April 15 2020- Pemphigus and Pemphigoid Side Effects Patient Education Call

Becky: Welcome everyone. This call is now being recorded. I would like to thank you for being on the call this morning. Our speaker today is Dr. Ron Feldman. Thank you for joining us. On the call today we will discuss symptoms of pemphigus and pemphigoid and please know we are planning to have another COVID-19 call so we'll save those questions for that next call. But first let me introduce you to our speaker today. Dr. Ron Feldman earned his MD PhD from the Medical University of South Carolina and completed his Dermatology residency at the Cleveland Clinic. He then completed an autoimmune blistering disease fellowship at the center of blistering diseases. Dr. Feldman is currently an assistant professor in the department of Dermatology at Emory University. He now directs the Emory Autoimmune Blistering Disease Clinic where they strive to better understand and develop novel therapeutics for these devastating diseases. Now, it is my pleasure to introduce Dr. Ron Feldman to answer your questions about Symptoms of Pemphigus and Pemphigoid.

Dr. Feldman: Hi, good morning. I think your signal is breaking up Becky. I'm having a hard time hearing you.

Becky: So have had quite a few questions about what to use on scalp lesions and if there is a best formulation of clobetasol to use on the scalp?

Dr. Feldman: Sure. Good morning everyone. Thank you for joining this webinar. Hope everybody's staying safe. So the questions about that formulation for the scalp. Yes clobetasol is all is a good option for the scalp. It does come in a shampoo formulation. Sometimes the difficulty is getting the shampoo covered by insurance. I would recommend folks go to Good RX to be able to get and shop around for prices for the medications and to get coupons to use. But clobetasol does comes in a shampoo and it comes in a solution or a liquid which can be applied to the scalp. You can also use other options. If the scalp is really crusty and scaly there's other medications called salicylic acid and one of them comes in a shampoo it's called Salex and that can be used to apply to the scalp area. And then after that you can apply the clobetasol on top of that.

Sound breaking up...

Becky: Dr. Feldman if you can hear me do you have a list of the questions that I sent you,

would you like to start answering those while I try to get a better connection?

Dr. Feldman: Sure. No problem. All right, so some questions that Becky sent me from folks one is about itch and pemphigoid. Why do patients it itch and why isn't it as common and patients with pemphigus? So in terms of the itch, we don't exactly know all the reasons why patients itch with pemphigoid. We do know that for example, there's a lot of inflammation in the skin and the inflammation in the skin can aggravate the mechanisms related to itch and some of those pathways are now being elucidated in potentially targeted for therapies. Pemphigus patients can itch actually in some cases as much as pemphigoid patients when pemphigus patients have skin involvement. And we actually just put out a paper on this recently describing this phenomenon. Things to help the itch, there's multiple options and I would encourage you to discuss with your treating physician. For sure over the counter lotions and treatments. Sarna lotions, one common lotion we recommend for itch. It comes in different formulations. The Menthol formulation tends to help a lot with itch. And in terms of the topical steroids tend to help with itch as well that you most likely be prescribed. Other itch mechanisms, we try to target different pathways. One of them is the antihistamine pathway. So some of you may be on antihistamines, some of those such as Zyrtec or Claritin or some of the more older sedating ones we call Benadryl or Atarax. Those can help with some aspects of itch. We also use medications related to blocking the itch pathway in nerves such as Neurontin or Lyrica sometimes we also utilize.

Dr. Feldman: Any non-medicinal ways to help with itching? Good question. I think this also applies to other behavioral techniques to help with itching. This is sort of an area of interest and research we don't know as much but there is some suggestion obviously reduction in stress, behavioral modifications in terms of meditation or yoga believe it or not have been utilized to help patients with itch.

Dr. Feldman: Another question was about cyclosporine. Cyclosporine has been used for three months and they haven’t noticed any improvement. Again, I would speak with your physician. I don't know, it could be the cyclosporine is not the correct dose or the optimal dose to keep. If it's not working for you it may be necessary to switch to another medication.

Dr. Feldman: This patient asked, what foods including herbs and spices aid mucous membrane pemphigoid. This is an interesting topic and I think we can address other questions with this one question. I don't know exactly what herbs and spices would aid mucous membrane pemphigoid oral lesions I suspect they're interested in knowing about. I do know that there are certain foods that can aggravate mucous membrane pemphigoid. Usually we say spicy food or acidic foods can aggravate lesions in the mouth with mucous membrane

pemphigoid or MMP. Also crunchy food can commonly aggravate lesions in the mouth with MMP. Some patients anecdotally mention bananas, onions, and chocolate sometimes can aggravate lesions with MMP.

Becky: Dr. Feldman. Can you hear me? I apologize for that. Does gluten or dairy have anything to do with oral lesions?

