator. Marc currently resides in Ventura, CA, with his wife Beth and his daughter Hannah.
My Ongoing Battle with Pemphigus Vulgaris

Corrie Sirota

Corrie’s story originally appeared on her personal blog (https://www.corriesirota.com/one-in-a-million/) on August 26, 2022.
On February 14, 2022, my husband Andy tested positive for COVID-19. I was upset because I had been planning to travel to Florida and visit my friend in two weeks’ time and now, selfishly, I had to worry whether I would be able to go. I took a rapid test and (thankfully) it was negative. Little did I know that getting COVID-19 would have been easy compared to what subsequently occurred.

It started as a rash. First, I noticed that I was itchy on my left forearm. That’s strange, I thought. What’s up with that? The itch spread to the other arm and became rather uncomfortable. I started to pop Benadryl pills and still thought nothing of it. When I left for Florida two weeks later, the rash had moved from my arms to my thighs and became bumpy, uncomfortable, and incredibly itchy but manageable. I resolved that I would simply enjoy my time in Florida despite having this bizarre, unexplained rash. Since it wasn’t going away, I decided to do schedule an appointment with my general practitioner.

When I returned to Montréal, I met with my doctor on March 9, 2022. She didn’t seem too concerned; however, she was unable to provide a diagnosis. She wrote a prescription to help manage the itching and referred me to a dermatologist to further investigate.

In the meantime, the rash continued to move about my body, never relenting. Then, one day, it just stopped. However, with my body no longer itchy, I realized I had neglected to pay attention to the fact that my mouth was bothering me. I noticed that it felt as though I had cut the inside of my cheek. My gums felt extremely sensitive. Yet, I went about my day, life, and schedule as if nothing were different.

On April 30, I attended a weekend training for a travel company I work for. I started noticing that it was becoming increasingly more uncomfortable to eat. Upon getting into bed on the first evening, I remember thinking that perhaps I had a tooth infection and that I might need an implant. I had just spent the past year dealing with a tooth extraction and implant, so this seemed like a logical conclusion. My ability to consume any food became increasingly difficult. When I returned home, I contacted my dentist for an emergency appointment.

On May 3, I went to my dentist and when I sat in the chair, he took one look at my mouth and said, “Hold on, I’ll be right back.” When he returned, he told me he thought that I had lichen planus, a condition that can cause swelling and irritation in the skin, hair, nails, and mucous membranes. He suggested that I apply an oral cortisone gel and if the condition did not improve in 10 days, to see an oral surgeon who could do a biopsy. A biopsy? This was starting to feel serious! The biopsy would determine the type of virus and the treatment I would require. While I wasn’t happy to hear this, I was not terribly concerned either.

Being a good patient, I did as my dentist suggested. However, 10 days later I was not feeling better. In fact, it occurred to me that I was developing unexplained blisters on the front and back of my torso. These blisters also appeared under my arm and there was one on my forehead. As the days progressed other blisters appeared on my back. Then, as if that wasn’t enough, I noticed some bumps on my head—one, two, three, and then more. By the time I spoke to the dentist again, I had managed to make an appointment with a dermatologist.

The dermatologist confirmed that something was amiss and referred me to an oral surgeon who contacted me one week later. As soon as the oral surgeon looked into my mouth he said, “It appears to be Pemphigus Vulgaris (PV).” Pemphi-what? He advised me that he would do a biopsy to confirm this diagnosis, and that he was going to refer me to the Montréal General Hospital for treatment with the dermatology clinic immediately. I was scheduled to work in Europe at the end of June and he told me that wouldn’t be possible. That’s when things started to feel real for me.

I was in complete and total shock. Me? I’m in great shape. I eat well and I take care of myself. How? Why? What does this mean? A part of me knew that my life and lifestyle were about to change, but at that time I didn’t know exactly how.

On June 6, I met with the most professional and compassionate dermatologist, Dr. Therese El-Helou
(Montréal General Hospital), who walked me through the process of the potential diagnosis and treatment options for PV.

