

Becky: Welcome everyone. This call is now being recorded. I would like to thank you each for being on the call this evening. Our speakers today are the IPPF Peer Health Coaches, Marc Yale, Janet Segall, Mei Ling Moore and Rudy Soto. So thank you for joining us today. The call today will be a question and answer session with the peer health coaches to provide answers to your tough questions about living with pemphigus and pemphigoid. So first let me introduce you to our speakers. Marc Yale is the Executive Director the IPPF and also works as a peer health coach. 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 in finding a knowledgeable physician. Eventually, Marc lost his vision from this disease. This inspired him to help others with the disease. In 2008, he joined the IPPF as a peer health coach. He works with people to improve their quality of life and encourage them to become advocates. In 2009 he helped develop the pemphigus and pemphigoid comprehensive disease profile, giving experts insight into the patient perspective. He is a two time Rare Voice Award nominee, a Global Genes Rare Champion of Hope nominee and a national advocate for rare disease.

Becky: Mei Ling Moore was diagnosed with Pemphigus Vulgaris in February of 2002. After dealing with the challenges of a rare autoimmune disease for 10 years, she became a peer health coach in the fall of 2012 when she went into remission. In addition to being a peer health coach, Mei Ling along with Marc Yale organizes the southern California patients support group meetings. Prior to getting PV, she has spent over 25 years in the entertainment industry and 10 years as an administrator for an award winning architect.

Janet Seagall is the founder of the IPPF. She was diagnosed in 1983 and started the foundation in 1994 with the express purpose of giving people living with this very rare disease, a place to go and find information and find others also living with pemphigus. Janet graduated with a Bachelor's of Science degree in Psychology. After College, she worked at Pilgrim State Mental Institution on Long Island. Upon moving to San Francisco, she volunteered at San Francisco mental health department assisting the psychiatrist in the psychodrama group. She also worked for the Women's Refuge in Berkeley, California. In 2009 Janet received her certification as a hypnotherapist and began working with the foundation as a coach 2015.

Becky: Rudy Soto is from the great state of Texas. He was diagnosed in 2009 with Pemphigus Folioscus and has been in remission since November of 2006. Rudy likes outdoors, cooking and spending time with his loving, supportive family. He's been married to his wife, Jennifer for 25 years and have four children. And now it is my pleasure to introduce the IPPF peer health coaches here to answer your questions about living, managing and thriving with pemphigus and pemphigoid. So welcome team.

Marc: Thank you Becky.

Rudy: Hello

Becky: Great, so let's just jump right in if it's all right with you and we're going to start asking questions. So our first question is, does this disease ever go into permanent remission?

Janet: Well, the answer is yes and no. You can go into remission for very, very long time, the rest of your life but the disease could also come back at any time.

Becky: And Janet, how long have you been in remission?

Janet: Since 2002 but I still get lesions in my mouth from time to time. They just go away by themselves.

Becky: Great. And which of our other coaches are in remission? If you wouldn't mind just sharing with our listeners, how long have you each been in remission?

Marc: Go ahead and go Rudy.

Rudy: Oh, hello. My name is Rudy Soto. I have a PF and I've been in remission, it'll be three years this November.

Becky: Well, congratulations Rudy.

Rudy: Thank you.

Becky: Mei Ling are you on the call with us too? I know Mei Ling was having a little bit of a problem with her audio there for a little bit, so I don't know if she was able to get back on.

Mei Ling: Hello, it's me. It's Mei Ling. I'm sorry, I'm back on

Becky: Oh, hey. We were just saying, what disease do you have and how long have you been in remission?

Mei Ling: I have pemphigus vulgaris. I got it in October, 2001, diagnosed in February of 2002 and finally it went into remission in 2012, which I've been in ever since.

Becky: Congratulations and Mei Ling just beat me to the punch by a little bit. This is Becky and I have pemphigus vulgaris and I've been in remission since the end of 2013. So great question. So Janet, there's a question and they ask, how does the disease work?

Janet: Yeah. So what happens is, is that a genetic marker and anybody who has pemphigus or pemphigoid has this genetic marker and something turns it on. Something gets into your body, we don't know what it is. It could be stress that lets in a virus or bacteria, it could be a virus or bacteria on its own. It could be some sickness that you had. It could be a drug that you took and we don't know exactly what causes it. But what happens is, is that the genetic marker then sends a signal. You have an immune system that is made up of T cells and B cells basically. And T cells tell B cells what to do. So what happens here is that the genetic marker will tell the T cell to tell the B cell to make antibodies against the glue that keeps skin cells together and then that's what causes the blister. I hope that was clear and everybody understood that.

