

**Amethyst:** Welcome, everyone! This call is now being recorded. I would like to thank you for being on the call this evening and to our Sponsors Genentech, Principia Biopharma, Argenx, and Cabaletta Bio for making today's call possible. Today's topic is Peer Support to answer your question about living with pemphigus and pemphigoid with the IPPF's Peer Health Coaches.

***So before we begin, I want to take a quick poll to see how many of you have connected with an IPPF Peer Health Coach (either by phone or email)?***

While you are answering the poll let me introduce you to the IPPF Peer Health coaches:

**Marc Yale** is the Executive Director of the IPPF and also works as a PHC. Marc was diagnosed in 2007 with Cicatricial Pemphigoid, a rare autoimmune blistering skin disease. Like others with a rare disease, he experienced delays in diagnosis and difficulty finding a knowledgeable physician. Eventually, Marc lost his vision from the disease. This inspired him to help others with the disease. In 2008, he joined the IPPF as a Peer Health Coach.

**Becky Strong** is the Outreach Director of the International Pemphigus & Pemphigoid Foundation and also works as a PHC. She was diagnosed with pemphigus vulgaris in 2010 after a 17-month journey that included seeing six different doctors from various specialties. She continues to use this experience to shine a light on the average pemphigus and pemphigoid patient experience of delayed diagnosis and bring attention to how healthcare professionals can change the patient experience.

**Mei Ling Moore** was diagnosed with Pemphigus Vulgaris in February of 2002. After dealing with the challenges of the rare autoimmune disease for 10 years, she became a PHC in the fall of 2012 when she went into remission. In addition to being a PHC, Mei Ling along with Marc Yale organize the Southern California Patient Support Group meetings.

**Rudy Soto** is from the great state of Texas. He was diagnosed in 2009 with Pemphigus Foliaceous and has been in remission since November 2006. Rudy likes the outdoors, cooking and spending time with his loving supportive family. He has been married to his wife Jennifer for 26 years and has 4 children.

**Carolyn Fota** became a Peer Health Coach with IPPF in August 2019. Carolyn was a newly retired U.S. Army Lieutenant Colonel Medical Service Officer when she first experienced the undiagnosed symptoms of Bullous Pemphigoid in October 2015 following a simple same day medical procedure. Carolyn experienced body wide blistering to include oral lesions for almost

six-months before being correctly diagnosed and treated. Carolyn has actively participated in Rare Disease Week on Capitol Hill since 2017, writing articles for IPPFs magazine, Mid-Atlantic Support Group Leader, Representing IPPF at various events and meetings and last year serving as a Peer Health Coach.

So, thank you, everyone for being here. And it looks like, based on the poll, it looks like a little over half of you have connected with a Peer Health Coach, while others have not. So, I hope today's call is very educational. For those that have either connected or not connected with the peer coach. I will do my best to ask questions in the order that they are received. For callers asking a question sometimes, I can see your name, your phone number, or it says blocked. So depending on how you have it set up I may say out loud the last 4 digits of your phone number or your name.

The coaches are here today to answer your questions about living with and managing pemphigus or pemphigoid. They are here to also share their tips and tricks based on their personal experiences. If you ask a question that does not pertain to the call subject I will have to ask you to email me after the call. Also, while we care about everyone's individual journey it can take up too much time on the call and then we can't get to everyone's questions so please if you could just introduce yourself and ask your question we would greatly appreciate it. We received many questions before the call so we will try our best to answer as many questions as we can during the call.

To get us started, for those of you who are either newly diagnosed, or are still kind of learning about the disease. Marc, can you briefly explain the difference between Pemphigus and Pemphigoid?

**Marc:** The difference between Pemphigus and Pemphigoid kind of in simple terms is they're both bullous, immune blistering skin diseases. However Pemphigus generally affects the upper layers of skin like the superficial layers of skin on your body, while Pemphigoid affects the underlying or the lower layers of skin on your body along with what they call the basement membranes. Generally speaking, that's the basic difference, at least, how it appears. There are a few other differences between the diseases, including the type of antibodies that are present in your body. Pemphigus and Pemphigoid have different antibodies that are attacking

your immune system and then they also can present differently because of where they're located in the layers of skin.

Pemphigus Foliaceus is any of the diseases that fall under that umbrella will look differently that you'll still get blisters but they will appear sometimes more like plaques. Versus a disease like blisters where and a lot of times those blisters are actually filled with fluid so they can appear differently. Or do they have different antibodies that kind of distinguish the two, and they affect the different layers of skin. Basic ways that the two diseases differ.

**Amethyst:** Great, thank you. Becky, did you have anything else you'd like to add? I know maybe you wanted to share a little bit on our website about how you can learn a little bit more about the difference of the disease.

**Becky:** On our new website, I just wanted to share with everybody. I know Marc gave a really great overview of what Pemphigus and Pemphigoid are. But if you go to this first tab on our website, you can find out more about Pemphigus and Pemphigoid. If you just click on the header and there's a great overview here and then on the drop-down menu, there's more specific information for Pemphigus and Pemphigoid as well.

**Amethyst:** Great. Thank you. So, we had a question come in. Beth said that she was diagnosed with MMP by an oral pathologist with confirmed blood work, nikolski sign and a biopsy in 2016. Three months after she was diagnosed with MMP, she was then confirmed with Bullous Pemphigoid with a biopsy and multiple blood tests by her dermatologist. that says that she has read about people with BP that may have mucosal involvement. But not anyone that's been diagnosed with both. She's wondering why she's not finding any information on people with both forms of Pemphigoid?

**Marc:** Yeah, that's that's a great question, Beth. Yeah, the reason you're not finding a lot of information about it is it's pretty uncommon to have both diseases but it can happen. So, basically what that means is that you have both the antibodies or MMP and the antibodies, or BP or Bullous Pemphigoid. So, you have both of those antibodies present in your body.

For example, for Bullous Pemphigoid it's BP180 or BP130 and then for Mucous Membrane Pemphigoid you have again the antibodies that are present for Mucous Membrane there isn't a lot of literature about having those diseases, But you can have mucosal involvement with both diseases, both, with BP, and MMP. The, I don't wanna say, good news, because obviously

there's not a lot of good news when it comes to having the disease. Generally speaking, the diseases are treated similarly as are the medications that you would use. Depending on the extent of your mucosal involvement, you will certainly determine the type of treatments that you have, or that your doctor is going to prescribe. But, generally speaking, the treatments are similar. I think where the peer health coaches can really help you, is kind of understanding how to manage the disease or diseases that you have, whether it's oral involvement or other mucosal involvement, areas of involvement. The peer health coaches can certainly help you with trying to understand how to manage that. So, feel free to reach out to any one of us.