And so the process began with blood tests (12 vials), a gastroscopy, and seven vaccines administered due to a susceptibility to infection. I had to protect myself in every way I could. My new normal was a mixture of medications, vitamins, prednisone tablets, calcium, vitamin D, five different topical creams, and two rinses to help mitigate the lesions on my body that had spread to my head, trunk, mouth, eyes, throat, and other areas that shall remain nameless. This treatment was done to help keep my condition stable while I waited for approval for Rituxan®, a well-researched infusion treatment that targets certain parts of the immune system to help fight PV. In order to receive Rituxan®, I needed to be up to date for my tests and vaccines and I needed insurance approval due to the high cost of the medication.

On July 20, I received the approval from my insurance and I was able to set up the procedure. Due to a series of miscommunications, I still had to wait three weeks to schedule my first six-hour infusion. The first infusion happened on August 17, and thankfully, I had virtually no side effects. My second infusion took place at the end of August. With the start of Rituxan®, I hope to be able to wean off prednisone. I am determined to remain optimistic that this is what will put me into remission.

What have I learned from my experience? Like the philosophy of Gerald Jampolsky, MD, author of Forgiveness: The Greatest Healer of All, I believe that attitude is everything—it determines how you experience every aspect of your life. I have never been one to feel sorry for myself, but this felt different. I had to put my whole life on hold. I couldn’t travel, eat many of my favorite foods (I wasn’t able to eat anything solid for quite some time and still currently cannot eat many foods), and take care of my own body (my husband had to tend to my open blisters). I could not be in the sun and I could not drink alcohol. But most notably, I hadn’t ever felt so scared.

This condition has a life of its own and each day I’m unsure how I will look or feel. I’ve had to learn to live in the moment and appreciate what I’m still able to do. I’ve reached out to a former student who has been living with PV for the past seven years and they have turned out to be an incredible source of information, support, and guidance. I joined the PV Facebook group and met incredible, supportive warriors who offer kindness, care, advice, and compassion. I’ve also received support and guidance from the IPPF.

I’ve received overwhelming support from my friends and family from offers of soup delivery, rides to appointments, gifts, and consistent check-ins. I’ve been able to attend family events and continue laughing throughout life despite what I’m going through. I appreciate the incredible support from health care providers who’ve taken the time to listen, explain, and advocate for me.

Ironically, I have never felt closer to my clients than these past few months. As a grief counselor I am acutely aware of how fragile life is, how one minute your life is one way and then in the blink of an eye so much can change. I feel like I’ve been able to empathize with my clients more than I was able to before my PV diagnosis. I’ve learned how important it is to reach out and lean on the people around you during a challenge like this. Sometimes there isn’t a quick fix for grief, or PV, where there isn’t a known cause or cure. And yet there are many ways to help and support people by accompanying them on their journey.

I remain eternally grateful to the plethora of people in my life. Without them, I don’t know where I would be. It is true that the best thing in life is to hold onto each other. One of my colleagues told me, “That disease picked the wrong person to mess with!” and I constantly remind myself of this. I still have no idea what the future has in store for me, but then again, does anyone ever?

Corrie Sirota, MSW, PSW, is a Clinical Social Worker, Psychotherapist, and Grief, Loss, and Bereavement Specialist. She is the author of “Someone Died...Now What? A Personal and Professional Perspective on Coping with Grief and Loss” and the co-host of the radio show Life Unrehearsed. She lives in Canada and is married and has two daughters and one son-in-law.
KSL Beutner Laboratories Launches First Blood Test for Mucous Membrane Pemphigoid

Highly accurate IIF serological assay for Laminin 332 Mucous Membrane Pemphigoid reduces the wait for confirmed diagnosis from months or years to 72 hours or less

The following was published on July 27, 2022

KSL Beutner Laboratories (Beutner), a global leader in immunologic testing for the diagnosis of bullous, vascular, connective tissue and inherited skin diseases, has launched a first-to-market indirect immunofluorescence (IIF) serum blood test in the U.S. that positively identifies laminin 332, an antigen associated with the chronic, debilitating autoimmune disease mucous membrane pemphigoid (MMP).

Without a definitive interpretation, patients experience considerable pain and suffering due to misdiagnosis and treatment delays. Long sought after by oral pathologists, oral surgeons, periodontists and dentists, in addition to dermatologists, Beutner’s laminin 332-specific serological assay reduces the time to confirmed diagnosis from as much as two years to 72 hours or less.