Becky: Yeah. Well I think that was a great explanation. Thanks Janet. Rudy, this might be one in your wheelhouse. Margolet is saying that she has been having a strong breakout of PF even though she has no special worries or in your usual circumstances now. PF has been with her for a number of years, but thanks to taking Cellcept she's been able to take it for many years and had a good result with minimal outbreaks. So she's strongly wondering why these past couple of weeks, she's had an intensified breakout of strong itching, red marks on her arms and upper body, the normal signs of a PF problem.

Rudy: Oh, that's a good question. Now, what was, what was her name?

Becky: Margolet

Rudy: Well, I know sometimes, having PF, sometimes you get little itchy red marks. It could be the sign of maybe not a flare, but maybe a blister happening. And a lot of times it happens for me when I'm outside in the sun for a very long period of time. Before a little small blister forms, for me, I get a little redness and it becomes a little itchy. So what I do is I use an anti-itch lotion. I have been able to find it over the counter. I usually apply some of that on there. And that usually helps with the itching and then it kind of goes away within a couple of days. I was also prescribed an ointment that I use if I were to get any kind of small little blisters or anything from being out in the sun for a long period of time.

Becky: Great. To any of the coaches have anything that they would like to add? Okay. Well Sue is asking a question that I think that as a coach I've had asked of me quite a bit. She was wondering if we could offer any help for potential PV food triggers. She says many articles and people discuss garlic, the onion family and lemon and citrus as big no nos. But there are so many other potential triggers and I'm particularly struggling with navigating fruits and vegetables. Short of trying an autoimmune protocol diet, is there any advice such as not eating veggies raw or buying organic when possible? She says that she understands it's highly individualistic, but anything that we could help share would very much help her.

Mei Ling: If you are having a lot of activity in your mouth at the moment, I would recommend staying away from sharp things, crackers or celery, carrots that are hard. I used to get blood blisters, just from Romaine lettuce cause the stem of the central part of it was irritating. If you're not really having a lot of activity, you still would want to keep a daily diary of every food that you eat because in case you do get a flare up you might be able to go back and see what was it that caused you to be so sensitive to have a blistering.

Janet: I also think that sometimes when you first start with a disease, any food could trigger a blister in your mouth. When I was in remission or when I was in the drugs, but I wasn't having any blisters, I would do my own thing with garlic and I could see if garlic caused any problems for me. So I would test myself a couple of times and see if it made a difference. And I had to eat quite a bit of garlic for it to make a blister in my mouth but this was after I was still on drugs but had no blisters. So it a really individual

thing and it could be any food at the beginning that triggers it, but what Mei Ling was talking about was great.

Marc: I wanted to just add a couple of things. I think both Janet and Mei Ling did a great job of expressing the foods that probably should be avoided. And actually the IPPF recently helped recruit patients for a study that was just published in the Journal of Venereology and Dermatology in the European Academy of Medicine. And we actually had 200 patients that I participated in this study. And the results were very interesting and we found that through this study a number of things irritated patients when they were consuming them, including things like alcohol, citrus, fast food and fatty foods, fried foods. Garlic as Janet and Mei Ling both mentioned were high on that list. Nuts, so many, nuts, which can be particularly irritating to the mouth and can cause trauma in the mouth when you're eating it. And then spicy foods. So these are all things that can certainly irritate and exacerbate the disease, particularly if you have them in the mouth. And you know, those types of foods should definitely be avoided if possible.

Becky: Great. I'm going to throw my two cents in here too. I had a real problem and I didn't even realize it with pineapple. I love pineapple if I ate it, when I had the disease I would be out of commission for days. And it turns out that there's a protein in pineapple that helps to break down proteins. So you're not supposed to use it as a marinade. Well, apparently it was in my mouth to shreds. So that's one thing. The other thing is if anybody is listening, if you're not keeping a food journal, that was one thing that really helped me. I would take a picture with my cell phone and I could just put it into my food album and I could notice if later that day or even the next couple of days if my mouth really kind of ballooned up with lesions and was a lot worse, I could look back to see what I ate and see if I could find any correlation. For me, the big things were pineapple and sharp foods. I still get an occasional lesion from time to time. And one of my precursors is I like spicy foods. And so things that I can normally eat spice wise become very intolerant where I'm going to be getting a lesion. So, I don't know if anybody else has anything like that, tips like that, but I thought maybe that would help. The other thing is I've heard different patients tell me like going gluten free and wheat free or dairy free or sugar, reducing the amount of sugars has helped. But I am not aware of any research. Are any of the other coaches aware of any research toward those things?