**Amethyst:** Ok, great. Laura has a similar question. She was recently diagnosed with PF, Is there any information about the chances of potentially developing PV as well?

**Marc:** Again, it's extremely rare. With PF, you know the antibodies that you will have are desmoglein 3 and then with PV it's desmoglein 1. Sometimes the body can produce both of those antibodies, desmoglein 1 and desmoglein 3. So again, you can have both PV and PF. The treatments and therapies are similar, but it's good to know which antibodies you have because certain therapies are able to treat those antibodies more effectively. But you can indeed have both PF and PV. Same thing, there's not a ton of literature out about having both of those subsets of the disease, but again, it is possible. I encourage you to reach out to Becky and Rudy. Both of them have excellent experience and can really guide you through the treatment odyssey and things that you're going to be experiencing. So, again feel free to reach out to the IPPF. We're here to help.

**Amethyst:** Great, thank you. Mary says that she would like recommendations for the type of doctor to go to for treatment. Right now, she has problems in her mouth. However, she has noticed some small lesions on her genital area. Mei Ling, do you have any suggestions?

**Mei Ling:** If you're seeing inexperienced dermatologists at the moment, you could ask your dermatologist who he or she would recommend for your oral care. If you have, for ladies, OB GYN and for men, probably an internist. I'm not really familiar with Men's Health, but I would think that the internist would get help. Hopefully, the internist and the OB GYN would be familiar with it. I will say I've had below the waist activity luckily my dermatologist at UCLA, who understands the female body and was able to help me with that. But for anybody else, I would suggest that you ask your doctor if they can take a look, if it needs more investigation, then ask if they can give you a referral.

**Amethyst:** Perfect. And Becky, for patients that are maybe new to the community and just were recently diagnosed, what's a good way for them to find a doctor that either has experience or knowledge with treating their condition?

**Becky:** Again, if you go to our website, and under this first link of What Is Pemphigus and Pemphigoid. The first link that comes down is Find a Doctor. If you click on that, you can definitely add some information and you'll be able to access the map. There should be some information there. And then our Find the Doctor map will pop up. Let me just pull that up real quick. And doctors are listed on there, who are able to, who we believe are familiar with. There's a number of different specialities on there. And so you would be able to find a doctor. The easiest way is to search by your zip code or your postcode or the city that you're in. In this example, I'm just going to use Sacramento because that's where the foundation is located. And then, from here, depending on where you live, sometimes we have a lot of doctors. Sometimes there's just a few doctors. Each of these pins on the map represents a doctor, and you may need to come over to the corner to zoom in and zoom out using your plus minus sign over here. Also, worthy of mentioning on our website you can zoom out and then also, if you are not that fluent in English, we also do have some ability to translate our website into a few chosen languages.

**Amethyst:** Wonderful, thank you. We just had a question come in from Judy along the same lines of recommendations for doctors. Judy says she has reinins and asthma under active thyroid alopecia eczema and now diagnosed with BP. Would you recommend that she rheumatologist or who should she see?

**Becky:** I think she has a lot of issues going on, a lot of health issues. And even under the best circumstances, normally, it takes a whole team of doctors to see to take care of us properly. And so a rheumatologist may be familiar with some of the conditions. a dermatologist would be definitely familiar with a Bullous Pemphigoid and her internist, her primary care physician, would be the one that would help her coordinate that care with all of the different specialists and kind of oversee some of the side effects. But she definitely needs to get her doctors talking to one another, and at very minimal, they print out for your after visit paperwork. Definitely keep those in a binder, share them with the doctors. The same things, you should be getting copies of her medical records than any tests, just in case, and she has to take that binder with her, to all of the appointments. So, just in case. We've become very digital, and a lot of times are, there's a lot of information that can be shared in a medical record, an electronic medical record. But, she would have the hard copy to be able to share what doctors are doing with one another, if they are not communicating on their own. But, generally, one thing that I did was, anytime I saw a doctor, when I was newly diagnosed with Pemphigus I asked that it be sent to my medical primary care doctor. That the information was shared with

my oral medicine doctor. That it was shared with all of my different specialists. And so, everybody was all on the same page, and are aware of what one hand was doing.

**Amethyst:** Wonderful. That's a great recommendation. Moving on to some of the symptoms of the diseases. Tom asks, what are some strategies for pain management for blisters? I know, Rudy, you're on the line. Would you like to answer that question?

**Rudy:** So I've found that sometimes doing over the counter pain reliever helped. But also, a lot of my pain came from whenever I would take off my shirt because my blisters were on my torso and lesions were on my torso. So, the shirt, Of course, sticks to the lesion and causes it to begin to heal. And then when you would pull off your shirt it caught, it tears off the existence scab that's coming off so it causes pain. But what I've found to do is, I would put a light, thin shirt that's not really cotton, but there's a thin material that breathable and it helps. And then it also helps put up some kind of ointment on there to kind of create a barrier between the shirt and the lesion. And for sleeping at night, I would also do the same thing. I would create a barrier between my lesions and the sheets or comforter or whatever I was in on and I would do that by using cornstarch. And that kind of creates a little paste and a little layer of a barrier to kind of protect that lesion from oozing out and from sticking to the sheets or whatever you sleep on or your pajamas.

**Amethyst:** Great, thank you. Any of the other coaches have anything to add about pain management and the blisters.

**Mei Ling:** For the ladies, if you happen to have blisters under your breasts, there are bra liners. You can find them on Amazon, and they're very good to create a barrier between if you're wearing a bra, or maybe even a sports bra can be helpful. And you can even put cotton between you and the sports bra if you happen to have blisters there and we're happy to have clusters there. The same thing if you have a waistband male and female, maybe loosen it. If you can, they have buttonhole extenders. you could find online so that you could extend the waste of slags for your jeans or whatever that can be helpful too.