“The development and launch of Beutner’s much-anticipated serological assay for laminin 332 MMP builds on the pioneering work of Dr. Ernst Beutner, the father of immunodermatology,” said Dr. Lakshmanan Suresh, Technical Director at Beutner and Chief Medical Officer of KSL Diagnostics, Inc. (www.ksldx.com), which counts Beutner among its clinical laboratories. “The innovative methods he established for diagnosing autoimmune blistering diseases are used worldwide, and we are proud to continue his legacy.”

MMP is an autoimmune blistering disease characterized by multisite lesions on mucous membranes. Anti-laminin 332 MMP lesions often scar and can lead to serious complications depending on the mucosal surfaces affected. Oral mucosa -- gums, inner lining of the cheeks and lips, palate and tongue -- are involved in 80-90% of cases. Scarring of ocular mucosa, present in half of patients, may lead to blindness. The disease can also impact mucous membranes in the nose, throat, genitals and anus, causing severe, irreversible damage. Another complication in 25-30% of patients is increased risk of cancer malignancies, primarily adenocarcinoma in the gastrointestinal and genital mucosa and lungs. The condition can be fatal if left untreated. Considered a rare disease of unknown cause, MMP occurs mainly in people between ages 60 and 80 and, infrequently, in children. Women are affected twice as often as men.

“The first assay of its kind available in the U.S., Beutner’s IIF serum blood test verifying the presence of specific autoantibodies for laminin 332 MMP now enables clinicians to accurately identify this hard-to-diagnose disease much faster,” said Dr. Raminder Grover, Laboratory Director at Beutner. “This in turn allows for available therapies to be started much earlier to alleviate patients’ significant ongoing discomfort and spare them the long-term medical complications of this devastating disease.”

The most common autoantigens associated with MMP are BP180 and laminin 332. About one-third of patients are afflicted with the laminin 332 variant. Since 2002, blood tests to detect BP180 have been widely available, but a laminin 332 serum test has not been approved in the U.S. until now. Anti-laminin 332 MMP cannot be differentiated from other forms of the disease based on clinical examination. It can only be distinguished by a serological test for IgG antibodies of the variant. Because it mimics other diseases in the mouth, patients may suffer increasing pain and decreased quality of life for six months to two years before obtaining conclusive test results. Reaching a positive diagnosis as soon as possible is critical so that physicians can begin treatment.

Confirming laminin 332 MMP relies on several laboratory tests offered by Beutner: first, a direct immunofluorescence (DIF) microscopy of a skin biopsy to detect tissue-bound immunoreactants. Although DIF is the gold standard for investigating all forms of MMP, it does not always differentiate between variants. Next, indirect immunofluorescence is applied to identify circulating antibodies targeting the autoantigens in the basement membrane zone (BMZ) of the skin. This type of analysis is done in two parts -- an IIF test on salt-split skin and an IIF serum test on transfected cells for laminin 332 IgG/IgG4 antibodies using a EUROIMMUN assay. Beutner’s IIF serological assay has a sensitivity of 84% and a specificity of 99%.

Beutner recently received approval from the New York State Department of Health to perform the laminin 332 IIF blood test for U.S. customers at its Buffalo, NY lab.

Dr. Ernst H. Beutner, a co-founder of Beutner Laboratories, pioneered the DIF test 40 years ago after he and his associates at the University at Buffalo (UB) discovered the role of autoimmunity in pemphigus and pemphigoid. Development of defined, quantified immunofluorescent methods now used worldwide for the investigation of autoimmune skin diseases began with studies at UB led by senior researchers from Beutner, UB and Harvard University.

KSL Beutner Laboratories provides anti-laminin 332 IIF serological testing as a service, as well as several other assays for laminin 332 MMP and for the BP180 variant of MMP. Tests can be ordered at https://www.beutnerlabs.com/.

About KSL Beutner Laboratories

KSL Beutner Laboratories, a global innovator in immunodermatology, specializes in immunologic testing for the diagnosis of bullous, vascular, connective tissue and inherited skin diseases. Beutner’s staff maintains a long tradition of excellence in laboratory service and brings a wealth of experience to diagnostic immunologic research of the skin and mucous membranes.