Janet: Well there was something about a lot of chocolates. Dr. Worth in Philadelphia told me years ago that she had a patient who ate five pounds of chocolate and that's a lot of chocolates who got lesions in their mouth. I don't know how anybody can actually eat five pounds. But that's what she said. And she said they got pemphigus. So, I know that if I eat too much chocolate, I'll get some sores in my mouth. So lots of chocolate.

Marc: We lost Becky so I can actually jump in and navigate here. So there was a question about going out into the sun. So maybe Janet, you could maybe make a few comments about going out into the sun, if you have the disease whether or not you should be going out into the sun or not?

Janet: Well, you can go out in the sun but don't stay out there very long. Keep covered, especially if you have lesions.

Marc: Mei Ling, did you have anything you wanted to mention about that? There was some concern about patients not going out into the sun.

Mei Ling: Yes, especially if you play tennis or you golf, you need to wear a cloth hat because the sun's rays will filter through straw. And everybody you need to use sunscreen, but if you can use a 50 plus SPF. 35 minimum. And if you're doing waters keep reapplying. And also you do need sunscreen and for the ladies there is some sunscreen put into makeup bases, so you can look for an SPF there as well. But on a cloudy day, you're likely to think that the sun can't harm you because it's not shining. It actually is and being filtered by the clouds. So you have just as much of an intensity from the sun's rays on a cloudy day as you would on a regular day when the sun is bright. If you go camping, there's SPF clothing that you can wear, cover your arms. Don't forget that water reflection, just like being in the snow when you go skiing you can get a sunburn that way. You can get a sunburn if you're just staring at the sidewalk and it's too hot. Not that anybody stares at the sidewalk, but I'm just letting you know that the intensity from it on a very hot sunny day can be dangerous. So you want to limit yourself. Also, remember it used to be between the hours of 10:00 AM and 2:00 PM the sun's rays are at their strongest. It does also depend upon where you live, so you can decide that for yourself.

Becky: Great. Thanks Mei Ling. Rudy, do you have any advice to pass on as well?

Rudy: To go along with what Mei Ling said, there's a couple of companies out there and one of the companies they make real thin, moisture wicking fishing shirts that I bought. And that's what you use in the summertime when I'll go out in the sun. And a lot of them, the ones that I use are long sleeve and they're also got the SPF and UPF built into the shirt. They're generally about 35 UPF or SPF. There's also some other companies that are a little bit more expensive out there that make those type of shirts in a real thin, they're good for the summertime. They pull the moisture and sweat from your body, so it kind of cools you off of as the wind blows. But that's what I didn't really use and they work really well.

Becky: Great, I just want to add one thing for me, the chemical sunscreens tend to irritate my skin. I had sensitive skin before I had PV. But I find that there are a lot of lotions and creams out there on the market for your face as well as your body that have the titanium oxide in them. And titanium oxide, for those of us that are old enough to remember is like the white creams that lifeguards used to put on their noses and on their faces. And I will say that you do get a little bit of a sheen not anywhere near back to what the lifeguards had. But they really look pretty natural and they do help protect. And I tend to layer different sunscreens. Big thing when you're putting sunscreen on, the average adult human takes an ounce of sunscreen and it usually needs to be reapplied according to the manufacturer's instructions, but generally every couple hours. So Marc there was a question and Zina is asking what were the symptoms that you were having in your eye that caused the blindness?

Marc: Yeah so I did lose the vision in just one of my eyes, my left eye. The first symptoms were it was red, it was itchy and I thought, well, maybe I just had an eye infection. And so I went to an ophthalmologist to see if I had an eye infection because those were kind of the symptoms of a sort of infection. And then I started to also have some discharge come out of the eye. So those symptoms are very common. Some of the other symptoms include photo phobia, which is light sensitivity. You can also

get what they call like a gritty feeling in your eye or it feels like maybe there's something in your eye like sand when you blink. So that's very common symptom. And then of course, the other symptom is your eyelashes start to turn in and grow inward, which causes other issues with ocular disease. So those are probably the most common symptoms that you'll notice. And then of course, as the disease progressive you will end up getting scarring or you may have scarring in your eye, in the lower area of your eye. Your eye might eventually kind of be droopy and you might have a hard time keeping it open. So those are definitely some of the more progression symptoms as time goes on. But, you know, I just recommend that if you feel like you have any of the symptoms that I mentioned in your eye that you want to reach out to a qualified eye specialist to make sure that you get it checked out. And the IPPF has a list of those physicians to refer people to.