Dr. Feldman: We don't think so but again, we don't have much evidence either way. Anecdotally some patients think that an increase in processed foods or high sugar content foods may aggravate some lesions. But as far as gluten outside of the 1 autoimmune blister disease, we call dermatitis herpetiformis or DH where we know gluten plays a strong role with that condition,we don't have much evidence and dairy either. We don't know.

Becky: Great. Kate says, I have been getting lesions on the soles of my feet. What is the best way to care for these?

Dr. Feldman: So lesions on the soles of feet are relatively common, especially in pemphigoid. I don't know the best way to care for these but there's many different options. I think there was a suggestion that you should pop blisters? I typically recommend popping blisters. If you don't pop the blister that pressure from a blister itself tends to make the blister expand so I do recommend popping blisters either in the office and or at home if you just get a sterile needle Or have a needle and heat it up so you can sterilize it. You can use that to pop blisters, gently pop it, don't remove the whole blister just pop it and gently push down with some gauze to remove the fluid and keep the actual top of the blister as natural Band-Aid, so your skin acts as a natural band-aid and then you can cover that if you want to with some gauze or we call non stick telfa dressing, gauze on top of that and wrap it up.

Becky: Great. Thank you. So Timothy asked what causes my skin to develop blisters and he wants to know why pemphigus blisters seem to be more fragile than pemphigoid?

Dr. Feldman: Sure, why what causes the skin to blister? It’s obviously related to the auto antibodies so patients with autoimmune blistering diseases have unfortunately their immune system is generating antibodies, that's a substance that's attacking proteins in the skin. And unfortunately when the skin proteins are attacked, they no longer function normally so the skin becomes fragile and falls apart. With pemphigus the protein that is attacked, it tends to be higher up in the skin which is why these skin blisters tend to be more fragile where as pemphigoid, the antibodies are attacking a protein a little bit lower down at the junction of the

upper layer and lower layer of the skin so that blisters tend to be a little more tense as we call them or a little stronger and do not break as easy.

Becky: Great, should patients be leaving lesions open or should they be covering them? I guess there's a lot of talk in different Facebook groups about what's the best way to have lesions? So what is best for patients?

Dr. Feldman: Right, for the most part we tend to recommend keeping them covered nowadays. It helps improve wound healing and less chances of also getting the wounds dirty or getting them infected for other reasons. So I would recommend you typically wash them gently with whatever is recommended by the treating physician and then apply the topical steroid and you can apply a dressing on top of that until the lesion heals, that is until the skin starts to form again on the surface.

Becky: Yeah, and it seems like covering that would help with lesions sticking to clothing as well. Are there any other recommendations to help stop lesions from sticking to clothing?

Dr. Feldman: That's a great question. I remember a patient whose name is David, he unfortunately passed away a few months ago, he was a pemphigus patient but unrelated to pemphigus. He had a whole regimen of ways he used to do his morning routine before he got dressed in order for the clothes not to stick to him. And what I can do is I can forward to the email Becky you're happy to send it out.

Becky: Great. Thank you.

Dr. Feldman: With some suggestions he had and some websites where he used to get some of the various clothing and materials. So I'll send that to you, it's a good way to start because it is an issue especially if the pemphigus is very new and inflamed on the trunk. It is a big issue with clothes sticking easily.

Becky: Great, that would be very much appreciated. Our next question says can you visually tell the difference between the lesions of activity of pemphigus and pemphigoid? Susan says she can't get a biopsy with everything going on. Is there a way to diagnose lesions using telemedicine?

Dr. Feldman: Very relevant question now as we as I've been seeing patients remotely. I'll say

there's no easy way to diagnose using telemedicine. There are certain clues you can utilize to be able to narrow it down. Obviously if there's oral involvement, more common in pemphigus than bullous pemphigoid. Depending on where the skin lesions are as mentioned earlier if the blisters are really tense and don't break as easily we tend to think more pemphigoid than pemphigus but there can be overlap there. Location of blisters to, and pemphigus tends to affect mostly above the waist in terms of skin involvement and pemphigoid can affect really anywhere but does tend to prefer the lower legs in many patients. So there are certain clues potentially and where the lesions are may be helpful in terms of getting a good clinical history, but it's very difficult without a biopsy to be honest with you. The other option would be, and again and I don't necessarily recommend going to labs yet but if you had to you could also get blood work done. Where you can look as specifically which antibodies are circulating in the blood would help you narrow down the diagnosis too.

Becky: Great information. Thank you. Mark asked, can I put topical steroids like clobetasol on open lesions and are there any steroids that I shouldn't put on open skin?

Dr. Feldman: Yes, you can use clobetasol on open lesions. Any steroids you can use on open lesions.

Becky: Great, can people use clobetasol in their mouth?