**Becky:** One last thing is I wouldn't be shy about letting your doctor know about how much pain you're in. They're probably going to do some examination and ask you some additional questions, but it may be necessary to get a prescription medicine that may be able to help with

the pain. And the doctor may have some other non pharmacologic options that he has found effective for their patients as well.

**Marc:** Especially, like, if you have oral lesions, too, pain can certainly be problematic. If you have oral lesions, you want to be careful as far as what you're eating in your diet. Also, your physician can prescribe what they, you know, magic mouthwash, or something that can kind of help alleviate the pain in your mouth if you have oral lesions or some sort of topical treatment for oral lesions. So, you know, the best advice is, as Becky said, is just talk to your physician. Don't be shy. Tell them, you know, the extent of the pain you're in and, you know, they can help you, try to manage that.

**Mei Ling:** If I could add another thing, for all of you patients who have blistering all over the body I was there. I did on my upper back and it looks like a minefield. I couldn't shower and I know that a lot of you have that same situation and if you have scalp to shower pressure hitting your scalp. Though, I use buckets of water, lukewarm. You might want to take a lukewarm bath. You might just want to be able to pour water or make sure that when you wash your hair, your scalp, that you're not using your fingernails. Use the pads. Use the pads of your fingers and do a massage, not scratching because then you're going to scratch the scabs. Using water for yourself until you're able to heal will be able to help you.

**Amethyst:** Great, Thank you. That's wonderful advice guys. We have a question that came in from Mo and Mo asked if anyone can talk about their experience with how long it will take for the scars on their skin to fade and become normal looking skin again. And if there's anything to do to speed up. Mei Ling you said it was a bit of a landmine on your back. Any experience from the coaches about what to do with some of the scarring?

**Rudy:** It is a good question. And it all depends on the patient. As far as the scar goes, and how deep the lesions were, because the deeper the lesion and the bigger the blister it's going to take a lot more time to, of course, to heal. And then leaving a scar it takes a lot more time for them to fade away. But, they do fade away in time. I'm still seeing some of the spots leftover from where there was the skin discoloration from the new skin that forms so it's a little bit lighter than the normal pigmentation but they do take time but the more that you're outside and doing things and you'll start to gain your normal, pigmentation back even on the scars that were formed.

**Amethyst:** Moving on to some medicine and treatment questions. How do I know if a medicine is not working for me, and when should I consider trying something else? And it looks like she

says she's been on Rituxan, but she had an allergic reaction. She's on prednisone and cellcept and also uses clobetasol on her skin.

**Marc:** Great question. Generally speaking, these medications have what you call an efficacy rate, so they take a certain amount of time to start to become effective. And then, depending on the dosage of those medications, they can be either more or less effective. Then every medication has a range of dosage kind of a lower dosage to a medium, to maybe even a high dosage. So, depending on the medication you're on, the dosage and the efficacy rate of that medication will determine how long you should kinda wait to see whether or not the therapy or the medication is working, Generally speaking. And the best way to kind of gage whether a medication or a therapy is working is to really look at your body and say, okay, how many lesions do I have? And how frequently are the lesions coming or returning? How long are they staying on my body and how long are they lasting?

So, if the blisters that you're having aren't resolving or they're lasting longer, or they're coming more frequently, generally speaking, the medication is not working. Because the medication should reduce the amount of disease. If the medication you're using is making the blisters not appear so frequently, you're getting less of them. And the amount of blisters you have is getting smaller. Then the medication is working. All of the medications work slowly. You're not going to be able to take a pill or get an infusion and then wake up the next day and then everything's going to be better. It's just not gonna happen, unfortunately. Everything is going to take time. So, the best thing to do is maybe keep a log of your disease activity. So, you can see if it's getting better or if it's getting worse. Like, write it down and on a piece of paper or on a log. That's really a great way to kind of tell whether or not medication or therapy is working.

Then ask your doctor how long those therapies take to start working or how long does it take for therapy to get you to go into remission? Or what's the maximum dosage you're allowed to have? So those are all really good questions to ask your doctor to help you kind of gage whether or not you're on the right amount of cellcept. Or you're taking the right amount of Prednisone to really help you kind of gage whether or not your activity is improving or if it's getting worse. I will say, though, that medication for a long period of time, and that you haven't improved at all. Then, it might be time to ask your physician about other optional therapy's, because if you're on medication for a long time and it's not working or not helping, then you might want to try something else.



**Amethyst:** Great, thank you. We had a couple questions here about prednisone. Sarah asks, what are the long term side effects of prednisone?

**Mei Ling:** We know that long term you can have bone problems, so you do need to take a supplement for your bones and usually like vitamin D, calcium, Magnesium. I recommended extended use can give you problems with diabetes, too, cataracts. But there are side effects and I know a lot of people don't want to go on prednisone. They hear that word and they go, Oh, my god. But we need to remember that aspirin has side effects as well. Now, everybody's different though. No side effects for me, different than it would be for Becky or Marc. Not everybody has the same, but it depends also on, if you're on an immunosuppressive, which will help you taper off the prednisone. So, you may not have the same side effects as somebody else does.

Again, like Marc said, if you notice something is different, since you've been taking it, muscle weakness, muscle spasms, being tired. Also, if you're on a high dosage it's recommended, you take it first thing in the morning so that by the end of the day when you want to go to sleep at night, you're not exactly so manic that, you know, you still can calm down enough to be able to rest. So those are a few of the side effects that if you notice other things going on then discuss it with your doctor.

**Amethyst:** Great. Thank you. Now I know a side effect that a lot of people talk about while being on prednisone is some excess weight gain. So, does anyone have any recommendations on what to do to help weight gain while they're on steroids, and is it ok to exercise even if they have open skin blisters? I was hoping Carolyn might be able to answer this.

**Carolyn:** I was on steroids for quite awhile initially. And the big thing that I did but I did have a lot of sores, head to toe. I also wanted to exercise and I was on a good dose of steroids at the time 60 milligrams for several months. Then, my doctor gradually reduced me. I had a great talk with my doctor at the time. I was into running, yoga, locking a lot of outdoor sports, and golf. And when I was on the medication, my doctor advised me, and the side effects, as Mei Ling had stated, can be serious with steroids, elevated blood pressure, elevated blood sugar. It can have an effect on the bones, the joints. And I did have a lot of skin damage still. And so my doctor said instead of, you know, your usual biking and running and cross-country and a lot of these things at that time, my doctor advised me. 15 minutes of walking in the morning, 15 minutes walking in the afternoon. Yoga, Gentle stretching, was what they had me on for quite a while, because I didn't damaged skin and I was wearing a lot of bandages on my arms and legs, but they wanted to make sure the skin wouldn't get irritated that I would not overdo it or

have another injury. And then, as I gradually reduced and my skin healed, we then, month by month would make small changes to my activities. It was about a year later when a lot of my skin had really healed over. I was able to do more but I was glad I took it easy in the beginning. I was walking twice a day, gentle yoga and stretching in the beginning. I hope that helps and it was important to protect the skin. I hope that does help.

