

May 21, 2020- IVIg Patient Education Call Transcription

Becky: Welcome everyone. This call is now being recorded. I would like to thank you for being on the call with us today and a big thank you to our sponsors Genentech, Principia Biopharma, argenx and Cabaletta Bio for making today's call possible. Today's call topic is IVIg for pemphigus and pemphigoid with Michelle Greer. So first, let me introduce you to our speaker. Michelle Greer is the senior vice president in charge of sales for Nufactor. She has 15 years of experience with plasma administration management and sales. Michelle has strong national relationships with key opinion leaders in the autoimmune and immune deficiency arena. Michelle has achieved a Bachelors of Science in nursing as well as an immune globulin certified nurse credential. So now it is my pleasure to introduce Michelle Greer to answer your questions on pemphigus and pemphigoid. Welcome Michelle.

Michelle: Hi everybody. Thanks for having me.

Becky: Thank you for joining us. I'm going to launch a quick poll here and start off our call. I'm just curious today to see how many people have ever received IVIg for pemphigus or pemphigoid as part of their treatment? So if you could take a couple seconds, if you're on the call and answer the question, we would really appreciate it. As people are starting to fill out the survey, Michelle, can you explain what IVIg is and what it's made of?

Michelle: Sure. So IVIg stands for intravenous immunoglobulin and immune globulins are antibodies and they're found in blood plasma. IVIg is made from many donors. From many many human donors and what they do is they take plasma and they separate out the immune globulins, and then they purify it so that it's free from any virus and they turn it into something that is infusible into the human body.

Becky: Great. It looks like Michelle that about 30% of the people on our call have had IVIg as treatment. So our first question from a patient says, Jacob asks that when I receive IVIg will each day's infusion be made from a different batch of serum, or will it be the same blood mixture makeup as the first day or will it be different?

Michelle: So that's a really good question and the answer is maybe, maybe not. Traditionally for pemphigus and pemphigoid the dosage is pretty high. It's based on body weight and it's 2 grams per kilogram is the typical dose. That's a lot of drug to receive in one shot. So what they typically do is break that up into about a five infusion, sometimes it could be a little less three to five. It depends on how people tolerate it. Typically that one, five days therapy will come from

one batch. However the next dose, a month later, because it's a monthly infusion for quite a while. The next dose could be a different lot number. But it'll be the same brand but it could be a slightly different composition.

Becky: Great, thank you. Who can get IVIg? Is there any part of our population who can't get an IVIg infusion?

Michelle: So again, it all depends on how people tolerate. Anybody's a candidate even if you have things that might make the infusion more complicated. So for example, all drugs have something called a black box warning. So on the package insert, there might be a black box that will say, pay special attention to people with this or pay attention to this potential reaction. So for example on IVIg, the box warnings are renal failure that was associated with a product from many years ago that contains sugar as its stabilizer and that product is no longer on the market. All of the products since then whether they have sugar or not, a lot of them have different stabilizers like amino acids these days but that black box warning remains. So we're always going to look at somebody's renal status, their kidney function. We're going to monitor that if they're at higher risk for kidney complications, but doesn't mean that a patient can't get it. The other black box warning is clotting, a thrombotic event it's called. People who are at higher risk for a blood clot or have had things like deep vein thrombosis, heart attack, or stroke that was due to a clot. It doesn't mean they can't get it, it just makes the infusion a little more challenging. It's really up to the doctor looking at each individual patient deciding if IVIg is a good treatment for that patient.

Becky: Great. Thank you. You mentioned earlier that somebody got an infusion over 5 days. Is that the typical length of time that it's given and how long does an infusion take?

Michelle: So again, it's 2 grams per kilogram is the typical dosing. So if I am 75 kilograms that means I need 150 grams of drug. Conservative would be 30 grams a day for 5 days. If you tolerate that or if you can tolerate more than typically, we follow like a 10-gram an hour rule at home. So, it all depends on if the patient wants to get it in the shortest amount of time and the doctor's okay with that. We might be able to do 50 grams a day for three days. Something like that is totally doable. So it depends on the patient's weight and their total dose because the larger amount is infused in one sitting. As the infusion goes on and you're getting that drug most of the reactions are infusion related so if you get a lot in one sitting you might be more prone to some side effects. We start conservative and we can ramp it up if somebody tolerates it no problem to make it a quicker infusion or over less days. Does that answer the question?