Dr. Feldman: Yes, I recommend clobetasol particularly the gel format for the mouth and for the vaginal area too. For the genital areas you can use clobetasol on both. What I would recommend though is just have your treating physician tell the pharmacist it's okay to use in the mouth because typically the pharmacy will not recommend it in the mouth based on what's written on the tube. But many of my patients' clobetasol gel, it's very effective in the mouth.

Becky: Great, and you said it can be used vaginally as well?

Dr. Feldman: Yes it can.

Becky: Perfect, thank you. Beverly says that she's been on prednisone for about three years for a PV. She's usually on 30 mg but now on 25 mg. I know you should get off prednisone, what is the recommended time and amount for decreasing prednisone and how much should it be lowered for each drop in dosage?

Dr. Feldman: That's a very loaded question. A good question, but a loaded question and I

think it's worth discussing with your treating physician. Yes, we have patients who have blistering diseases who are on prednisone for long periods of time. For those patients, yes there are unique issues that should be addressed particularly related to bone loss for example in patients. So if we're going to have patients on prednisone for extended periods of time it's important to make sure we cover all of our bases and at least look into something such as osteoporosis. So you want to have discussions about getting a bone density scan if you are going to be on prednisone for such a long period of time. Again, it's a case-by-case basis whether the patients have issues with blood sugars and diabetes, high blood pressure, hypertension. All of which can be exacerbated by long-term use of their prednisone and of course we tend to prefer obviously to be able to transition off or lower doses of prednisone by using what's called steroid sparing medicines. So again, it depends on the case by case basis and I will discuss in more details about the necessity of being on such a high dose for so long and what are potential other treatment options?

Becky: Great. Thank you. Changing topics here a little bit, Peter asked with summer coming and is being outside okay and are physical sunscreens that contain titanium oxide better for patients or the more sunscreens that are the more chemical kind?

Dr. Feldman: Good questions. I don't want to tell patients to avoid going outside but there needs to be some caution with blistering diseases for the sun does exacerbate these conditions. So patients I would recommend trying to avoid direct sun if you can between 10 a.m and 4 p.m. Especially in the middle of the summer, try to go out earlier in the morning or later in the afternoon. If you do have to go out, physical barriers are most important. So hats, wide brim hats, and sun clothing. There are a lot of manufacturers now that make clothing that have sun protection called UPF and you can just Google that online. There are multiple companies now that make sun protective clothing much more widely accepted now than it used to be. Sunscreens, I agree, the barriers are better than the chemicals. So that would be sunscreens containing titanium dioxide zinc oxide. We tend to prefer to have better protection and more broad protection than chemical sunscreens. So those are the main tricks I would recommend. And of course don't forget the lips, especially with pemphigus patients you want to make sure you get some kind of sunscreen and or chapstick with sunscreen to protect the lips.

Becky: Great. I think we tend to forget about the lips and so that's a great tip as well. Brenda asked what are some things that I can do to boost my immunity or keep me from getting sick while I'm on the medications to treat my disease?

Dr. Feldman: Great question. I think some other folks were also asking similar type questions.

What can you do to boost your immunity? There really is nothing that we know of that can boost your immunity and there is no blood test believe it or not where we can tell you how intact your immune system is. The main thing that we know can help keep your immune system functioning as well as it can, is to reduce levels of stress. We know stress can lower the immune system and play a role with these diseases. Getting a good night's rest, we tend to say around 7 hours seems to be the magic number nowadays that would help keep your immune system healthy. And getting a regular amount of exercise if possible. And of course a well-balanced diet too, lots of fruits and vegetables. All those things we know help keep your immune system as healthy as possible. And in terms of medications, again it's a case-by-case basis. I would have this discussion with your treating physician in terms of if you are on immune suppressing medications, does it need to be modified at least as far as I suspect this is related to the COVID but I think I would have this discussion in terms of should the doses or should the medications be changed and that needs to be a discussion between you and your treating physician. Of course washing hands is very important, social distancing is important, too very helpful as well.

Becky: Great with the COVID-19, everybody is talking about washing their hands for 20 seconds. Is this a good length of time for us all to be washing our hands no matter what's going on or should we be washing our hands longer since we have these autoimmune diseases?

Dr. Feldman: I think 20 seconds is a reasonable number for all patients including patients with blistering diseases.

Becky: Great. Thank you. Gordon says they seem to be having watery eyes and I usually have bad seasonal allergies. When do I go see a doctor and what type of doctor do I see about this?