**Amethyst:** Great. Thank you. I know you're talking about some of the things that you're doing while tapering. Julie asks, When tapering prednisone? Is there a tendency to be very tired? She thought it might be the methotrexate that she on, but she is also tapering her prednisone. Mei Ling, any thoughts on tiredness that may come with tapering.

**Mei Ling:** I think you just have to go. I'm a big believer in the mind body use. I know that we worry about being on the steroid, we would worry about having this disease. We're creating a mental exhaustion, so that kinda helps it too. I was sort of high doses like Carolyn, but I started at 100, which was difficult. And I was on 100 for over a year, and I was kinda manic. I mean, at some point, I felt like I could go run around a football field 10 times. So it just depends on the level of activity in your body and how much your dosage is. I think it's something you need to discuss with your doctor and find out if an adjustment can be made and let your doctor know. I'm really tired. I can't move, I have no energy. What can you suggest? What can we do to counteract this? I think having that conversation with your doctor is the best thing to do.

**Becky:** I was going to just add one little thing to that is that sometimes as we come down on the prednisone our body's natural version of it, in my case, my body got pretty lazy, and it wasn't ready to kick up to the level that I was dropping my medicine with. And so we had to go back up on the medicine and then come down at a slower rate. So, I think the biggest key, like anything, with a rare disease is to keep those communications open with your doctor. Don't be afraid to have embarrassing conversations. I think a lot of times, like the tiredness or extremely painful joints. That can happen as we come down on Prednisone, too. I think those things kinda get lost in the shuffle that were so anxious to come down off the prednisone, and it is important to get off the Prednisone as well. But I think we need to be cautious and really listen to what our bodies are telling us and share what our bodies are telling us with our doctors and then they can help us and kind of guide us through the process.

**Amethyst:** Great. Thank you. We've had a lot of questions come in about getting a flu shot. Specifically, one person says that they're taking 10 milligrams of prednisone daily. Is it ok to

get a flu shot if it is a killed vaccine? And the same question, I think, applies for many people who are curious about getting the shingles vaccine, or the pneumonia vaccine.

**Mei Ling:** Generally speaking, for the general population, the flu shot is recommended for anybody over 60. For somebody with the auto immune, compromised condition, the flu shot is definitely recommended. It is a dead virus. So, no matter what the steroid dosages, you will not have any interaction with that. And pneumonia shot. If you do, they do recommend that. That is also a dead virus. The shingles, there's two types because I think this is something I'm not an expert with the shingles I think that is a conversation that a patient would need to have with their doctor.

**Marc:** Yeah, I mean, I think you hit it right on the head. The flu shot is recommended, as long as it's not a live vaccine. pneumonia, certainly recommended, as long as any of these are not a live vaccine. The shingles is a live vaccines. So, I would definitely consult your physician first. I've spoken with people that have gotten the shingles vaccine, had no problem, and I've spoken with people that have that issue. So, you know, I would definitely consult with your physician regarding the shingles vaccine and know that the whole vaccine question obviously is, you know, there's a lot of questions around vaccines, and then of course, you know, what happens when there's a COVID vaccine? What are we going to do? Right? So, we don't really know at this point what that's going to look like, and so the IPPF will certainly work hard with our medical advisers to consult with them on what's the best path forward when it comes to a COVID vaccine, when it becomes available. I guess we just have to stay tuned to find out what that's gonna look like. But, definitely consult with your physician if you're unsure or reach out to a coach. You know, we have lots of medical advisers that work very closely with the IPPF to really kind of help guide us in these recommendations. And, again, as Becky mentioned earlier, you can find a lot of that information on our website, as well.

**Rudy:** Going back to the shingles vaccine, like Marc alluded to, it is the live vaccine. When I started my Rituxan infusions, that was one of the things that my oncologist was worried about, because due to the fact that was the live vaccine. And I had had Chickenpox when I was younger. And I guess, apparently, if you've had the chickenpox, you're susceptible to maybe developing shingles. But my oncologist and my dermatologist recommended oral preventative medications for shingles, which helped me. And I only took it for maybe about a month or two when I was, first beginning my Rituxan treatment. And then I kinda tapered off of that medication, also. So that means there is another alternative to just to do it in a shingles preventative medications other than the vaccine, if that's what you're upset about or somebody's worried about.

**Amethyst:** For a patient that is newer to the community, or maybe on a certain medication now and not sure, if they want to stay on it, how do they know what treatments are safe and effective for their disease?

**Becky:** First and foremost, talking with their doctor, their doctor is going to be the wealth of information and have the most knowledge. If you are looking for a primer on the IPPF website, again, I'll share my screen. You can go to What is Pemphigus and Pemphigoid, the link down here called treatments. And this link discusses commonly used treatments that are used for both Pemphigus and Pemphigoid. Again, there's some information on here about the different phases of treatment, and what that means.

Then the types of therapies with corticosteroids, immunosuppressants, and then with generic and brand names like Rituximab. And then if you keep scrolling to the bottom of the page, there are side effects. Mei Ling kind of already alluded to it like drinking water has a side effect and aspirin and very bluntly considered benign treatments.

These are probably a little bit more scary, but it's definitely something to talk to your doctor. What is common? A lot goes into determining what treatments that are used, including age, our overall health, other different conditions or comorbidities. You might hear that word your doctor using. Definitely educating yourself on what you're comfortable with, talking with your doctor, communicating any fears that you have, or concerns that you have, and then together answering them, but they're on our website, that's a great resource as well. And if any of the patients listening or family members have our patient guide, there is a list of the treatments in there as well.