Beutner Laboratories was founded by industry pioneer Dr. Ernst H. Beutner and his wife Gloria Beutner in 1992 after Dr. Beutner and his associates at the University at Buffalo (UB) discovered the role of autoimmunity in pemphigus and pemphigoid. The Beutner team developed reproducible diagnostic methods such as immunofluorescence (IF) for detecting the pathogenic autoantibodies that cause these diseases. The lab’s immunology and pathology experts have authored over 300 publications in immunodermatology and skin immunopathology. For more information, visit https://www.beutnerlabs.com/.
Share your experience with BP and help define the patient experience for use in new treatment development

Background

- In this study, the focus is on patient experiences with BP and its treatment.
- Participation will involve one interview session (one hour) conducted via telephone.

Purpose

- This research will focus on the symptoms you as the patient experience - how they impact your daily life and how they make you feel - and what it is like to be a patient with BP.

Getting Involved

- Your participation in the study is completely voluntary.
- All information that can identify you personally will be kept strictly confidential. There will be no way to identify you from the reports that will be generated from this study.
- You do not need to do anything to prepare other than be ready to talk about your experience living with BP and your experience with treatment.
- During your consent process, you can invite a caregiver to join you in your interview. They will also be given the chance to consent to the interview.
- You (and/or your caregiver) can withdraw from the study at any time.

Next Steps

- If you are interested in this study, you will need to provide consent to participate.
- If you are selected to participate, you will be contacted by a researcher to schedule your interview.
- If you have questions about any part of the study please contact contact@global-patients.com
With the rising prevalence of complementary and alternative medicine (CAM), patients have been increasingly turning toward the use of alternate modalities to supplement their conventional treatment plans. A review of survey data looking at the trends in the use of CAM found that 86% of U.S. patients seek alternative medicine to treat their skin conditions. The National Center for Health Statistics (NCHS) reports that the most used CAM by U.S. adults were non-mineral, non-vitamin, natural products. Similarly, the most common alternative therapy used by dermatology patients are herbal and dietary supplements. Despite being used as a natural remedy for immune boosting or anti-inflammatory effects to either supplement conventional treatment or as stand-alone therapy, there is little-to-no compelling evidence of treatment efficacy.

While there is limited information on the benefits and effectiveness of herbal supplements for any disease, there have been increasing reports of their various negative medical and dermatological effects. Multiple studies have demonstrated the autoimmune disease inducing properties of various herbal supplements, including echinacea, spirulina platensis, and chlorella. In addition, there are documented cases of autoimmune skin diseases, including pemphigus, that were either started or worsened by taking these herbal supplements.

Echinacea has been suggested to induce a pemphigus flare in a man with a previously stable disease. The patient had an upper respiratory infection (URI) and decided to take echinacea supplements...
for the first time. Within one week, the patient developed blisters on his head, trunk, and in his mouth. Another man with pemphigus vulgaris (PV) experienced a severe flare of his disease after starting a food supplement containing ginseng, ginkgo biloba, and spirulina platensis. Following this report was a case of a previously healthy patient who developed mixed features of pemphigus and pemphigoid after using a spirulina-based product for a year. Given these reports, the use of “immune boosting” herbal supplements especially echinacea and spirulina by patients with autoimmune bullous diseases, raises concerns of the effects of the immune-enhancing properties on disease activity.

Previous studies have shown that spirulina’s immunostimulatory effects appear to be mediated by both our early line of defense, the innate immune system, and our more sophisticated adaptive immunity. Numerous studies have shown that spirulina stimulates the immune system by activating several inflammatory cells, including our white blood cells in particular natural killer (NK) cells, T cells, and monocytes. Isolated components of spirulina increase numerous inflammatory markers, which are proteins made by certain cells and cause inflammation. Activation of these cells in autoimmune bullous diseases is certainly problematic.