Becky: Yeah, you did great. Thank you. Are there different brands of IVIg? And can you explain the differences and is there a difference in formulation from IVIg brands?

Michelle: Sure. So there are many different brands and after the call I'll forward you a link to Nufactors website. They actually have a brand comparison chart you can either order one or just view it online and it'll show you the different brands. Some of them are just for IV and some of them are just for subcutaneous administration, which is more given to people with immune deficiencies because the dosing is lower. Then some brands are okay for both IV and subcutaneous use. The difference, a lot of them there are some newer formulations and then there's some that have come off the market. In general they're very similar, but there are differences. Typically the concentration can vary. For IV it comes in 5% or 10%. 10% would be half the volume of a 5% solution and the stabilizer might be different, the manufacturing process might be different. But once you're stable on one product you should stay on that unless you have to change for whatever reason such as shortages or insurance changes or something like that.

Becky: Great speaking of insurance. We got a question from one of our listeners who want to know is IVIg covered by insurance in particular Medicare?

Michelle: It is, so with Medicare there's several parts of Medicare. There is Medicare Part B which covers home care. Part D if you opt to have a prescription drug plan through Medicare. You'll have a Medicare Part D plan. If you get it in the hospital or a physician's office, it's going to be covered under Part B, if you in the home setting it'll be covered under Part D through the prescription drug plan.

Becky: Great. Thank you. So how does IVIg work to control pemphigus and pemphigoid?

Michelle: So IVIg works in a lot of different ways. It depends on the disease state, but there's a lot of different theories on how it works in autoimmune conditions. In pemphigus and pemphigoid, well let me back up. In autoimmune diseases like pemphigus and pemphigoid there's a lot of inflammation that's going on and IVIg will help to control the immune response in terms of suppressing that auto antibody production and then the inflammation can subside. So it's not a cure but it can control the condition and it can put you into a permanent remission or long term remission.

Becky: Great. Thank you. So you mentioned before some of the side effects of IVIg. Can you go over what some of them might be?

Michelle: Sure. So again, a lot of the reactions are infusion related. So the infusion should really be tailored to the patient. Anything else, any additions that they might have, any past experiences with IVIg, any allergies and we tailor the infusion to the patient. We like to start out slowly and based on their response we will increase it in 15 minute to half-hour increments until we get to what is their maximum rate. So half the infusion is spent ramping up and the other half of the infusion is at the maximum rate. Typical side effects that might occur are headache, nausea, vomiting, blood pressure fluctuations, fatigue, flu-like symptoms because you're getting all these extra antibodies, you're creating an immune response. So you might feel achy, you might have a fever, things like that. Those are all manageable through either the pre medications, they are key. Hydrating before and during and after the infusion is really important. The nurse being there in front of the patients so if they're having some reactions, they might slow the rate back down again things like that. Some of the more serious complications that are way less common are the Black Box warnings that I talked about before, so renal failure, thrombotic event and another one is aseptic meningitis. That's different from a typical headache that you might experience. It's very severe headache, light-sensitive, vomiting. You could end up in the hospital with that or at least the emergency room. It doesn't mean you can't continue with the therapy. Again, that would be up to you and your doctor based on what you're experiencing, what you want to do and if the physician feels comfortable continuing.

Becky: Great, and whether we get it at home or an infusion center, our nurses that will be there with us they'll be able to handle and they'll be monitoring to intervene as quickly as possible, correct?

Michelle: Correct. That's really important to understand the therapy. At Nufactor a pharmacist will call the patient before starting the therapy and do a health review and a lot of education just so you know what to expect and it helps them to tailor the infusion and the infusion rate and everything and pick the brand if they have the choice. A lot of times there's formulary brand selection by the insurance company. So we have to try that but just knowing what to expect going in. Being able to tell the nurse, you know what, I'm starting to get a little bit of a headache she can back off. You can repeat the pre-meds which are Tylenol and Benadryl, typically.