Dr. Feldman: That's a good question. I guess that I'm inferring that this patient may or may not have eye disease related to blistering conditions in addition to seasonal allergies. I will say patients with what we call ocular pemphigoid or disease pemphigoid that can affect the eye. It's kind of tricky in spring because patients can also have seasonal allergies. The pollen is very bad this year. So sometimes it's hard to tell whether the eyes are really watery and itchy from the pollen or from their underlying pemphigoid. But of course, I would start with the eye doctor. In terms of which eye doctor, the optometrist typically will be the sort of the front lines. They are the ones that are mainly dealing with the fraction of the eyes, so various contact lenses or glasses. The ophthalmologist will deal with more medical related issues. I work with multiple ophthalmologists here at Emory University. And so either one is reasonable to start with. If it's specifically related to the pemphigoid I recommend an ophthalmologist who may

have more experience than an optometrist. That being said sometimes patients have eyelashes and I think there were some questions about this. With pemphigoid eyelashes can be an issue in which the eyelashes can bother. They can rub against the cornea and in that case it's good to have them removed and either optometrist or an ophthalmologist for the most part can help remove those eyelashes.

Becky: Great, speaking about the eyes we also had a question: are contact lenses okay to use when I'm on treatment for my disease or just when I am having specific eye symptoms?

Dr. Feldman: Contacts and again assuming, it depends on how involved the eyes are but for the most part, yes contacts are okay for patients with ocular pemphigoid. We also utilize what's called scleral lenses, their specific contact lenses to help protect the cornea for patients with ocular pemphigoid. In terms of eye drops again I would have this discussion with your either optometrist or ophthalmologist what they recommend in terms of the correct eye drops. Yes, you can also use eye drops to help with the eye symptoms and they range from topical steroid formulations to antihistamine formulations to the saline products are safe to use in the eyes and saline drops or saline ointments are available over-the-counter actually too.

Becky: Great, Charlie says I've been having oral pemphigoid lesions and my dermatologist wants me to see an ENT and gastroenterologist. I'm wondering if I'm also experiencing vaginal lesions too, should I mention this to my dermatologist or are there OBGYN who specialize in patients like me and if so, how do I find them?

Dr. Feldman: I would definitely mention this to your dermatologist for sure. It's most likely that you do have what we call mucous membrane pemphigoid that can involve multiple areas of the body including the mouth as you mentioned, the swallowing tube or the esophagus sometimes can be involved and the vaginal or anal area can be involved. As far as OBGYNs, I would definitely also recommend discussing that with the OBGYN. I will say from my experience not many OBGYNs are as comfortable or have seen many of these patients so they may not be helpful in the situation. You may rely more on the dermatologist. I don't have a specific way to find them perhaps they have IPPF has a way to liaison with various OBGYNs across the country, but I will say it's an area where we need more help and we're looking for OBGYNs to help co-manage many of these patients.

Becky: Great. This is great information. Samantha wants to know should I say an internist or another physician to help me deal with the drug side effects?

Dr. Feldman: Great question. So typically again, it depends on which drugs we're starting here but for the most part many patients will be started on courses of steroids. And as I mentioned earlier many of the potential side effects relate to other conditions such as high blood pressure, diabetes, potentially high cholesterol. So I do tend to partner with an internist to help co-manage many of these other issues because many times it's outside the comfort zone of a dermatologist. So for example, managing blood sugars, I think would be helpful if you have a patient who has diabetes and you need to start prednisone. You could co-manage with the internal medicine doctors to help manage the diabetes as you manage the blistering condition. So yes, I do encourage everyone to get a primary care physician anyways, but it's always helpful to co-manage together to treat the blister and conditions if they're going to need specific drugs that have side effects such as prednisone.

Becky: Great, thank you. Elmer asks how common is it to have oral lesions in pemphigoid?

Dr. Feldman: Again, it depends on which type of pemphigoid. With bullous pemphigoid oral lesions can occur but not as common. Obviously with mucous membrane pemphigoid, very common because the condition involves mucous membranes, which are those areas of the body that are exposed to the outside, so mouth, nose, eyes, genital area. So very common and mucous membrane pemphigoid, not as common and bullous pemphigoid.

Becky: Great and this I think applies to both pemphigus and pemphigoid patients. If I'm having lesions, should I still go to the dentist?

Dr. Felman: Good question. So many patients along their journey right with oral lesions tend to first go to the dentist. Dentists may or may not be able to do biopsies so sometimes the dentist will want to manage aspects of the hygiene related to care of the gums obviously care of the teeth and may not feel comfortable necessarily with conditions that may involve autoimmune diseases at the mouth. So oral lesions for example will be diagnosed at the dentist. But most likely they'll be referred to an oral surgeon, ENT and or dermatologist to be diagnosed. So I think primarily the dermatologist will be treating the oral lesions. The dentist is great though to partner with in terms of once the disease gets under better control the dentist can help with managing oral hygiene and taking care to reduce levels of bacterial plaque which can sometimes aggravate these conditions too. So I typically recommend that if the disease is very active in the mouth have the dermatologist help to reduce the inflammation of the mouth. Once the sores are mostly healed then I do recommend every four to six months to have a gentle dental cleaning to make sure the level of plaque is reduced. And then the dermatologist again will be the primary driver of the lesions in the mouth and this case. Of course topical lesions you can use we talked about earlier Clobetsaol gels is an option, other oral options would be steroid rinse. So one's called dexamethasone rinse is an option too in

terms of a you can switch it out and spit out the medicine that helps also to control if lesions in the mouth, especially if the lesions of the mouth are extensive. So topical options are definitely available for patients.