We found that water-soluble spirulina significantly stimulates inflammatory protein production, primarily driven by the activation of various types of monocytes. Monocytes play an important role in immune defense, inflammation, and homeostasis by sensing their local environment, clearing pathogens and dead cells, and initiating long term immunity. We sought to evaluate whether spirulina’s immunostimulatory effects differ in healthy controls (HCs) compared to dermatomyositis (DM), another autoimmune skin disease. We performed experiments on spirulina stimulated HC and DM human immune cells. These experiments demonstrated similar effects in both HCs and DM, with spirulina significantly increasing inflammatory cytokine levels. Our studies utilizing healthy and DM cells suggest that spirulina may promote autoimmunity via production of inflammatory proteins in monocytes, thereby implicating spirulina in the development of DM in some susceptible patients.

Such immune stimulation would not be desirable for those with autoimmune bullous diseases and could trigger a new disease or worsen a current diagnosis.

Understanding how spirulina and other immune stimulating herbs function is critical to better understand how they induce inflammatory proteins in DM and other autoimmune patients, and to ultimately inform physicians and patients to make health-conscious decisions and guide the clinical management of patients with autoimmune skin diseases. It is important for patients with autoimmune diseases to discuss any planned use of herbal medications with their health care provider prior to starting them.

DeAnna Diaz, MS, MBS, is a fourth-year medical student at PCOM and pre-doctoral research fellow at the Department of Dermatology at the University of Pennsylvania.

Josef Concha, MD, is a postdoctoral fellow in autoimmune skin disease in the Department of Dermatology at the University of Pennsylvania.

Dr. Victoria Werth is a Professor of Dermatology and Medicine at the Hospital of the University of Pennsylvania and Chief of Dermatology at the Veteran’s Administration Medical Center. Her clinical and research interests lie in autoimmune skin diseases, including autoimmune blistering diseases.
Thank You

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FOR THEIR CONTINUED SUPPORT OF THE
IPPF AWARENESS PROGRAM
Our Spotlight section features a medical professional whose work regularly impacts the lives of pemphigus and pemphigoid (P/P) patients. Get to know a new physician, researcher, or other medical professional who knows these diseases best. This issue, we’re featuring Dr. Cory Simpson.

Dr. Simpson is a board-certified dermatologist and Assistant Professor in the Division of Dermatology at the University of Washington. In 2012, he graduated from the Medical Scientist Training Program at Northwestern University in Chicago, earning both his PhD and MD. His thesis research focused on the cellular functions of desmoglein 1, which is targeted in pemphigus, and led him to pursue clinical training in dermatology. After an internship in internal medicine through the University of Chicago, Dr. Simpson moved to the University of Pennsylvania for dermatology residency and served as Chief Resident in his final year at Penn. Upon finishing his clinical training in 2016, he obtained grants from the National Institutes of Health and the Dermatology Foundation allowing him to return to research, applying genetic tools and imaging techniques to a model of human skin grown in the lab.

In 2021, he was recruited to the University of Washington, where he is building a lab focused on understanding how the skin is compromised in dermatologic disorders and identifying potential new treatments to promote tissue regeneration for those with rare skin diseases. Dr. Simpson’s weekly clinic in Seattle focuses on both autoimmune and genetic skin diseases, including pemphigus, pemphigoid, Darier, and Hailey-Hailey disease.

How did you become interested in P/P?

During medical school, I completed a PhD in cell biology working under Dr. Kathy Green (Northwestern Medicine), whose lab has made critical discoveries about the biology underlying pemphigus. My project in her lab focused on the function of desmoglein 1, which is one of the proteins targeted by auto-antibodies in patients with pemphigus. This research inspired me to pursue specialized training in dermatology so I could combine my research expertise with the practice of dermatology. When I saw my first patient with pemphigus as a senior medical student, it was a transformative experience to realize that the protein I studied in the lab was actually at the root of the person’s illness. I also learned first-hand how challenging it can be for patients to have a blistering disease, which can be difficult to treat. That inspired me to become involved with the IPPF and its efforts to advocate for patients with these rare disorders, helping connect them with reliable information, committed clinicians, and a supportive community.

What is one thing you’d want all patients to know early on in their journey with P/P?