Becky: Well, great thank you. Is IVIg typically given with other therapies or is it given alone by itself as a standalone treatment for pemphigus and pemphigoid?

Michelle: So again that depends on your physician and the severity of the illness. So I'm sure that some of the people on this call have or are receiving Rituxan for their pemphigus or pemphigoid that can be used as a standalone treatment as well. Sometimes somebody might start with IVIg and if they're still having flares or they're unable to taper prednisone or

something like that you could either switch to Rituximab or there are a few physicians that will use a combination therapy and they have a certain protocol that they follow where they'll have IVIg and Rituximab. So basically Rituximab depletes antibodies. So it works by trying to remove and stop the production of the auto antibody that way and then IVIg actually gives a lot of extra antibodies and hopes of suppressing the immune response. So there are some doctors that will give Rituximab to deplete and then IVIg to put back the antibodies so they don't deplete your antibodies. You might be on steroids while on IVIg which again the goal is to wean off because there's so many nasty side effects of steroids and you shouldn't be on them high dose for a long time or other oral agents such as immune suppressants.

Becky: Great. Thank you. I think we're kind of touching on our next question. Tahira says I am due for a Rituxan booster as I'm starting a mild flare-up of pemphigus vulgaris. Due to the COVID situation my doctors put me on 20 milligrams of Prednisone to be reduced to 15 milligrams until I get Rituxan next month. I have had IVIg in the past. Should I suggest to my doctor the IVIg instead of Rituxan?

Michelle: I mean you could certainly ask your physician if that's something that you can get at home while you're waiting to go back to outpatient. Or if you want to try that and switch over to home infusion of IVIg and see if that will control your condition. That's certainly something that you can discuss with your doctor to see if that's agreeable for the two of you.

Becky: Great. Thank you. And this is another COVID question. In light of COVID-19, I've heard that IVIg can be given at home. Is this right? And is it easier now since the great pandemic has hit us?

Michelle: It's really no different. Before COVID-19 you could get home infusion of IVIG. I think some people would prefer that now because they don't want to have to go to a hospital outpatient setting. Conversely there's some people who don't want anybody in their homes so they're going to go outpatient. So much unknown right now still with COVID and it's all about somebody's comfort level. So if you are comfortable going out and you prefer outpatient infusions, that's what you should do. If you're not comfortable going out and you want and you're okay with somebody coming into your home you should absolutely look into home infusion services. Aside from COVID there's a lot of benefits to home infusion, again it's all about what you want. There's the convenience of scheduling. We infuse a lot of people who are working on the weekends and around their work schedules in the evenings. So it's really just up to you.

Becky: Some people do work overnight shifts and are used to being up at night. Can a person get their infusions on a midnight shift or an afternoon shift or is it purely during the day? How does that work Michelle?

Michelle: I don't know that we would give it overnight but again, it's about finding a nurse. If you wanted to infuse from 5 to 10 at night and we found a nurse who was willing to do that in a patient's home, which I believe we have, then yes, that's possible.

Becky: Great. Thanks.

Michelle: Overnight is a little less desirable because if there is a problem you don't want to have to try to wake the physician or the pharmacy. You'd rather have it during working hours or at least semi working hours.

Becky: No, that's a great point and things to consider when we're thinking about doing a home therapy. Our next question comes from Bonnie and she asks, is there a current shortage of some brands due to the use of IVIg for COVID-19.

Michelle: So shortages, it really just depends on who you're asking. At Nufactor right now we're not having any problems getting any brands. Certain hospitals might. It is currently another link I can send which I discussed with you earlier. It actually has a listing of any drug including the different formulations of immune globulin that are in shortage. Right now all of the brands of IVIg or at least most, the ones that I checked they are on this list. But again, I'm not really hearing that it's a problem like it has been in past years and even last year. There are definitely less people donating plasma right now due to COVID and so there could be shortages that are more notable coming up. So donate your plasma or encourage people to donate plasma.