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Becky: Be recommended to help with the pain.

Dr. Feldman: I only got part of that, but I think it's related to painful blisters in the mouth and throat. To relieve pain, good question. I typically recommend using at least starting with a lot of lidocaine rinse you can use it’s something called viscous lidocaine and it comes as a prescription your dermatologist can give this to you and it's a rinse you can use to help relieve some of the pain or you can use it locally to some areas. Otherwise, it would be traditional pills, oral medications to help with pain relief including Ibuprofen or Tylenol. Certain foods, I think we mentioned earlier in terms of foods that can aggravate would be usually spicy or acidic type foods or crunchy foods can really aggravate. So patients who have really active lesions in the mouth I tend to recommend my normal slogan is boring, mushy and bland foods for the next few weeks until this kind of settles down. So you really have to be careful in terms of keeping the food very soft. So that doesn't aggravate the lesions in the mouth.

Dr. Feldman: Great. Thank you. Maxine says I get cold easily and my temperature at those times is under 96 degrees. Warm tea and more layers help little and warm showers help a little as well. This predated my rituximab and my doctors don't seem concerned since all of my labs are within normal limits. Could this be a symptom of PV or a side effect from the mycophenolate?

Dr. Feldman: I don’t know. That's what that's a difficult question without knowing more of the context but I would suggest that if you always feeling kind of cold or always too hot one of the lab tests you can do is a thyroid test because a low thyroid or hypothyroidism can go along with pemphigus. But otherwise, I don't think it's related as far as I know as a very common symptom of pemphigus or Cellcept.

Becky: Great. Thank you. Bob is newly diagnosed and has noticed that he doesn't have many blisters and some seem to come and go does this mean that he doesn't really need to be on any medication?

Dr. Feldman: That's a good question. So in other words we would call this sort of mild disease

is what I interpret this anyways. So not necessarily we tend to leave if it's a mild disease and it's not too bothersome to patients you don't have to. One thing I would recommend though if possible would be to get some blood work to see if you can pick up the level of antibody in the blood. And if that also is very low that to me anyway is encouraging that it's mild disease there's very low levels circulating. So you don't necessarily have to do anything other than you could recommend a topical medicine and that would be all. But it would be important to follow up though. I would recommend every few months or at least stay in contact with the dermatologist in case things change. Sometimes the disease can get more active unexpectedly.

Becky: Great. Thank you. Beth asks should one taking losartan and several other medications for high blood pressure try to change? I see a lot of conflicting information regarding blood pressure medicines and pemphigoid.

Dr. Feldman: It's a good question in terms of how medications can potentially play a role in triggering these conditions. There is some evidence at least published and previously about some blood pressure medicines, particularly in what's called the ACE inhibitors that can be related to pemphigus, causing pemphigus potentially. Some of the other medications related to causing pemphigoid are more related to the diuretics we call Lasix or furosemide has been shown in many patients to potentially trigger pemphigoid. Losartan, not necessarily as common reported as causing pemphigoid but I would say in all cases a good clinical history. If it seems like the patient started a new medication whether or not related to high blood pressure and developed pemphigoid shortly thereafter it may be worth doing some detective work to see if it may be related. I find in most instances it's probably just coincident, but I think the timing is relatively consistent, you may want to discuss switching off or stopping to see if it does result in improvement in the skin condition.

Becky: Okay, great. Ali says he was diagnosed with pemphigus over 10 years ago and was given Rituxan but hasn't been on any treatments for 4 years. However in the last few weeks it started to get some red spots and pain in his mouth. What should be done for these sores right now, especially with the limited access to being able to see a doctor.

Dr Feldman: That's a good question, what to do? Yeah suspect the easiest thing to do would be to use go with the topical medications as we mentioned earlier to get rid of some of the spots and to help with the pain. That would be the Clobetasol gel, Dexamethasone rinse for example, and or lidocaine rinse. And be careful with the food, so avoid crunchy, acidic foods which may aggravate those mouth lesions. And I think it's important to be in contact though, if

possible, at the treating dermatologists to discuss whether or not anything else needs to be done. Again it depends on a case-by-case basis and whether or not this is truly what we call a relapse or a flare or the disease is trying to come back. I don't know from this brief history here, but I would say it's worth a reach out to your dermatologist. Most dermatologists nowadays are able to access by patient portals or by what we call teledermatology. So hopefully this patient will have access to his doctor at least remotely.