Becky: Great. Marc real quick, when somebody is diagnosed with pemphigoid, especially since we know that it tends to affect the eyes more than pemphigus, what are some recommendations that you can give? Like should somebody see an ophthalmologist even before they're having any symptoms in their eye or any of the things that you've talked about?

Marc: Yeah, I would certainly recommend once you're diagnosed to go see an eye specialist just to kind of rule that out. So it can't hurt to go see an eye specialist. And then from there on out, certainly you want to make sure you have regular appointments with the eye specialist because the disease can certainly progress. You want to make sure that you're getting checked out frequently. So yeah, I definitely think it's a good idea to see an ophthalmologist, especially someone that maybe has experience with ocular diseases. Someone like a, an optician or optometrist or somebody that you might get glasses from is probably not the right person to see. You want to see someone that actually has experience with cornea issues and that sort of thing. So definitely an eye specialist and I definitely would recommend you see them right away.

Becky: Great, thanks Marc. Dolores is asking, to what extent might pemphigus complicate dental work given I have artificial joints and require antibiotics prior to deep cleanings and other significant dental work. I'm in remission and I'm concerned about the use of antibiotics.

Janet: My suggestion would be if the doctor wants you taking antibiotics, you don't have to, but if doctor wants you to take antibiotics, you probably should, but don't take penicillin. They don't give that very often anymore, but penicillin might be a trigger. There was some clear indication that it is. But you could take an antibiotic it just depends on what the doctor thinks that's a good idea or not great.

Mei Ling: I've had occasion like in the winter, like if I'm getting a virus with a cold or whatever, my doctor will give me a z pack and it has never affected me. I am sensitive Amoxicillin 'm allergic to, more or less, but I haven't had a problem with the Z-pack and I think it just depends on the individual physiological makeup because everybody's different.

Becky: Great, thank you. There was just a call last month to, Dolores, Dr. Joel Laudenbach discussed oral issues with pemphigus. And if you send me an email after the call, I'm happy to send you the link from

our website to that call as well. Our next question comes from Salma who asked after 20 years of PV, is there a chance of remission? If not, should I be looking for an alternative to Cellcept only treatment?

Janet: A lot of people have been on medications for a very, very long time. Cellcept is one of those medications that usually people are on for a very long time. If you wanted to get off the Cellcept you can ask the doctor about Rituxan. That might be another drug that could help you get off that drug. 20 years is a long time to be on one drug. So you could still have remission with it, but it's one of those drugs that most people have to take for a very long time.

Marc: Yeah concur with Janet, if you've been on Cellcept for 20 years and you haven't had any remission, then I would suggest you speak with your physician about maybe an alternative treatment like Rituxan. If you haven't had remission in 20 years, it's probably not going to happen. So if you're looking to get off the medication altogether and be in remission then I think it would be wise to take some other alternatives like Rituxan.

Becky: Great, and just as a reminder on the IPPF website there is a list of treatments. Selma especially, but anybody if you want me to share that link with you, please feel free to send me an email after the call. Linda says I have pemphigus vulgaris in my mouth and my question is for the people in remission, what medication were you using? I'm currently on methotrexate and some days are worse than others.

Janet: I was on mostly Prednisone because I was diagnosed in 1980 and only Prednisone was available. But I took it for a very long time and went into remission. Then came back to Prednisone again, went into another remission. But then as the third time when it broke out, they put me on Imuran and I was on Imuran for five years. Methotrexate is not considered the best choice of medications to take for pemphigus because it's milder but some people do very well on it. So if you're not doing well on it, then you may want to discuss changing drugs.

Becky: Great. Thanks, Janet. Anybody have anything else they'd like to add?

Marc: I would, similarly to the last question about the Cellcept and I think Janet's right, if you're on methotrexate or you've been on methotrexate for awhile. If these medications are controlling your disease and it's manageable I think that's okay. But again, if you're looking to get off medications altogether or you're looking to go into remission then again, you might want to have that conversation with your physician about the other options that are available. The one thing I will say with methotrexate to the caller's question is to make sure that you're taking folic acid along with the methotrexate because the methotrexate does deplete your folic acid. So please make sure you're taking folic acid and you should consult with your physician about the dosage for that.

Becky: Great. Thanks Marc. We've kind of been talking about remission and there is a question on our list. Patients want to know what do we mean by remission? What do the doctors mean? And is there even a test to test if we're in remission?