Becky: So you kind of addressed the next question that was submitted to us. Has COVID-19 affected getting plasma for IVIg and although patients like us probably are not candidates, especially with active disease to donate, are there ways that the IPPF and our community can encourage others to donate?

Michelle: Being you know an advocate for blood donation in general and encouraging friends and family members to donate plasma right now is crucial.

Becky: If you're listening and you're one of the people that are getting IVIg therapy, explaining to your friends and your loved ones too might help them encourage when they know how it's helped you to be able to help you and our patients in the future by donating.

Michelle: You can look up specifically plasma collection centers for the various brands or manufacturers of IVIg in your area and there's ways to donate. So I can send you that and a couple of those links as well.

Becky: Perfect. Thank you. I imagine it takes a while to make IVIg. If somebody donates today, how long until the plasma that they donate is a usable product and can be given to a patient.

Michelle: 18 months.

Becky: Wow. Wow. I was not expecting that answer. This next question says can I get COVID-19 from IVIg?

Michelle: No, COVID-19 is an airborne virus. So you will not get it by any type of blood or body fluid including IVIg. Now at some point there may be donors that have antibodies to COVID and actually there are some manufacturers that are looking for those specific donors right now to help develop some treatments. It's very desirable to have those but you will not actually get the virus, sick from the virus by receiving antibodies. It's actually good.

Becky: Our next question is along the same lines, will getting IVIg cause me to test positive for COVID-19 or another disease if somebody donated and has it?

Michelle: So when you're looking for active conditions, like where you actually have that disease state they're looking for antigens not antibodies. So if you take a blood test and say you have antibodies to certain disease states, it may be because you've had a vaccine or you've been exposed prior to something and you've created antibodies to it. From what I know and again, I'm not an expert on this, but from what I know the COVID test is actually a nasal swab looking for what we call antigens. Antigens are the foreign bodies that are not supposed to be there. So they're looking for that on the nasal swab results. If you have antibodies received through IVIg, that's not going to make you test positive on a nasal swab.

Becky: Okay. Thank you. So our next question, from the patient's perspective getting IVIg from Nufactor or another company, is it just like like Rituximab where your doctor would submit to the insurance for pre-approval and then the patient just shows up at the infusion site and used

by their doctor or is there other steps involved? If I can get my doctor to write a prescription for IVIg, now what? What happens and what's the process I guess is a better way.

Michelle: So if you want a home infusion and you want to use Nufactor or another home provider and again your insurance company may mandate you use a certain one or give you choices and want to be contracted and be in network with your insurance company. At Nufactor we like to get our own authorization. So all your doctor would have to do is send a prescription over and then some notes describing pemphigus or pemphigoid. Sometimes the insurance companies like to actually see the proof of the diagnosis. So biopsy and maybe some labs that were done that confirm a diagnosis of pemphigus or pemphigoid. Some insurance companies also like to see that you tried and failed other medications. Steroids being the most common one and sometimes immunosuppressants as well. Then we would submit that to the insurance company, get authorization, gather all the supplies, arrange a nurse, get a pharmacy console. That's kind of like the Reader's Digest version of what happens.

Becky: Great. Thank you. And you kind of alluded to it, sometimes insurance companies mandate who we go to for our home infusions or even in facility infusions. How do we notify our doctor or let them know we would like Nufactor or another company to be our infusion company?

Michelle: So this happens all the time. If the doctor or the patient has a preference, they'll send it to that provider and the provider will verify the coverage and then they'll know if they can take the case or not. There are some companies that will take a patient out of the network and that's fine. But you would want to understand how it's covered and what the patient responsibility of any co-pays or deductibles and things like that would be before you go on service and you would want to know that it was authorized. If somebody were to provide services without proper authorization, IVIg is extremely expensive. It's been a while since I've heard of this happening, but there have been some patients that the provider did not get authorization and the patient got a huge bill. You would want to understand that it's approved and how it would be covered for you. If the provider can't accept the case because of insurance they would advise the physician no, we can't take it. Here are the providers that can, who would you like to use and in the referral would go there.