Becky: Great, we've gotten a few questions about breakouts of pemphigus vulgaris related to stress. Is there a relation between stress and autoimmune disease and do you have any recommendations on how to handle the stress right now?

Dr. Feldman: There is a fair amount of evidence that stress plays a role with autoimmune disease including pemphigus and pemphigoid. Again, we're in time now with this COVID crisis where everybody is under high stress. So realistically how do we reduce that? It's not so easy. But as I mentioned earlier a good night's rest if possible, regular exercise and a well-balanced diet. These things can help with stress. Other behavioral techniques there are some evidence actually that meditation. We're lucky here at Emory, we have a Tibetan monk population that studies here at University and they give classes on stress relief including utilizing meditation. We had a previous resident who did some work showing that itchy patients who underwent the meditation course had reduction and levels of stress and itch. So the exact mechanism is not quite clear how stress does play a role or potentially induces these conditions or aggravates autoimmunity but it does and anyways, you can to mitigate the stress if possible can help your condition as well.

Becky: Great. Thank you. Nancy says that she's scheduled for a gum graft on May 27th, and she's unsure if she should have it. Do you have any experience or know how people with MMP react to gum grafts?

Dr. Feldman: That's a tricky question to answer again sort of a case-by-case basis, but in terms of in other words surgical procedures and patients with mucus membrane pemphigoid what my opinion is that if the disease is under good control then I'm typically okay with patients undergoing surgical procedures including grafting if it needs to be done. Obviously if the procedures don't need to be done or they are more cosmetic then I tend to prefer patients not to undergo them just in case it may aggravate things. But assuming this patient is under good control I don't think it's not unreasonable to go ahead with the procedure but it's always good to reach out and get in touch with the dermatologist and they can discuss with whoever's doing the procedure whether it’s the periodontist and can discuss the condition the comfort level of the periodontist, have they treated pemphigoid patients before. Whether or not the patient can undergo a quick burst of steroids around the time of the procedure just to potentially head off flaring of the condition or if something else needs to be adjusted. So as many different options

I think the main thing is communication. Just to make sure everybody is on the same page and to address the condition very carefully and the various treatment options.

Becky: Great, that's great information. Thank you. Sharon says, I've noticed that my skin around my fingernails is really red and my fingernails have lines in them and split easily, is that a symptom of the disease?

Dr. Feldman: Well I'm not sure which disease but yes fingers and fingernails can be involved with pemphigus and or pemphigoid. You can get inflammation around where the nail is generated and resulting in a defective nail coming out or the nails sometimes falling off. So it's not uncommon. It can be a symptom of blistering diseases. Obviously just like the skin outside of the nail region we tend to treat the underlying condition. Once the underlying conditions are treated the nails tend to get better and come back normal. For the short-term treatment options you can use topical steroids in those areas. It tends to help, you can also do Epsom salt soaks to help some of the soreness and then put on the topical steroid as well.

Becky: Great, Linda asked, can you just stop taking Methotrexate or do you need to wean off of it?

Dr. Feldman: Again, that is something that has to be discussed with the treating dermatologist in terms of the particular style or comfort level of the dermatologist in terms of utilization and these medications but you can. All the immunosuppressant medications outside of prednisone, you can stop. The question is what is the purpose of stopping in this case? I'm not so sure or we tend to want to wean Methotrexate more for disease issues and potential side effects. In other words, we want to see what the lowest level of Methotrexate we can go down to which would keep the disease quiet and allow us to avoid obviously using steroids. So it really depends on the situation. But yes, technically yes, you can stop Methotrexate cold turkey or any other immunosuppressive drugs. Prednisone is the one that you don't want to do that necessarily from a high dose to stopping very quickly because of the potential issues with blood pressure. But I would definitely suggest talking to your local dermatologist who is working with you.

Becky: Great. Thank you. Henry asked, what can I do about the pain? Sometimes it's uncomfortable to even sit or lie down when I have blisters on my back.

Dr. Feldman: That's a good question. Again, I'm going to send Becky here the recommendations. A lot of the pain comes from obviously the rubbing, so the clothes rubbing against the skin. So something's to try to at least to have clothes not to be so bothersome, I

think will help with that. Again popping the blisters and getting the fluid out will help too because the pressure of the fluid causes a lot of pain. Using topical lidocaine is an option too. Your dermatologist can give you topical lidocaine or Lidocaine patches and in some cases those can help. It's tough to say I think I don't have one standard answer to help with all the pain management. I think the dermatologists working with you will have to discuss that. In some cases it's really severe, we sometimes get our colleagues in pain management to help us, too.