Marc: I was going to say, so generally the consensus in the medical field is that a remission means that you have no disease activity whatsoever for at least two months. So that is the clinical definition of remission. Now bear in mind that that can happen in two ways. One could be on medication with no disease activity or one could be off of medication with no disease activity, but you have no disease activity for at least two months and that is considered to be clinical remission. And then as far as a test is concerned there really is no test to see if you're in remission because as a patient or as a person with Pemphigus or Pemphigoid we will always have these antibodies in our system. So if they do a test a blood test or they do some sort of test to see if the antibodies are present in your system, it's highly likely that those antibodies are going to show up and you may not have any disease activity but they do a test, they're still going to show the antibodies are present. So you always have those underlying antibodies in your system. Even though you don't have any disease activity, you'll still have those antibodies present. So the best way to really tell whether or not you are in remission or not is based on that clinical activity and whether or not you have any disease activity happening in your mouth or on your skin or anywhere. So, but two months is kind of that bar, that's where we set the bar. So no disease activity for two months.

Becky: Great. Thanks Marc. Anybody have anything else they want to join in? Okay, so I've gotten quite a few questions about this so I'm going to just read one and this person says, well, just for a minute, can we talk about the itch? I seem to have an insatiable itch even when I don't even have any pemphigoid lesions. What can I do about it?

Janet: Yeah. Well it's hard to get rid of itching. There are topicals you can use. The allergy medicines can sometimes help with itching. Benadryl sometimes if you can take them. That's a difficult question to answer. Itching can be really annoying for sure. I had hives recently and I just took over the counter stuff but there may be stronger stuff that the doctor could recommend.

Marc: Yeah. Just a couple of quick comments on itching. At the onset of the call, I didn't have an opportunity to share but unfortunately I'm not in remission at the moment and itching is definitely one of those symptoms that I've been experiencing quite a bit of. And Janet's, right. I mean, there are some topical things you can use topical creams, anti-histamines, those types of things. But currently there aren't any major treatments that really address the itching which can be very burdensome and super annoying, right? So it's just really itchy. And I understand completely, so I know that there are a couple of therapies that are being looked at currently by a couple of different companies to maybe address the itching component of the disease but none of them are on the market or in clinical trials at the moment. But I think it's something that is being recognized as being a challenge and discomfort for patients. And I think, you know, as we move down the road with new therapies, I think we're gonna see that issue being addressed with the medications that are coming out to trial and hopefully coming to market.

Rudy: What I've tended to use is, I just went and found some over the counter lotions. It's an anti-itch lotion and it really helped out with the itching. It wasn't as bad as it as it would've been. These are lotions that I could find at my local grocery store or a local Walmart supercenter. They're really not very expensive and if you have it on, like I had it on my arms the itching, and my legs. So because it was also a

lotion and also moisturized my skin, so it would prevent it from drying. I think one of them was Aveno anti-itch and then the other one was a Curel.

Becky: Great, thanks Rudy.

Marc: Hey Becky, one more comment on the itching if I can?

Becky: Yeah, absolutely.

Marc: Something I just thought about. So I don't know if everybody understands why the disease causes the itching. And so I just wanted to just quickly talk about why itching is associated with this disease. And it's because in your layers of your skin, there are sensory nerves that are next to located adjacent to where the skin is separating and that's what helps cause that itching of the skin. And so a lot of times patients or people will go to the doctor and say, you know, I'm really itching, I don't have, you know, any blisters but I'm itching really bad. And the doctor might look at them a little sideways, like maybe it might be in their head. But the reality is that because this is a subcutaneous disease that you can't always see what's happening. And so if a patient has a lot of itching they definitely need to address that with their physician. It's not psychological often about in their head its actually physically occurring in the skin.

Becky: Great. That's great information, Marc. Thank you. Linda says that she's been taking Methotrexate and the folic acid and she's been on it for a month, but her doctor wants her to take Rituxan. She is afraid of any of the meds because of the side effects. So has anyone taken Rituxan and how did it work for you?

Rudy: I went through all the protocols of having all of the oral medications and none of them worked for me. So we finally had to do the Rituxan treatment. There are side effects associated with using it. Some of them could be real mild as a sore throat during the treatment. Or some itchiness of the skin or where they do the IV. Because it's an IV treatment there could be some itchiness or redness there. But if you experience any of those side effects while you're doing it typically there's a nurse there with you and just let the nurse know. And what they'll do is they'll give you some better Benedryl to counteract the side effect of what you're having and then they'll slow the treatment down on the Rituxan. I had seven treatments and through those seven treatments I didn't experience any type of side effects during treatment or after treatment at all.

