

# Real-Life Tips/Tricks for Living with Pemphigus and Other Blistering Diseases: An Interview with Becky Strong

*By Dr. Brittney Schultz*



Becky Strong is a patient with pemphigus vulgaris. For Becky, her journey included 17 months, 7 physician specialists, multiple encounters with her dentist, and several rounds of incorrect treatments and procedures before correctly being diagnosed with pemphigus vulgaris. She's a registered nurse living in Michigan with her husband Tim and 2 children. Currently, Becky is the Outreach Director for the International Pemphigus & Pemphigoid Foundation, an organization that supports patients with these two rare autoimmune diseases. Becky is responsible for medical and dental professional, dental student, and patient education at the IPPF. She also spends time advocating at the federal level for patients with rare diseases.

I first met Becky at an American Academy of Dermatology meeting where she shared her experience as a patient with pemphigus. She is the unique blend of patient and advocate and she is one of the reasons I chose to focus on autoimmune blistering diseases. I hope her experiences will help you with your patients!

## **Tell us about your diagnosis of pemphigus and what you went through prior to diagnosis.**

- My lesions started in my mouth in September 2008. I had these water blisters that would appear, and they wouldn't hurt but they would break easily when I ran my tongue across them. They would go away and a few days later, reappear in a different place in my mouth. The nurse in me attributed the lesions to the spicy salsa I was eating, and I didn't realize this was a problem until they started turning into what I thought were large canker sores.
- About a month after this started, I had my 6-month check-up with my dentist. I told her that my mouth just felt slimy all of the time and these canker sores were very painful and the scabs came off the lesions very easily. At this time, she told me that I had bad oral hygiene and I needed to brush more. She also recommended an over

the counter debriding mouth rinse and told me to use it to make things better. It was extremely painful, but I did exactly as I was instructed but things were getting worse.

- I also was diagnosed with anal fissures in October 2008, and my gastroenterologist was struggling to find treatments to work as the traditional treatments did nothing to provide relief. A year later in October 2009, I was still struggling with the anal fissures - and now I had 3 of them. It was at one of the follow-up appointments that I mentioned the oral lesions to my GI doctor. He looked in my mouth and cringed - and if you can gross out a gastroenterologist, you really feel like somebody! Anyway, he called in his physician assistant who was a dental hygienist in a former career. She took a look at me and they went back into the private office and called her former employer, a dentist. When my GI doc came back, he had a prescription for "Magic Mouthwash" - part dexamethasone for the inflammation, part lidocaine for the pain and part Maalox to help it all stick. I was to swish 3 times a day for 10 days. When I used it, my mouth got remarkably better, but when I stopped it came back with a vengeance.
- In November, I made an appointment with my primary care internist. I explained what I had been going through and told him that I think if I could use the Magic Mouthwash for 14 days, my problems would go away. He took a look in my mouth and told me that the amount of steroid was too much to be on that long, and besides, I had cold sores that would go away on their own in 14 days with a prescription or if we did nothing they would go away in 2 weeks. This was after I had told him that this had been going on for months without relief.
- Back at my dentist in December, I asked her to refill the Magic Mouthwash and she too balked that it was too much steroid. Instead she gave me an alcohol based mouth rinse and told me "I don't know if it will help, but it won't hurt and you need to brush more!" By this point, I couldn't even turn on an electric toothbrush and eating a simply cup of chicken noodle soup took over an hour and felt like I was chewing glass.
- In December, my GI doc scheduled me for a colonoscopy in case I was experiencing a chronic or acute colitis. He also wanted to try an off-label use of Botox in order to help my anal fissures heal. During the colonoscopy, he did a procedure that injected Botox on either side of the largest fissure allowing it to heal. This resulted in a complete loss of bowel control - I'm an OR nurse and I had to wear an adult diaper to work, let my boss and co-workers in on what happened, and when I called for relief, it needed to come quickly.
- By January, I was getting to the end of my rope. I asked my dentist who I needed to see, and she told me that no specialist would do anything else other than what she's already doing. My husband and I had a very difficult conversation - I told my husband of barely a year that if he ever wanted to leave me, he could - no questions asked. I had no idea what was wrong with me or how much worse it would get, but I married a good guy who told me he married me for better or worse and this just happened to be "worst". Years later, he admitted to me that he thought I was literally dying, and he loved me so much that there was no way he would let me die alone. He also made an appointment with his internist at the University of Michigan. It was the first time in my whole journey that I felt somebody was listening and not supposing what was going on with me. I told her everything that I thought could possibly be contributing to my current situation - childhood immunizations, allergy testing, professional hazards, and my overseas medical missions. She told me that she loved a good mystery and ran a bunch of bloodwork that all came back pretty normal. She wanted me to see the rheumatologist but didn't understand how the oral lesions fit in, so she sent me to the oral surgery clinic at the University.
- At oral surgery, they told me that I needed to have a biopsy. They took 4 biopsies from different places in my mouth - 2 for H&E, and 2 for direct immunofluorescence. On February 10, 2010 - 17 months after my journey began - I finally received the diagnosis of pemphigus vulgaris.

## Is there any advice you would give dermatologists for patients who see patients with pemphigus?

- If you don't know something, it's okay to tell us patients that you don't know. But we will want you to help us to find the answer that we are looking for.
- Being diagnosed with any disease, let alone rare disease, is a very hard thing to comprehend. We patients are quickly overwhelmed with information and the medical jargon around our diseases are hard to understand and want to get better right away. Having handouts for patients is very helpful, and pathways to reiterate the good teaching you're doing.
- Also, please be patient with us. It may be necessary to book a double appointment until we get comfortable with our disease as we're going to have a lot of questions.