Becky: Great. Thank you. The next question comes from Robert and he wants to know is IVIg plasma matched to your specific blood type or is it a universal product?

Michelle: That's a good question. So it is a universal product the manufacturing process will make it so that it's usable for anyone. However, there is one side effect called hemolytic

anemia and that has to do with if your blood type is positive and negative and it's in your blood type you could have that reaction. But in general anybody can receive IVIg again, it's all based on tolerability.

Becky: Great. Thank you. After you mentioned you wanted to make sure that it is covered by your insurance. We got a whole bunch of questions that came in, how much does it cost without insurance?

Michelle: It's a lot. Again for pemphigus and pemphigoid it could be upwards of \$10,000 a dose of therapy. So it's not something that you want to pay cash for. Really with this condition it's not something that is typically denied. It's addressed in pretty much all of the payer policies. If for some reason there is a denial we can successfully appeal by showing that although it's not FDA approved for pemphigus or pemphigoid, that in no way means that you can't receive and get insurance approval because it is a standard of care for this condition. We don't really see a lot of issues with getting it covered by insurance.

Becky: Great and is IVIg used worldwide specifically there's a question that came in is it available in Australia?

Michelle: It is. In fact, one of the manufacturers is headquartered there. Yeah, so it is used worldwide. I believe you have people outside the US on your medical Advisory Board.

Becky: Yeah, and we have a find a doctor map on our website as well and that you can click search by ZIP code or postcode or city and state or Province and you can search for doctors. You may have to zoom in and zoom out, but there are international doctors on our list as well.

Michelle: I don't know about home infusion in other countries, I can't speak to that but you can absolutely receive IVIg for this condition in most countries, I would imagine.

Becky: Great. Thank you. Our next question and we've touched on it a little bit. You were talking about monitoring kidneys and cardiac function, but is there any special testing that I need to go through prior to having infusions and deciding if IVIg is the treatment for me?

Michelle: Not typically, but because of the Black Box warning even if a patient has no prior issues with their kidney at Nufactor we do yearly and BUN and creatinine. BUN stands for blood urea nitrogen and creatinine, those are two blood tests that give an indication of kidney function. Even in completely, otherwise healthy individuals we would monitor that annually,

maybe more frequently if there were some concerns about that or it's up to the physician as well. But that's about it. Other than that, there's not really anything that you need to have done prior or while on therapy for pemphigus or pemphigoid.

Becky: Great thank you. So before our next question, I'm going to do another poll of the people who have received received IVIG. If you could just take a quick minute and answer if you've had it at home? It looks like about 80% of people have not had it at home. So about only about 20% of the people. So the next question that came in is what do I need to do to prepare for an infusion in my home?

Michelle: So you'll get all of that education up front. There's a lot of little things. Drinking unless there's a contraindication for you having extra fluids, we really encourage people to drink a lot of water. Hydration just seems to make a world of difference. Some doctors will even order concurrent hydration like intravenously to offset side effects. So that would be one thing. We definitely want you to take your pre-meds and just get proper rest around the infusion. We want the home environment to be conducive to home infusion in general. You want your supplies stored in a clean safe place. Your IVIg should be refrigerated and taken out about an hour before or a couple hours before. It is stable at room temperature for a certain period of time but we like to have it refrigerated. But you don't want to infuse it cold, so take that out. Hydration also helps with the nurse being able to start an IV, so that's important. Having had the education about what to expect so, you know what to look for and to report during the infusion.

Becky: Great. Thank you. So what equipment will I need and will my insurance company or will Nufactor send it to me? Or what do I need to get on my own to be able to be prepared for an infusion?