**Amethyst:** Great, thank you. We've had a ton of questions come in about Rituxan. I'm going to jump right into those. Lynn asked which Rituxan protocol is better? Is that every other week infusion or weekly infusion better and which one is which protocol? I know Marc and Rudy have had experience with Rituxan, so if you guys would like to chime in.

**Marc:** Yeah, so I'll take the latter part, first which one is which? The every other week protocol is what they call the Rheumatoid Arthritis protocol and that's where you get an infusion every other week, two infusions one every other week, one cycle of Rituximab. The other protocol is where you get a weekly infusion and that is Non-Hodgkin's Lymphoma protocol. And that is a

weekly infusion. And there's four of those once a week, and then, that equals one month or one cycle.

Which one is better? I don't know which one is better. I've tried both. There's different opinions on which one is better. Which one's not better. I think a lot of it has to do with your overall health: If you have other medical conditions that could play into that which one might be better and the extent of your disease activity. If you have mild activity versus severe disease, actually, I think all of those things kind of play into that. Generally speaking, If your doctor has recommended Rituximab, I don't think it really matters which one you do just as long as you do it, because I think in the long run, there are benefits to taking it as long as you don't have any type of reactions to the therapy. I don't know if there's a better one than the other? What's your thought Rudy? I don't know what did your doctor say?

**Rudy:** The best thing would be to discuss the plan your doctor comes up with. The plan is like Marc said, depending on your disease activities, that would probably determine which protocol is better, and what you're going through. Like Marc said, I don't think there's a difference, I think it's like you said, I'm going to reiterate. As long as you're getting the treatment, in the long run, it's going to help you out.

**Amethyst:** Sue wants to know, what are the pre medications that are given? Rituxan infusion.(49:20)

**Marc:** Pre medications generally consist of Tylenol, Benadryl, in case you have any type of allergic reaction. And then most physicians prescribe some sort of steroids, like a medrol or a solu-medrol, to make sure that you don't have any type of reaction to the therapy. Those are really the three medications that most physicians use.

I know that sometimes I say medrol or steroids, patients don't want to be on any more steroids than they may already be on, but if you're taking Rituxan, I always think it's better to be safe than sorry, so, even if they do give you some sort of solu-medrol a corticosteroid to help guard against any type of reaction. Those steroids will wear off pretty quickly versus the long term effects of taking oral steroids for a long time. They affect your body differently. One injection versus long period of oral steroids are two different things. You're gonna feel that the injection of steroids, but it will last as long as the oral steroids that you're taking.

**Amethyst:** Great, thank you. Kim from Charlotte, North Carolina just wrote in, and she said she's newly diagnosed with PV. She's currently taking prednisone and hoping to start Rituxan infusions shortly. Of course. I know many people say that they have some of those kinds of pre-fears with some of the unknown. What can she expect from the treatments? Maybe, like, what happens on an infusion day? How does it work?

**Rudy:** On infusion day and it's always best to discuss with your dermatologist or whoever your doctor is, about any medication that you're taking at the moment and if you need to either stop taking those prior to infusion or if you can still take them the day of infusion. I guess some of them may or may not interact with the treatment. So, it's always best to discuss that with them first. But today, the treatment pretty much is you would go in depending on where you do it. Oncology Center or Cancer Treatment Center or rheumatologists and they pretty much talk you through every everything. There's a nurse there present with you at all times. Like Marc shared with those three medications. They give you the three medications prior to treatment, to make sure there's no side effects in could help with the side effects. And I also was given an antacid or any kind of nausea with the amount of medication going into your system. You could get nauseated, so that would help out. But other than that, you just sit back, relax, the treatments began, and you just let the treatment do its thing. The nurses are there to monitor every 15, 20 minutes, or so, they may just drip. They'll start at a very low drip to make sure that you have not experienced any side effects or not. Then, they will continue to Drip a little bit, at a time, Usually like 15 to 20 minutes. It is a long process, Generally takes anywhere from 5 to 8 hours. I think my first treatment was eight hours long.

So, you know, you can usually take your snacks, take something to read, take some headphones, take a laptop, you know, and you will get sleepy with the Benadryl they give you. So, if you fall asleep, it's Ok, just sit back and relax.

**Marc:** The only thing I would add is, I always recommend drinking a lot of water before, during, and then after your infusions. It does just kinda help assimilate any of the medications that you might be taking. Just make sure you stay hydrated. It's also helpful if you're hydrated, because they're going to need to give you an IV. Easily accessible. And so, hydration I think, is a really important part of any type of infusion therapy strength.

**Amethyst:** How long after your Rituxan infusions did you guys start to notice any difference with them, your lesions and your disease activity?

**Rudy:** Well, you know, it all depends every person reacts to the medication differently. It could take 4 to 6 weeks for others it could take it a little bit longer. Generally somewhere between 4 to 6 weeks before you start seeing some kind of improvement and improvement can come into the way of an existing lesion beginning to heal. It could come as maybe you, you see a new lesion appear, but it may not be as pronounced, or as big as other lesions that you've had in the past. So, that's progress, because the lesions are smaller, so that means the medication is beginning to take effect as to what it's supposed to do. It kind of varies on the person, and of course, other medications that they're currently taking, also steroids to suppress. And also that could help them help them speed it up if they are on those two.

**Marc:** Rudy is exactly right, I mean, you're gonna see a slow progression of improvement and goes back to kind of what I was saying before. Take notes, like notice that the lesions are not as big as it was last time, or it's resolving a little bit faster than it was before. Then those are small signs that the medication is working and things are improving. So, it definitely varies per patient. I saw a slow improvement from my last infusion, soft, slow improvement over six months until eventually I didn't have any lesions. They were all gone. And they didn't come back and I was in remission. It really depends on the patient, but you have to be patient and you have to kind of, you'll see a slow improvement.

**Amethyst:** Great, thank you. We still have lots of questions about Rituxan, but I'm gonna ask two more and then we're gonna move on. I know we're running out of time. Hopefully, we can get a few more questions in here. Deborah asks, can you share how restrictive life has become after you completed Rituxan treatment while you build your immune system? Should you be staying at home or not participating in any activities?