## Any tips or tricks in managing pemphigus that you have learned along the way that might benefit other patients?

Sure! These have worked for me:

- Great skin care is essential. Start with the basics and explain the difference between fragrance-free and unscented, ointments and creams are more moisturizing than lotions.
- Sunscreen is important, but more important is that you use it properly.
- SPF clothing and hats, as well as sunglasses, are as useful as sunscreen.
- Water comes out of showers at a very high rate of speed, so baths are better to help keep your skin intact. Water shouldn't be hot or cold as both can damage the skin.
- Fragrance-free, dye-free, paraben-free, alcohol-free body washes are less drying than bar soaps.
- Nasal saline drops help to keep nasal lesions moist and helps to prevent them from bleeding.
- Saline drops for eyes can prevent dryness and help to wash away any irritants that get into your eyes.
- Use a toddler toothbrush as they are generally smaller, rounded and very soft when you have oral lesions. Toothbrushes should be manual as the ultrasonic action of electric toothbrushes can disrupt the tissues.
- Using toddler toothpaste is also great as mint flavoring burns, and they tend to be gels which are less abrasive - just be sure to use fluoride to protect the teeth.
- Flossing tape can be less painful to use than dental floss.
- Frequent dental appointments help to decrease the inflammation from plaque and tartar.
- Consider seeing a registered dietician to help learn about the pH of foods and nutritious ways to prepare foods that won't hurt the mouth or throat.
- Use cornstarch/talc powder to prevent skin from sticking to clothing - and wear breathable fabric that wicks moisture from skin.
- Use a watering can with a long pour spout to pour water over yourself when you urinate to stop the burning of below-the-belt lesions.
- Use tight t-shirts or leggings to help hold dressings in place as an alternative to tape.
- As hard as it is, DON'T SCRATCH! Consider over the counter and prescription medications and topical treatments. Wearing cotton gloves at night as well as clothing to cover lesions to prevent causing micro abrasions or tearing lesions.

## Any well-meaning tips or tricks you were given that you would NOT recommend?

There are a LOT of internet groups for our diseases who share information that may not be medically sound. Even if the advice is good, it may not be right for you. Before trying anything that you see on the internet or hear from another person, please be sure to have a meaningful discussion with your doctor to be sure it is right for you.

## What resources does the IPPF have for patients?

The IPPF offers lots of free things for patients, caregivers, families and friends to help all those who have been affected by pemphigus and pemphigoid to lead happy and productive lives. These include:

- Educational resources
  - The IPPF Patient Guide - a medically reviewed guide that provides information on the diseases, how they are diagnosed, and common treatment options
  - Patient Checklist - a document to help organize questions and information around doctor appointments and follow-up
  - Tip sheets about a number of topics important to patients in our community
- [Find a Doctor Map](#) - The IPPF maintains a map of medical and dental professionals at no cost and without preference to any physician listed herein. The purpose of this map is to provide patients with contact information for medical and/or dental professionals familiar with P/P. This map is not inclusive by any means. It has been developed from contact with P/P treating physicians, recommendations from patients, and referrals from other physicians.
- [Peer Health Coaches](#) - The IPPF's Peer Health Coaches (PHC) are pemphigus and pemphigoid patients who help more than 1,000 patients and caregivers each year. These specially trained PHCs reduce patient anxiety and uncertainty while providing unbiased disease and treatment knowledge. The goal of our PHC program is to ensure we help every person who needs assistance in the shortest amount of time possible.
- [Patient Education Webinars](#) - The IPPF offers Patient Education Series Webinars featuring leading pemphigus and pemphigoid physicians and scientists. These free webinars are a friendly and casual environment to learn more about specific disease topics.
- [Regional Support Groups](#) - The IPPF has 10 regional support groups around the U.S. and works with many international organizations for these diseases.
- Annual Patient Education Conference - A once a year event that allows patients to gain information from experts and to network with each other in a social and caring environment
- Natural History Study - Launched in March, 2017, the IPPF Natural History Study is a patient registry sponsored by the National Organization for Rare Disorders (NORD) and the US Food and Drug Administration (FDA). This online data system collects, stores, and retrieves patient data for analysis in research studies.
- Advocacy - The IPPF has led the way in voicing the needs of the pemphigus and pemphigoid community. We work closely with our like-minded advocacy partners on the federal and state level to inspire policies supportive of the needs of P/P and other rare disease patients and their families.
- Research - Before new treatment therapies can be developed, investigators need to better estimate the prevalence of P/P, study their pathophysiology, understand treatment patterns, and assess the burden of these diseases on patients. The IPPF encourages research by conducting our Natural History Study, hosting scientific symposia, periodically offering research grants, and maintaining a library of clinical trial information for researchers and healthcare professionals.

## How would a dermatologist best connect their patient with the IPPF?

A physician looking to connect their patients to the IPPF can give them [our website](#), email us at [info@pemphigus.org](mailto:info@pemphigus.org), or have them call us at 916-922-1298.

## How would a dermatologist get involved with the IPPF?

The IPPF is always looking to engage more dermatologists and to connect you to your peers through the IPPF:

- We encourage anybody who is knowledgeable and comfortable treating patients with pemphigus and pemphigoid to contact us so we can add them to our Find a Doctor Map.
- Volunteer to be part of the IPPF Medical Advisory Council
- Become a panelist on one of our Patient Education Webinars
- Become a scientific speaker and present at medical and dental conferences
- Engage with us or partner in research
- Advocate for patients with rare autoimmune blistering diseases at the federal, state and grass root level
- Become a Healing Hero by making a monthly sustaining tax-free donation to the IPPF

We're open to other ideas and are happy to collaborate with you. We look forward to connecting with you!