Michelle: Not really anything. So the way that insurance covers IVIg at home is kind of like in three parts. They cover the drug, they cover the nursing, and they pay something called a per diem. The per diem is like, for every day of infusion you get a small amount of money, maybe \$50 or \$60, it depends on the insurance. You get a small amount of money for every day of infusion and that will pay for the supplies, the alcohol wipes, the tubing, if there's a pump. We like to run it via gravity to simplify things. You will get a collapsible, portable IV pole for that as well. If you wanted a pump so you could walk around that's available too. I know a lot of patients sometimes get concerned with the amount of supplies that they get and they worry that they're going to be charged or their insurance company is going to be charged more if they get a bunch of supplies. That's absolutely not true. You get that per diem if you send 1,000 gloves or you send one pair. So it's really up to the provider to determine what they feel is necessary to safely and efficiently complete an infusion each day. And send that, they don't want to overstock you but they certainly don't want to deprive you of anything that might be needed.

Because you are in the home, you're not in the hospital where the nurse can just run down the hall and get something else that's needed. We like to get that right balance of supplying what you need without over supplying you because you do have to store it and have your house turn into a pharmacy. Does that answer the question?

Becky: Yeah, and I guess as a patient at home who might have a big box lands on my porch. Is it good to keep it in that box? Is it better to put it like in a tackle box or a separate basket that I can put away in a cupboard? Or what do you prefer as the nurse?

Michelle: It's up to you. It's up to what works for you. Again, the IVIg should go right in the fridge and everything else should be kept in a clean and dry, safe place. So, if you want to keep it in the box and put it in a closet somewhere. That's fine. If you want to unpack everything and put it in a plastic bin so you can put it up in your cupboard, that's fine. As long as it is safe, clean, dry, doesn't get mixed in with kids toys and dog food. It depends on the patient and what works for their house.

Becky: Okay, and are there any special precautions that I need to take in light of COVID-19 both to protect me and the nurse coming into my home?

Michelle: So what we're doing at Nufactor is the nurse will call the patient the night before the infusion and there's a list of questions that we'll ask about any potential symptoms that they might be having. If there's any concern about maybe the patient or the family member having certain symptoms that might be indicative of COVID-19, we would want to postpone the infusion. If the nurse assigned to the patient had any of those symptoms themselves, we would want them to notify the agency and Nufactor and we would want to reschedule that or get a new nurse depending on timing and everything how that would work. But our goal is to keep the patient and their household safe and the nurse safe. We are wearing masks. In the home again, it's up to the patient, if they want to wear a mask. We actually provided all of our patients this week Nufactor scarves that we gave them so they can use them during the infusion or outside if they wish. But we're following all of the CDC guidelines in order to make safety in that regard.

Becky: Great. Thank you. And then one thing I think to consider too is that my IVIg nurse isn't going to just be standing there the whole time. We should probably provide you a place to sit or anything like that as well, correct?

Michelle: Correct. It is a working infusion and it is one to one. Whereas, in the hospital outpatient that nurse may have multiple patients. But at home it is one-to-one. So yeah, they would sit down during the infusion because it's several hours. But we are monitoring you the entire time. So we will take baseline vital signs and we take vital signs before every rate increase and then minimally every 30 minutes once we reach the maximum rate. We're always asking, how are you feeling or do you have any symptoms? It's not like, let me just plug in this IV and watch soap operas with you for five hours. It's going to be a working infusion where we're actively engaged and making sure you tolerate the infusion.

Becky: Okay, and then you said I might have 3 days or 5 days of infusions and then is that it until next year? How often does that cycle need to be repeated?

Michelle: So it is definitely a long-term infusion again. It's up to your physician to determine how long. A lot of the patients that we have on it with pemphigus and pemphigoid stay on for a few years. So it's a monthly infusion for a few years, a year or two years. I have a couple patients right now who are into their third year, but they're weaning off. They are stable and have been stable for a while. It is definitely something that is a long-term treatment.

Becky: When you said that Marcia wrote in and said that she's been receiving monthly infusions since 2013. Is this typical?

Michelle: That's a long time. Without knowing the details of her particular case, if she's had flares in between or anything. But that's up to the physician how comfortable they are with this diagnosis. I know a lot of the patients with pemphigus and pemphigoid end up with the specialists in that area and there aren't that many. But seven years is a long time but again without knowing the details of her case, I'm sure there's a reason.