**Marc:** It's not worse than COVID that's for sure. You have to be cautious like you would like you would because you have a suppressed immune system. So, you know, don't share utensils, don't drink out of other people's water cups. If you notice someone that's sick, you probably want to avoid them if they're visually ill. But you can go out and you can participate and do normal activities as long as you feel like you can do that. So, you know, goes back to what we were saying about listening to your body and listening to how your body is feeling tired, rest. If you feel like you have some energy, then it's ok to go out and do something, Now with COVID everybody's wearing masks. You're probably ok because everybody's wearing masks. So. It's about the same. I mean, you have a suppressed immune systems and Rituxan is gonna suppress your immune system even more. So, you just have to be cautious. But you don't have to be isolated and let you know and live in your house, 24 hours of it, that you don't have to do.

**Marc:** Ok, real quick, Donald asked, how rare is it for a patient to develop acute heart failure after three rounds of Rituximab his dermatologists thinks that it may have caused his heart condition.

**Becky:** Amethyst, you shared that question with us beforehand. I did reach out to some of our medical advisers and they said that they haven't seen any literature on that. But the Rituximab can cause some heart arrhythmias and some other issues. And that there's also a fluid load issue. And so that they recommended that while they haven't heard of that, that they have a complete markup by a cardiologist and have the dermatologists. and cardiologists really work through to see what the root cause was. And also it may be related to the prednisone if he was on prednisone. So, those were some of the issues I've reached out to a couple of our medical advisers and those are the issues that they pointed out. So, I hope that's helpful and that he can share the information with his doctors as well.

**Amethyst:** Great, thank you. Thanks for doing some research on that Becky. We've had a few questions about scalp lesions. Both Rhonda and Tammy asked about how do you manage scalp lesions? What's the best thing to do for that?

**Mei Ling:** I think when I first got it, nobody really knew what to do. I used Johnson and Johnson's baby shampoo until I finally got a dermatologist, I think clobetasol makes the shampoo, Clobecks. Any shampoo doesn't have a lot of additives. My reason for using Johnson and Johnson, if it doesn't make a baby cry, that it would make me cry. It was awful, I had stuff falling out of my hair. But like I said, use the fingers to massage. Just pour water, gently, maybe from a faucet, big pots of water, because the shower pressure can hurt your head as well. When I went to the hairdressers, please don't pull on my hair like this because it would be hurting my scalp. I am in remission but I still have moments when my scalp will feel like it's got a heavy sunburn. So you have to be very careful with that.

I will also add, though, if you have scalp issues, especially if you are going up in the sun to wear a cloth hat, do not use a straw hat, because the sun's rays filter through, and you can get a sunburn.

**Carolyn:** I think you made a lot of good recommendations and that there is a shampoo available and I think Marc mentioned it too that does have some steroid clobetasol in that and you need to make sure you pay attention to the directions on that, how much to wash and how frequently and that can help a lot with any scalp lesions. And then just keeping those lesions clean and just using a very gentle shampoo. My dermatologist had prescribed one to me at the time. And actually just recently and just keeping the scalp clean, not using products. What



helped me was using a baby comb and just gently, when I cut my hair, not using a product to include a hairdryer or a curler. For those who may want to use that just coming gently, drying gently with a towel too. And I notice those kinds of things really helped, I also think wearing some kind of a cloth kind of cap. If you're going outside, it helps a lot too. Those kinds of things would help me a lot and make sure you're keeping it nice and clean and dry too. Cool showers, not hot. I like showers directly under the shower. Don't be hitting it so hard. On a nice gentle kinda shower. Put that on and cool too.

**Amethyst:** Does anyone have any experience with what to do if they have sores in their throats? Is there a throat spray that's recommended to use?

**Mei Ling:** I had it in my larynx. So, I remember those days there was a time when I only drank Ensure for three weeks. I wouldn't do that today. I would make my own smoothies. The magic mouthwash, I would gargle with it and be sure to spit it out, you could also gargle with, or what they call pulling. You can use coconut oil. But make sure that it goes in the trash can and not in your sink, because it clogs up the pipes, and then you end up with a plumbing bill. But otherwise, you may just want to ask your oral care doctor if you have a dentist oral care who specializes in helping or your dermatologist. Using the magic Mouthwash, helped me and the clobetasol.

**Marc:** Yeah, esophageal lesions are really, really challenging and fun. A lot of times, they're tied to the nasal cavity. So, you know, you may have them in your nasal cavity and then you're swallowing. And then you also have them in your throat or in your mouth. So, they do make like a nasacort spray that will help with. Then as far as the oral lesions are concerned, there's a couple other things you can do. One of the things that my physician recommended was a steam inhaler. You can buy the steam inhaler at the drugstore. What you do is, you put a little bit of distilled water in it. You plug it in and basically create steam. And then you put your nose and your mouth over the steam inhaler then create steam and it goes into your nose in your mouth and then, down into your throat. And that helps alleviate some of the issues that you might be having with the mucosal tissues in your nose and your throat. That's something you can look into seems to help quite a bit for myself and other people that I've spoken with.

**Becky:** And one other thing is I know it sounds funny because they're in our mouth, throat, and esophagus but let your doctor know. Especially if they're not getting any better because an adjustment may be needed to your systemic medication either in a change in dosages or a new medicine. And then, a lot of times, too, if you're especially having trouble, like when you're eating. One thing, that, it was kind of like a lightbulb that went on after the fact, and I really wish somebody would have suggested it to me, but seeing a registered dietitian, somebody

with a license. They have a lot of great ideas of how you can get really solid nutrition and ways to prepare foods. And they think about the PH of food. So, it's not stinging your mouth, and if you can't eat, you can't heal. And if you can't heal, you can't eat, it's kind of a vicious cycle. So, one thing that I wish somebody would have done for me very early on, instead of like pretty late in the process was to see a registered dietitian.

**Amethyst:** Thank you. And along those lines, Becky, I know we get a lot of questions about diet. You know, what should they be eating? Specifically Deborah asked, Has anyone tried a plant based or dairy free diet? She's been going vegan now for four weeks and that she thought she saw an improvement in her mouth and she's not on any medication at the moment. And I know along those lines a lot of people ask about these specific things. Should they be Paleo, Keto? Should they be taking things out of their diet? Any recommendations for what they should be doing with their diet?