Becky: Great. Thanks Rudy. Anybody else have experience with Rituxan that they would like to share?

Marc: I was just gonna kind of expand on what Rudy said a little bit. He is absolutely right there's a nurse with you there during the infusion and they'll monitor you very closely. They will also make sure that they infuse it very slowly particularly on the first infusion of the cycle just to make sure that you are handling the medication okay. They'll always give you what they call pre-medications to make sure that you don't have any reaction to the treatment. So typically they give you Benadryl, Tylenol and it's recommended that they give you an injection of Medrol or some sort of corticosteroid to make sure again that you are tolerating the medication. If you have an issue it usually will happen during that

infusion, and as Rudy said, you want to make sure that you notify the nurse immediately if you have any type of negative reaction to the treatment. But other than that the side effects, the typical side effects are pretty benign, not any worse than any of the other treatments that I was on. And I had quite a severe infusions of Rituxan myself. I had a little bit of constipation, a little bit of diarrhea, but other than that the side effects were as I said pretty benign.

Becky: Great. Thank you. This person says that they got Rituxan and their blood pressure got a little wonky. Is this a common side effect or is there anything that I can do or that the nurses can do to help prevent this from happening next time?

Marc: I guess it depends on what she means by wonky. They will certainly monitor your blood pressure throughout the infusion. If it gets really high I think that they may consider slowing down the infusion. But there could be multiple reasons why the blood pressure did get higher or wonky, including just the stress of doing the infusion itself can certainly cause high blood pressure. If you are ingesting caffeine, those types of things. I mean, I think there's lots of reasons why your blood pressure can be high. So it's just determining whether or not it's actually from the medication itself or some other reasons. But if it is from the medication or if they think it's from the medication, again, I think that the physician or the attending physician or whoever's on staff while they're getting the infusion would want to slow the infusion to make sure that you're tolerating it okay.

Becky: Great. Thank you. Barbara says, "I'm in remission right now, but if the pemphigus comes back, does it attack the same areas where you had pemphigus before or does it start attacking a different part of your body?"

Janet: I'd love to answer that question because I've been in remission off and on since 2002 mostly often I get a few lesions in the same place in my mouth. I don't know why it just happens to be an area in the back left side of my mouth that flares. And I don't know why. It seems to be the only place that I get a flare. So I suppose it could happen anywhere, but the fact that it does in that particular area for me, I know that it's going to go away by itself

Becky: Great. Anybody else have an opinion or want to share their experience? I can share my experience. My lesions generally happen in the same area of my body. If I get an occasional transient lesion, they happen on my tongue or on my cheeks or on my gums, it's not necessarily in the same place, but it generally affects my oral cavity. Some of us are lucky that way but I have heard other patients say that I used to get it on my leg, now I have it on my chest or vice versa. So I think it's a great question to ask the doctors, but I'm just not sure if there is a right answer to that, at least from the patient perspective.

Becky: So Debbie is asking, how soon would PML or this kind of side effects show up and is there a blood test that would indicate whether or not you carry a virus or anything prior to taking Rituxan?

Marc: I was going to say that if PML is potentially an issue, it would show up or almost immediately during Rituxan. For those of you who don't know what PML is, it's progressive multifocal

leukoencephalopathy. To my knowledge, I don't know if there's a test for it prior to the infusion, but there is some standard workup that needs to happen before you do Rituxan including blood work. Standard blood work should be done. Generally speaking they might want to do a chest X-ray just to make sure that you don't have any pneumonia or anything like that. You should also certainly mentioned any type of conditions that you have with your physician prior to them prescribing. But PML is basically in neurological condition. And again, to my knowledge, there isn't a test prior to determine if someone has PML. But I think that there maybe some type of markers in the blood work that would help physicians identify whether or not that's an issue.

Becky: Great, thank you Marc. There is another question. I am considering taking IVIG, what should I expect when I take an IVIG infusion?

Marc: Hey Becky just to jump back real quick because I know we were just talking about this PML issue, I know people may have a concern about this. It looks like there is what they call a JC virus. Maybe that person mentioned that can be detected but it's a DNA test. So typically patients don't get a DNA test prior to any this kind of thing. And just doing a DNA test is not considered a standard test to be done before any of these types of infusions. But I would say if someone had a history of neurological disease in their family it might not be a bad idea to maybe do that DNA test prior to getting the infusion.