Becky: Great. So our question I think we might have covered a lot of it. So let's talk through what will happen when the nurse arrives at my house. You said that I might get some premedication and they're going to ask me some questions, what else is going to happen?

Michelle: So the nurse, again, will call the night before to confirm the time. They might call before to remind you to take the pre medications which by the way the pre medications are also provided by Nufactor so you don't have to go out and buy Tylenol and Benadryl or whatever the pre medications may be. So remind you to take your pre medications. We're encouraging you to drink a lot of fluids the entire time. The nurse will get there and setup all the supplies to start the IV. We will start the IV then we'll prepare the medication. IVIg is not stable. You can spike the vials so there are vials of IVIg and they're designed to be able to stick the tubing spike right into the vial and infuse from that. Once that vial is spiked. It's good, it needs to be infused. For

whatever reason if a vial is spiked and then it's not infused, it's thrown away. And again going back to the cost that's not something that we want to do. So we always ensure that we have adequate IV access before we set up the medications. We will set up the infusion, start running the infusion, the vital signs, switch out the vials as we go depending on the dose. Once the infusion is done will either flush the line or remove the line depending on, everything is tailored to the patient. The patient can call Nufactor at any time if they're having any side effects or questions or concerns, whether it's from day to day in the infusion or two weeks out. As long as they're on our service if you have a question the pharmacist and the nurse are here for you.

Becky: Great. Thank you. So we've gotten a few questions about this. Can I eat and drink during my infusions? The other question that we've received while you've been talking is, can I also take my regular medications before my infusion or during?

Michelle: So I'm not sure what you mean by regular medications, but if it's something like blood pressure medication, we would absolutely want you to take that. There's really no complications with that. There's certain things but a lot of those are intravenous. But in terms of IVIg and oral medications, we would want you to be compliant with your regular scheduled oral medications. So there's really nothing that would say don't take that until later. As far as eating and drinking again, one of the side effects might be an upset stomach and that could or could not lead to vomiting. So it's again up to how you feel. If you want to eat and if you're hungry then eat. If you're thirsty again, we encourage fluids, non-alcoholic, of course. Again with the headaches, we wouldn't want you to drinking while you're on therapy. It's all according to how you feel as far as food and beverage go during the infusion.

Becky: Okay. Perfect. Thank you. Is it best to sit or to lie, or does it really not matter when I get my infusion?

Michelle: Again it's all about how you feel. So if you want to lay down you should lay down. If you need to get up and walk to the bathroom you absolutely can do that. However you feel and whatever is safe.

Becky: Thank you. And then are there any precautions I need to take after my infusion?

Michelle: Just again looking for the side effects. Not doing anything that might aggravate if you've had a past side effect. So again, the nausea, vomiting, blood pressure changes. It's all the same stuff before, during and after.

Becky: Great, how long after an infusion can I have a reaction?

Michelle: If you're going to have a reaction it's typically during or pretty soon after the infusion but there are delayed reactions that have occurred. So that's why we just like you to be aware of what they are so you can look for them at any given time and report them as they come up. For most people it's going to be pretty quick or pretty soon after the infusion is done, but we want to know about any weird symptoms you might be having at any time.

Becky: Great. Thank you. Does anybody else need to be with me for my infusion whether I have it at home or at an infusion center?

Michelle: We do like there to be somebody with the person who's getting IVIg, especially initially, but no, we do have patients who live alone, who don't want to have somebody with them during the infusion. Again, as long as they're safe and they tolerate it and there's no other factors that might make having somebody there warranted then it's okay for you to be by yourself.

Becky: Great. Thank you and just to clarify we've gotten a couple other questions, the home infusion company will send me my IVIg? This is not something I need to go to a local pharmacy and pick up or anything like that?

Michelle: Everything is shipped. Depending on the supplies and everything it could be a couple of boxes, but they would come together in one shipment. Because a lot of providers service the entire country and they don't have pharmacies on every corner so it will be shipped to you via FedEx or UPS whoever is used. Everything that you need would be in the boxes.