Becky: Great. Thank you. Iva who is a PV patient asks, she says that she has received Rituximab infusion and should she start Prolia treatment? She said she had a bone density scan and the diagnosis was osteoporosis induced by steroids in the spine and the endocrinologist recommended Prolia. She wants and says that she can get the infusion in the parking lot, but she's heard about some of the side effects and she just wondered if it was recommended?

Dr. Feldman: Infusion the parking lot, huh? Yeah, I don't as far as I know there's no contraindication from getting Prolia in addition to Rituximab, but I would have a discussion with the dermatologist and endocrinologist.

Becky: Okay. We've also gotten quite a few questions about pregnancy with pemphigus and pemphigoid. A specific question is that I have PV and I am currently in remission is it safe to consider getting pregnant? What if I go into a flare when I am pregnant?

Dr. Feldman: So if you've had pemphigus previously there's no guarantee that the disease will come back during pregnancy. I don't think it's a reason to consider not getting pregnant. Obviously if it does occur while you're pregnant, there's many treatment options. So again a case-by-case basis here but I think it's safe to get pregnant from my standpoint. We deal with it and if something happens we'll deal with it as we normally treat any pemphigus patient.

Becky: Great, that's great information. Thank you. Tom asks, what are your thoughts about using infrared light therapy for PV? He's read that red light treatment apparently has some evidence based in healing wounds and ulcers?

Dr. Feldman: I don't have experience with using blue light or red light for pemphigus. I can't answer that question. I don't know.

Becky: Okay, do you have any advice for bathing or showering so I don't worsen my skin

condition?

Dr. Feldman: When it's really active I counsel patients to be very careful with showers. A full-blown shower can cause a lot of trauma to the face and upper trunk. It can exacerbate lesions. So I would recommend that a gentle shower and or a bath is fine for patients with active disease.

Becky: Great. Thank you. Our next question says every morning when I blow my nose there is blood and a hard scab. The more I blow the more blood comes out until it turns clear. What do you suggest to help me with nasal blisters?

Dr. Feldmn: Good question, that's a very common problem actually. The simple suggestions would be, one, you always get a referral to the ear, nose, and throat doctor or the ENT, to give you some helpful suggestions. I usually recommend starting with simple options such as Vaseline or some kind of steroid ointment. For example Clobetasol, you can use with a Q-tip to gently apply to the areas that are sore in the nose. For other times you can use saline rinses as well. You can get those over-the-counter actually. You can use saline rinses to flush out the areas of the nose, so it kind of dries the area up. In some cases the ENT doctors we've utilized patients who have nebulizers can have a little bit of the steroid drops you apply into the nebulizer as we can actually use that in the steroid inside the nasal rinses. You can mix a few drops in there and that helps also to dry the inflammation in the nose. So there's many different options. Again, it's worth a discussion with a dermatologist and the ENT doctors are helpful too.

Becky: Great. We've gotten some questions in during the call about how I prevent my open sores from being infected and what kind of treatment do you usually use when lesions do become infected?

Dr. Feldman: That's a good question and this kind of goes back to the question earlier about should we keep things covered? So the best way obviously is to keep them clean and covered. So for example, if this is a sore from pemphigus or pemphigoid you want to do your normal routine, which would be to clean with soap and water, apply your topical steroid or your topical agent that's been recommended and then cover it. And that's your best way to keep this thing from being infected.So in terms of needing systemic and or antibiotic pills, it really depends on a case-by-case basis whether or not something else needs to be added. But in terms of preventing infection the best thing to do is to keep the skin quiet and covered.

Becky: Great. Thank you. Our next question says if I'm taking doxycycline does it also help

me from getting sick and getting my usual sinus infections that come with seasonal allergies? And if I get a sinus infection or get sick, can I still take another antibiotic?

Dr. Feldman: Yes, you can take another antibiotic. Again I would discuss that with your primary care physician or with your dermatologist. Helping getting sick, I don't know if that's going to be the case. Anecdotally some patients may say, I'm on this medication and it seems to be that my allergies are less but in general the evidence suggests that it probably will not have an effect on that at all. As you know, again, it's a different kind of process from seasonal allergies versus the antibiotic is reducing the inflammation from the pemphigoid or pemphigus.

Becky: Great, thank you. After getting IVIG I feel bloated and get headaches and feel like I'm just completely run down afterward. Is this due to the IVIG or the premedication that I'm given?

Dr. Feldman: Yes, it is a very common symptom after IVIG, headaches feeling kind of run down and tired, fatigued very common symptoms. It's the IVIG as far as they know not the pre medications. Ways to prevent that again, it's sort of a case-by-case basis. Sometimes it's very difficult. The headaches are very frustrating. So I usually recommend patients take Tylenol around the clock before their infusions and the day of infusions, sometimes even the day after. You can give some saline, extra saline upfront for pre medications through the IV line. Sometimes I can help and sometimes it doesn't but sometimes at least it's worth a shot. Other than that I don't know any other way to prevent fatigue unfortunately. It is a very common symptom afterwards.