Becky: Great. Thanks Marc.

Marc: I'm sorry, you were talking about IVIG.

Becky: Yup. What should somebody expect when getting an IVIG infusion?

Marc: I can answer that cause I've done IVIG a few times and I know we just recently also had a call about IVIG about a month or so ago. So IVIG as you mentioned is an infusion. It's infused very slowly through an IV so it takes quite a bit of time. So definitely expect that you'll spend a lot of time at either the infusion center or wherever the infusion is being done. Again, it's monitored by a nurse and they will check your temperature, your blood pressure that sort of thing throughout the infusion. okay. The other thing I think you should expect is because it's an immune modulator and it takes a long time to take effect you have to have multiple infusions over quite a bit of time. So if anything, I think it takes a lot of time to do. Again, it's similar to what we mentioned about the Rituxan. If you're going to have a reaction it usually happens almost immediately. Side effects again, were very similar to the Rituxan including headaches. Headaches can sometimes be a problem or problematic with IVIG, but again, they give you those pre-medications. But you can eat during the infusion, you can sleep during the infusion, you can pretty much do everything as normal it just takes a long time. Like my infusions usually take eight hours.

Becky: Well, great Marc. That's a lot of great information. The next question, Patty asks, how about effects on other areas of the body, not just the mouth? And how about avoiding tannins as recommended on the website. Any dietary advice for lesions on the body?

Mei Ling: Can you elaborate on that question Becky?

Becky: That's all it says so I guess they're asking like before we talked about how dietary, like sharp things and avoiding fruits, helped with oral lesions. Is there anything, as far as diet that is recommended to help lesions on the skin?

Mei Ling: Not really. I mean the diet is mostly for the oral matters. Anything to do with the skin would be topical.

Becky: I talked with doctor Donna Colton a little bit about this at the last patient conference. And just to throw my 2 cents in here, that was one of the questions I asked as far as diet is. Like what can patients do to help whether the lesions are in their mouth or on their skin. And she said the best advice that doctors can give at this time is to eat a healthy balanced diet in moderation. And then keep an eye like we had talked about for oral lesions, keep an eye on things and if you notice that after you eat a certain food, your disease gets worse, then you might want to consider taking that out of the rotation of your diet. But really there's no real research that has been done to extensively look at the effects of diet and the whole body and what could potentially be triggers. So I don't know if that helps. If anybody has anything that they would like to add?

Mei Ling: I would like to add a comment. I have been seeing a lot of comments from patients on the Pemphigus Vulgaris pages regarding whether the AIP diet is exactly the antidote or a gluten free diet or the Keto Diet, which is a fad and not recommended. There is no real diet that I am aware of that is going to help alleviate the activity from the autoimmune system for the blistering. I think if somebody, if you practice like deep breathing and yoga and doing this to take your mind off of the disease and relaxing, doing some deep breathing. Keeping the stress factor down is the most important ingredient to keeping the activity from spiking. And that's my personal experience on it and I know that a lot of people will say yes, after I had a stressful situation that it really went into overdrive. And so that's the only thing that I can offer on this. Maybe somebody else can add something else.

Marc: I would just say along the line of what Becky and Mai Ling said, eat fresh food and avoid processed food. Eat fresh green leafy vegetables. Eat in moderation. I mean the deal is you're immune system is under attack and so you want to try to make sure you bring your immune system back in balance. And by avoiding foods that are bad for you, overall will help your immune system. Don't eat fast food, don't eat a lot of sweets. Just eat fresh food and in moderation. I think that's probably the best advice that anyone can give.

Becky: Great. Well, coaches, this was a very quick hour, but our last thing, if we wouldn't mind just kind of go around amongst us. One thing with having any disease, let alone something that is rare, like pemphigus or pemphigoid, a lot of times you end up feeling quite down with this disease. It can be quite isolating. Um, what advice would you have for those out there who are listening that might feel the same way that we've felt once?

Rudy: First thing is, know that you're not alone. That there's others that are going through and have gone through what you're going through. Reach out to somebody, whether it's a peer health coach,

reach out to anybody that you know that can help you and listen to you and understand what you're going through. That, that was a big help for me knowing that I wasn't alone and somebody was out there going through the same things that I've gone through or that I was going through. That's my biggest thing.