Patients should know that the first clinic visit to discuss the diagnosis of pemphigus or pemphigoid can be overwhelming, but that initial appointment is an important step toward finding a therapy that will work for you. Allow yourself time after the appointment to process the information and consider making a second appointment so you can get all your questions answered and confidently decide on a treatment plan with your doctor. Though it can take months to get your disease controlled and you may not be able to tolerate every therapy, be assured that eventually you will find a treatment to help you heal.

What can patients do to better advocate for themselves?

One of the hardest steps with a rare disease is getting a firm diagnosis. Unfortunately, this can take years for some patients with pemphigus or pemphigoid, and they may feel hopeless during that time. Advocate for a biopsy if your skin or mucosal disease is not responding to initial therapies. For patients with a known diagnosis, if questions come up between appointments, be sure to write them down as they are not easily recalled during appointments. Finally, if you feel your treatment plan is not working, or you notice the side effects (which all treatments have) are not tolerable, don’t be afraid to express your concerns to your doctor.

What is one fun fact about yourself?

I am highly dependent on my daily coffee, and Dunkin’ Donuts cold brew is my favorite. Moving recently to Seattle, I realized there is no Dunkin’ here in Starbucks territory, so now I order it online and cold brew it myself in the fridge.
Spotlight

IPPF Board of Director’s Spotlight

Our new Board of Director’s Spotlight section features a board member who serves on the governing body of the IPPF. Board members strategize and provide operational oversight with the ultimate goal of sustaining the IPPF. Get to know our valuable board members and learn more about their story. This issue, we’re featuring David Baron.

David Baron was diagnosed with pemphigus vulgaris (PV) in 2005. He suffered from a severe and refractory case. He reached remission after three years of treatment with rituximab and IVIg. He is an airline pilot with a major airline based in San Francisco, where he flies the Airbus A320 series aircraft in domestic and international operations. He is a graduate of Purdue University with a bachelor’s degree in aviation technology.

When did you first connect with the IPPF and pemphigus and pemphigoid (P/P) community?

I first connected with the IPPF and P/P community in 2005. I was newly diagnosed with pemphigus vulgaris (PV), and I felt lost. The internet in 2005 wasn’t what it is today, but I turned to the internet to find more information about PV. That’s where I found the IPPF, and at that time the IPPF had an email list server which helped me connect with other patients. I found answers to so many of the questions I had thanks to the IPPF and other patients.

How has the IPPF benefitted you?

The IPPF has benefitted me in so many ways and continues to do so today. When I was first diagnosed and looking for answers, I found them through the IPPF. I also found connections with other patients, first through email, and later at my first IPPF patient conference in 2010. When I was in rough shape and not responding to the standard treatments at the time, it was the IPPF’s founder Janet Segall who directed me to an expert.

What motivated you to be more involved with the IPPF?

As a patient, I received so much help from the IPPF along my journey to remission. I felt it was my responsibility to give back to the IPPF and P/P patients. I could use both my experience with PV and my professional and volunteer background to give back. Volunteering is also cathartic. My journey with PV left scars that aren’t visible. Volunteering, helping P/P patients, and giving back to the IPPF helps heal those scars.

How can others take their first step toward becoming more involved?

There are several ways to get involved. We always need volunteers. Everyone has a talent and/or drive that the IPPF needs. Volunteers are part of the lifeblood of the IPPF. Reach out to any of the staff or board members, and we can get you pointed in the right direction.

Not everyone has the free time available to volunteer. Another way to get involved is through donating. Donations are the other lifeblood of the IPPF. Donations allow the IPPF to help patients. I strongly encourage everyone reading this to become a Healing Hero. If you are already a Healing Hero, thank you!

Is there a specific IPPF initiative you are especially passionate about (i.e., patient support, awareness, research, advocacy)?

I’m very passionate about patient support. As a patient myself, I understand the feelings of a newly diagnosed P/P patient. Patient support gives us the ability to help patients find information, medical professionals familiar with P/P, or even just a friendly ear.

What is one fun fact about yourself?

I was able to fly a plane before I could drive a car. I was flying gliders before I had my automobile driver’s license.
OUR COMMUNITY IS STRONG

Let's help it grow even stronger.

Join us for GivingTuesday on November 29. Donate to the IPPF to ensure our programs are available to all those who need them today, tomorrow, and for years to come.

#GivingTuesday
pemphigus.org/donate