Becky: That's great information, thank you. Our next question and maybe we should have moved this one up a little bit but they're asking what does it mean to have a flare and that I've also heard the term transient lesion. What's the difference? And when should I contact my doctor if I notice I'm having lesions?

Dr. Feldman: That's a very good question. So flare means that the disease is getting more active than it used to be. There's different ways to define that from our standpoint. And the main thing from us is going to be okay, what changed compared to previous and then is it transient or is it persistent? So in other words you may get a new lesion, but it may heal within a day or two, so that we would call it transient. Versus a true flare or relapse is that the disease is starting to come back the way it was when you were before your own therapy. So the disease is coming back and new blisters, new sores and they're taking weeks to heal, so that would be what we call a flare. It's reasonable any time to contact your doctor but usually tell patients, if you're getting new blisters don't panic if they're healing within one or two days. We don't necessarily change anything but if the disease is starting to come back and you get a lot more and they're taking weeks to heal, I think it’s worth a discussion that we may need to

change something, the disease is trying to come back worse.

Becky: Great. Our next question says that I am having a lot of hair loss. Is this related to my treatment, my disease or something else? Also, I'm thinking about getting scalp injections, do you have any thoughts on or experience on this?

Dr. Feldman: That's a tricky question because there's many things involved here. With hair loss, I don't know, I assume the patient may have pemphigus. Pemphigus you can't get hair loss if you have a lot of sores on the scalp and also MMP sometimes. Once treated though the hair does tend to grow back normally. Yes, hair loss can also be related to treatments. Some of the medications we give you can result in hair loss as well and it also tends to be reversible once you come down on dose or stop the medication. Scalp injections I assume the patient means scalp injections of steroids, we call intralesional Kenalog. Yes, we do that a lot. It can be very effective to get quicker control of scalp lesions.

Becky: Great, thank you. So I have mucous membrane pemphigoid and I've had two Rituxan treatments and will get a few lesions here and there from time to time. Is this something I should let my dermatologist know about?

Dr. Feldman: Yeah, this is in line with one of the previous questions in terms of the transient lesions. I think it's always okay to reach out to, like I said your dermatologist. I think you could get lesions from here and there and they're healing within one or two days or just a few days, I wouldn't get too worried. But like I said, if you are getting new lesions that are persisting then I think it's worth a discussion, maybe something needs to be changed. But in this patient, it sounds like it may just be transient lesions that are healing quickly. So I wouldn't necessarily do any different but I would definitely reach out to the dermatologist either way if you're not sure.

Becky: Our next question says my mom has pemphigus vulgaris and needs to have open heart surgery. How many days before surgery should she go off medication and how long can she be off medication while recovering from surgery?

Dr. Feldman: It's a difficult question to answer. I don't know the scenario here and I don't know what medication the patient's on. Again, it's the same issue as I always try to harp on, really it's about communication. Just discussion between the dermatologist and the surgeon. Again, a lot of times it just depends on the comfort level of the surgeon in terms of utilizing medication. Sometimes the surgeon does not recommend or does not have an opinion about

whether or not a patient needs to come off medication and they'll leave it up to the dermatologist. In terms if the patient is on and again also depends on the level of the severity of the pemphigus. If the patients have a real severe pemphigus a lot of surgeons won't feel so comfortable necessarily even doing the surgery. So again, it's a case-by-case basis. If the patient's very severe then it's worth a discussion, the patient may need to go on high-dose steroids and or weaned off the other medication for the short term to get better control. If the patient has been in remission, then nothing necessarily needs to be changed. So again, it's a case-by-case basis. I would have the discussion between the dermatologist and ask the dermatologist to please discuss with the surgical team the plan.

Sound breaking up...

Becky: Can you hear me now? Is this better? I apologize for all the technical issues today. Thank you for being with us on the calls. It was extremely educational having you on our call. I would also like to give a huge thank you to everyone on the call for joining us today and thank you to Genentech and Principia Biopharma for helping to make today's call possible. Please join us next Thursday, April 23rd at 4pm Pacific/ 7pm Easter for our next Patient Education Call. Members from our medical advisory committee will answer your questions about coronavirus. Please submit your questions prior to the call. We need your help to continue to spread Awareness about pemphigus and pemphigoid. The IPPF's Awareness Program aims to accelerate the time it takes pemphigus and pemphigoid patients to get diagnosed by stressing the importance of a biopsy. Your tax-deductible donation will support our Biopsies Save Lives campaign that will educate and encourage dental professionals to consider a biopsy sooner in order to diagnose patients faster. 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.

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