Becky: Great. Thank you. And then if I get my IV infusion today and I get my series, how long is it until I should start seeing some relief of my symptoms for pemphigus or pemphigoid?

Michelle: So typically the response that we look for with pemphigus and pemphigoid. Is that, you're not getting new lesions and that your lesions that you do have would heal. You're going to just want to monitor your skin so that you're not getting any new outbreaks and any lesions that you do have are healing. So it's kind of hard to say. I don't think you're going to notice a big difference after one infusion. But as the infusions go on month after month, you won't see new lesions and you'll start to see the ones that you have start to heal and that would show that you're responding.

Becky: Okay, Pip is saying that the immunologist is refusing to try IVIg after two failed treatments. Is that a normal time when she should be seeing response and should they be looking at a different treatment or giving IVIG another try?

Michelle: I mean just two months of therapy, I would want to know what did or didn't happen? I don't think it's long enough personally. In my experience as a nurse that's not long enough to try IVIg. After only 2 months it's difficult, from what I've seen you're still on a pretty significant dose of steroids. But as the IVIg starts to work, the steroids are able to be weaned off. It's like spacing it out. You would want to see that the steroids can be weaned and you're not getting a new outbreak but I think w months is too short.

Becky: Okay. The last question is, if IVIg requires 5 infusions a month for a year or two, why would a doctor prescribe it instead of Rituxan?

Michelle: It's all about the doctor and what their experience. A lot of the physicians that actually specialize in this condition are researchers who are involved in ongoing research. There's definitely two camps in this arena. There's those that feel like Rituxan can give a very quick remission and it holds fairly long and it's not as many doses, but you can flare again. There's some doctors that might think I would rather have my patient stay on long term IVIg and then never need treatment again. There are some doctors that believe you can achieve permanent remission with IVIg. Then there's some doctors that believe in the combination therapy. So they start off on IVIg or depending on the severity start off on both but the Rituxan is more short-term and the IVIg is long term. It's just really dependent on the physician and how you feel and what you want to advocate for yourself to try a great.

Becky: Great. Our last question has come in, if my insurance denies me getting an IVIg will Nufactor be able to help me through the appeals process.

Michelle: Absolutely. We never just let the patient have to deal with that and we actually also like to help the physician. Really the only time that I've seen that is the insurance will say that's too long. You have been on therapy long enough. But there's papers that show that the mean time of somebody being on therapy I want to say is like 3 to 4 years. So we would write a letter and show that paper and a lot of times it's overturned. I can't really think of a time when it wasn't it. Sometimes there could be a struggle but we would never just leave it up to you even to start therapy. If the initial request for authorization was denied we would look at the reason for denial and write an appeal letter, send it to the physician to edit and sign off on. It would never be put upon the patient to handle that.

Becky: Thank you. That's really helpful. I can't believe how quick this hour has been Michelle. Thank you for being on the call with us today. 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:

Our next Patient Education Webinar will be on May 27th with the IPPF's Peer Health Coaches to answer your questions about living with pemphigus and pemphigoid. This is a great time to have your questions answered from a person who knows exactly what you are dealing with. You can register online today!

As you know, the IPPF's main focus is to improve the quality of life for all those affected by pemphigus and pemphigoid through early diagnosis and support. Day in and day out, we're here for you, whether it's by providing support through our peer health coaches, supporting the research of new treatment options, advocating on behalf of the rare disease community, or accelerating the diagnostic process. What you may not know is that we accomplish all of this with just 4 full-time and 6 part-time employees. Though our commitment is international in scope, the IPPF operates as a small nonprofit organization. Due to the ongoing COVID-19 pandemic, the world is facing unprecedented challenges. Unfortunately, the IPPF is not immune. Recent donations are down, and our usual sources of funding are now unreliable. In short, we need your help to keep the IPPF operating in the way you've come to expect. Together, we can keep hope alive. We need your continued support to reach our goal to raise \$30,000 by June 2. You can go online and donate today at www.pemphigus.org/hope. Principia Biopharma has generously agreed to match 100% of all donations raised until June 2. Give today, and you'll double your impact!

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. 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!

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, 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.