Janet: This disease can take a really big toll on your emotions, especially after you've been really, really healthy and you can't figure out why did I get this kind of thing. I mean, I've been so healthy all my life and this disease is not about health, but there are as Rudy said, people out there to support you. When I was diagnosed, there wasn't anything. So use the Facebook pages and Instagram and the Foundation to connect with people and contact peer health coaches. That's what peer health coaches are for, so you know you're not alone. We can't answer every question but we can certainly be there to support you. And I would also suggest talk to your family and let them know what you're feeling. Because I went a long time without discussing this with anybody and I think it was a mistake. So I think it's good to talk to your family, let them know how you're feeling because it can be misconstrued, especially if you're taking Prednisone.

Becky: Great. Thanks Janet.

Mei Ling: I can make another little suggestion here. If any of you would like to speak with one of us. Send us an email and say, please, I'd like to speak with you or your personal peer health coach you have been assigned to. You can ask a question at "Ask A Coach" or the Info email and say, I need to speak to somebody and one of us will call you back and we can have a conversation.

Becky: Great, thanks Mei Ling.

Marc: I guess the only thing I can add to all of those wonderful suggestions would be that when you are first diagnosed you feel like you have lost control and you don't, you have no control over your body and no control over what's happening. I think people need to find out how to get that right control back in their life. And that can come in many shapes and forms. It could be reaching out to the IPPF, as Rudy said, or talking with your family as Janet mentioned, or connecting with the IPPF or just even learning more about disease and become educated so that you are an expert on the disease. So I think there's lots of ways that you can take that control back. Advocate for yourself, be the expert. I think you know, as Rudy said, especially at the beginning to know you're not alone. There are other people that can share their experience with you and relate to what's happening and you can talk to them through social media or come to the patient conference and meet other people. So I think there's lots of ways to manage it, but don't be ashamed of the disease. Don't let it cause anxiety or depression. It can be very scary and daunting, but you're not alone, as Rudy said. And you know, just take that control back, figure out what it is to help you take that back and be in charge. Don't let the disease define who you are. Because once you do that, then you're headed down the wrong path.

Becky: Right Yeah. I agree with everything that you guys said. One thing for me, I was fairly healthy before all of this and my biggest health concern might have been the cold or you know, bronchitis or something like that. But I had to give myself a little bit of time grieve for my health. It was truly a major

loss for me. And I had to let myself feel that so I can get past it. With that being said, something that I wished somebody would've suggested to me a little bit earlier in my journey than they eventually did was to go talk to somebody. Even if they know nothing about Pemphigus, but going to talk to a counselor who kind of like helped you to feel okay with that loss of control right. That it's okay to feel that way, but then to help you take those steps to either to find some new coping mechanisms that are a little bit healthier than what I was using as well. And just different ways to take control back step-by-step were huge things for me. And then again, just like the other coaches finding somebody else and, and connecting. Right. So I didn't feel so alone like I was on an island. So all great, great experiences that we've shared today. So thank you. It was a really quick hours, a little bit longer than an hour.

Becky: I would like to say thank you to all of our coaches for being on the call with us today. It was extremely educational having you here. So thank you. And I'd also like to give a huge thank you to everyone who joined us on the call and listened in and participated. And a huge thank you to Genentech and Celgene for helping to make today's call possible. I do have a few announcements that the IPPF has kicked off our third quarter fundraising. Funds raised this quarter are used to support Patient Services such as this call today. Because the IPPF provides services free-of-charge to the community, your small donation will instantly make a huge impact! Other patient services include our Peer Health Coach Program and Local and Regional Support groups. Please consider visiting our website today to make a tax-free donation to support the valuable programs and assistance the IPPF provides to pemphigus and pemphigoid patients and families. The IPPF is also pleased to announce that we have opened registration for the 2019 IPPF Annual Patient Education Conference. The conference will take place in Philadelphia from October 11-13th. This is our 25th year as an organization and we hope that you will join us this year for an educational and fun weekend in the city of brotherly love! Early registration ends August 30, so go online and register before then to take advantage of your discounted registration prices. Mark your calendars, We hope to see you there!

If you have not heard, the IPPF has a natural history study! If you have not registered for the IPPF Natural History Study, we encourage you to do so. The IPPF Natural History study is a new 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. 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!** Our next Patient Education Call will be on Thursday, August 1st from 12-1pm EST with Dr. Kyle Amber, Dermatologist at the University of Illinois, Chicago to discuss your general treatment questions for the treatment of pemphigus and pemphigoid. Registration details for the August call will be on our website. Lastly, If you have a question that didn't get answered on the call, or have additional questions please e-mail me, Becky, at becky@pemphigus.org, or call me at (916) 922-1298 x:105, and I would be more than happy to help.

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