**Becky:** Sure. So, generally our medical experts our medical advisers recommend a healthy balanced diet in moderation. Really truly, one thing that worked for me was to use a food journal. And I'm really lazy. So, I would take a picture of my food with and then that way, I just put it into a food journal. And I would see what would happen the next day? If I got woken up with more lesions or less lesions, or if it hurt when I ate. And I would repeat it the next time that I ate the same meal. It was a trial and error. A lot of people say that including tannins in them makes their oral lesions worse, and that's what I refer to as everything good. In life like chocolate, wine, coffee, tea, those kinds of things, that they cut out of their diet, and made them feel better. I've heard other people say that when they cut dairy or gluten out of their diet, it's made them feel better, and I think it's really just kind of individual.

At this point, there was a study done by Dr. Kyle Amber, out of the University of Chicago. And he did study. If anybody would like to read that journal article, they can contact the Foundation, and we'd be happy to share that with them. And a little plug, I think he's also going to be, at our virtual conference, one of our speakers. So, if anybody is interested in learning more, from Dr. Amber, and what he has found out with his research setting food, I encourage them to attend and listen.

**Mei Ling:** Depending on your cultural background. Latino people have a lot of hot peppers. Obviously, if you're going to have oral lesions this is where I'm pertaining to. If you're having oral activity and you have enchiladas and so you might not be able to have them for a while because of the tomato and the acidity. Indian food is very high in curries and turmeric and gingers. I couldn't eat tomatoes for years until remission but now I can eat all the tomatoes I want. And also, if you have oral activity, salt because salt can hurt and irritate. Hard crackers.

A lot of patients will say, Oh, I got this blister in my mouth and it's red, and I'm bleeding. It's nothing to really get in about, because those go away. They dissolve within 24 hours. So, we all know you might want to be careful, and like Becky says, you know, keep a food journal or take photos. Then if you happen to get a blister or activity, you'll be able to look back and see what you ate that might have contributed to it. Mostly it's experimenting, trial and error. Maybe one day you can eat something with items, maybe another day you'll cut it out. Some people can't, we're all different so keeping a food journal is helpful but be very careful with condiments.

**Amethyst:** I know we've already run a lot of questions. Does everyone mind if we stay on for a few more? Alright. Thank you, everyone. So, Susan, just a messaged, and she wants to know, What is magic mouthwash. Hopefully, it's magic.

**Becky:** General. Generally, it's taking on that a lot of doctors have. And a lot of them have their own formula for it. Generally, it has some anti-inflammatory, whether it'd be prednisone. And some of them have lidocaine that helps. Or another medicine that helps with the pain is and mine had meloxicam and my doctor felt that helped it stick better to the surfaces, but they generally all have their different concoctions, but it's a medicine. That can help with the pain, or the inflammation and the oral lesions. I wish I could be more specific and talk about exactly what it is, but I think that's kind of a good overview.

**Marc:** I remember my doctor said it was called BMX like the motor Bike and I was like BMX? What's BMX? Vantage, drill my land enzyme. I came, so this with the X was for links disease. And so I was on the BMX. Benadryl, Mylanta and xylocaine. So, I was on the BMX that was a formula that I use one thing to do is make sure you ask your doctor about formulation. Because sometimes it can be more expensive, depending on the formulation, and so the cost of the BMX, or the magic mouthwash can be quite expensive. So you know, ask questions, ask your doctor, ask her about the formulation. You know, there may be a formulation that might be less expensive.

**Becky:** And generally too, a lot of pharmacies may or may not have the ingredients. I had to go to a specialty pharmacy to have them mix that up. So, if, by chance, your doctor does prescribe it to you, generally, they know where you can get it. So, that might be a question just to keep in the back of your mind as well.

**Amethyst:** Thank you. While we're talking about some oral care here. Jennifer said that she's been prescribed clobetasol and told to ignore the warnings that are not to be using it. And

also, along those same lines, does anyone have any recommendations for soft toothbrushes and toothpaste?

**Becky:** I'm happy to take this line. So if your doctor has prescribed clobetasol orally. I think it's rather in my opinion, like that kind of happened to me, to my doctors, or maybe red flag stickers and telling you don't use it internally. But trust me, it's going to be ok. And when I came. I wasn't prepared for the amount not to use internally. And I called her, anyway. So when in doubt, always call and ask your doctor. But I did use it internally in my mouth as well.

As far as toothbrushes and toothpaste. There are a lot of great brands on the market. Jennifer Harmon, I think it was last February on our website the archives for the Patient Education Calls, had a great call about different products to use. One of the things that she talked about during that time was a curaprox toothbrush, which is a bit of a fancy toothbrush. Personally, I would use a toddler's toothbrush in my own experience. They're generally very soft, very round and they're very small. And then she also had some recommendations and her call about what she lets her patients now use as far as toothpaste. But again, like the Becky method was to use a children's toothpaste. it wasn't meant to be flavored, and they tend to be gels instead of toothpaste, which tends to be a little bit less abrasive in my mouth. And then when I brushed, I truly focused on brushing one tooth at a time. It wasn't like trying to get the right side of my mouth or left side kind of a thing, but truly focusing on brushing and my technique, which is something that I think dentists should spend a little bit more time teaching us and brushing our teeth our whole life, but I think sometimes we can get some tips and tricks about how to do it right to help with the pain, too, though. Use your hygienist dentist. They're an underutilized resource.

**Carolyn:** And I want to add something to that as well. I'm only smiling because Becky was kind of my call out coach, supportive friend, as I was having a lot of happy dental work this spring, and even this summer with some activity. And I've used the clobetasol and am too and one thing that has helped is wherever that sore is to use something like a q-tip, and to place it on there. And I'll allow it to absorb and make sure not to be eating, or drinking right after that, to allow it to absorb and or talk to your dentist, too, about being fit for dental, and that's helped me a lot. And then I'll use a q-tip to put it on wherever I may have a blister or sore. And this is after I've done my dental hygiene enough, rinsed my mouth with lukewarm water, now put the clobetasol on that area that's bothering me. And then a dental tray, I'll do the same time to allow it to chore for a couple of hours before going to bed. And I also use a switch out to, for, like a night guard rounding and doing any damage to gums. They're a little bit on the frail. And

those kinds of things have really helped me in addition to what Becky has just said to those are really great suggestions.

**Amethyst:** Great. Those are some good suggestions on how to keep that clobetasol in your mouth. Thank you. Let's see, there are so many questions here. I know we get a lot of questions about different supplements that one should take. So, D asks, do you have any recommendations? Do you recommend taking supplements with Prednisone? Said, he was recommended a few supplements by a Naturopathic Doctor to support his liver at your immune system and gut biome, and it started having some pain after taking these supplements.

**Mei Ling:** If I can make a suggestion, at one point, because I was listening to everybody say, Well, you should take a whole lot. I went to my regular primary care doctor and said, should I be taking this? And he suggested taking a blood test that finds out if you actually are insufficient with certain vitamins. Don't just go out and arbitrarily take a lot of just because you think you need it, you might be doing more harm. The loop to find out am I really lacking in magnesium? And how much should I take? Do I take vitamin K, do I really need B 12 different? So I would suggest having that discussion first before you go shopping. That's great.

**Marc:** That's really a good point Mei Ling and the only thing I would add to that is there are some supplements that need to be taken with prednisone and methotrexate in particular. So, if your doctors prescribed prednisone. Generally speaking, you want to be on some sort of calcium supplement because it will deplete your calcium levels. And that's why a bone scan is recommended, when you start taking prednisone to make sure that you don't have bone loss because osteoporosis is one of the major side effects of prednisone. So, certainly calcium, but again, you want to do it under the recommendation of your physician.

And then the other one is vitamin D, D as in dog, that's another vitamin that gets depleted by the use of prednisone but I can definitely can soldier position first because you could have high levels of those. Your body could be producing high levels of those and you may not need to supplement. The other one is with methotrexate. Folic acid is recommended because the methotrexate depletes the folic acid in your body. So, folic acid is recommended with methotrexate.

The last one, actually is if for some reason your doctor has prescribed you dapsone, dapsone causes anemia. And so you want to get your iron levels checked by your physician. So those are kind of the three exceptions are three rules of thumb with those particular medications. But

other than that Mei Ling is right, I mean, you want to consult your doctor to see if you have any deficiencies in any of these vitamin levels before you just rush out and start taking a bunch of different vitamins because somebody recommended them.

**Becky:** Just one other thing, it's important to remember that the medicines that our dermatologists are prescribing or doctor treating, our Pemphigus and Pemphigoid are medicines are meant to knock down our immune system a little bit. Especially with some of the different supplements that are out there and even some of the vitamins they are immune boosting, which is pretty much the last thing that we want to do. So, any medicine, no matter what you think it is, whether it's over the counter or prescription, let your doctor know who's treating your Pemphigus and Pemphigoid just to make sure that you're not trying to fight the medicine, that they're prescribing as well, that everything is working in harmony.

**Amethyst:** Great, thank you guys. I have one other question about some alternative care, Brenda asks: Is chiropractic care good as an alternative option to management of Pemphigus Vulgaris since spinal health and alignment has direct impact on body function, illness and disease.

**Becky:** I haven't personally seen any research that supports chiropractic care and Pemphigus and Pemphigoid, that's not to say it might not help. I can't speak to that. One thing I learned is that in general, chiropractic care involves somebody pushing or pressing on your skin, which can cause little micro abrasions in your skin or little traumatic things, which could lead to if you have an open wound. Or if you have a lesion forming, it could break that lesion, or it could create an opportunity for additional skin lesions. So, I would just check it out. I kind of feel like I'm a broken record and I really hate that, but I would talk to your dermatologist or your treating physician just to see their opinion and to see if it might be recommended for you.

**Amethyst:** Great, thank you. I know we've definitely gotten past our time everybody and we do, thank you for staying on the call. I have one last question for everyone before we finish up here. What is the best way to connect with you guys after this call?

**Marc:** E-mail. It's our name, then, at pemphigus dot org, so that's the easiest way, so you can contact us directly, or you can go onto our website, and there's a section on our website called Ask a Coach. It's on the very first page. You can't miss it, if you scroll down, it says Ask a Coach, or there's a drop-down under Peer Health Coaches, And you can ask your question, and we'll contact you. We're here, We're here for you. Just feel free to feel free to reach out, every question is a good question. There's no such thing as a bad question. We're here to help

you, direct you to a qualified physician or to answer any questions you have regarding therapies or treatments or how to manage your disease. That's what we're here for.

**Amethyst:** Great, thank you. Well, thank you, everyone for staying on the call with us. That was definitely a quick hour and a half. We still have lots of questions to get to, so hopefully, we'll have another call with all of the coaches. I know you guys provided some great information for us, and they're so knowledgeable, so we appreciate you guys for being on the call with us today and for everyone speaking with us.

I would also like to give a huge thank you to everyone on the call for joining us today and thank you to Genentech, Principia Biopharma, Argenx, Cabaletta Bio for helping to make today's call possible.

Before we go, I have a few announcements:

I am excited to announce that this year's Patient Education Conference will be from Friday, October 2nd to Sunday, October 4th and will be virtual due to the COVID-19 pandemic. The IPPF invites you to attend the 3 day event from the comfort of your home. The 2020 Virtual Patient Conference will be an exciting and educational event for patients, caregiver, physician, researcher, and stakeholders in the field of bullous disease. Topics that will be discussed will include: Treatments, Oral Care, Diet and Autoimmunity, Research, Trials and much more! Registration will be opening soon so keep your eye out for an email as well as the registration link on the IPPF's website.

If you have not registered for the IPPF's natural history study we encourage you to do so. 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). You can register today at [www.pemphigus.iamrare.org](http://www.pemphigus.iamrare.org). This online data system collects, stores, and retrieves patient data for analysis in research studies. The more data we can collect, the better the information we can give to researchers, the sooner they can find better treatments, earlier diagnosis, and one day – **A CURE!**

The IPPF needs your help. Unfortunately due to the ongoing COVID-19 pandemic, the world is facing unprecedented challenges and the IPPF is not immune. Recent donations are down. You can help our community by becoming a Healing Hero you go above and beyond to support the IPPF community. A Healing Hero makes sustaining, monthly gifts to support the IPPF's mission of improving the quality of life for all those affected by pemphigus and pemphigoid. Every dollar donated makes a huge difference and even a small monthly donation will allow the IPPF to continue to help you and others in the way that our community has come to expect.

Lastly, If you have a question that didn't get answered on the call, or have additional questions please e-mail Becky Strong, at [becky@pemphigus.org](mailto:becky@pemphigus.org), or call (916) 922-1298 x:105, and we would be more than happy to help.

This call recording will be sent out with the survey following this call.