April 30, 2020 Oral Health Patient Education Call

Becky: Welcome everyone. Our 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 and Principia Biopharma for making today's call possible. The topic is oral health with pemphigus and pemphigoid. First let me introduce you to our speakers today, Dr. Tanya Gibson and Dr. Nasser Said-Al-Naief. Dr. Tanya Gibson is an Associate Professor at the University of Missouri Kansas School of Dentistry. She graduated from Meharry Medical College of Dentistry in 2001 and completed her residency in oral and maxillofacial pathology at Long Island Jewish Medical Center in 2004. She is a fellow of the American Academy of Oral and Maxillofacial Pathology and a diplomat of the American Board of Oral and Maxillofacial Pathology.. Dr. Nasser Said-Al-Naief is currently a Professor of Oral and Maxillofacial Pathology laboratory at the Department of Integrated Biomedical and Diagnostic Sciences at Oregon Health and Science University School of Dentistry and School of Medicine. Most recently he has served as Professor and Chair, Department of Pathology and Radiology at OHSU and as a director of the maxillofacial diagnostic pathology laboratory. Both Dr. Gibson and Dr. Said-Al-Naief are members of the IPPF's Medical Advisory Council. Now it is in my pleasure to introduce you to Dr. Tanya Gibson and Dr. Nasser Said-Al-Naief to answer your questions about oral health with pemphigus and pemphigoid. Welcome.

Dr. Gibson: Thank you very much for having us.

Dr. Said-Al-Naief: Thank you, appreciate you.

Becky: So I just want to start for those of you who are on your computer. If you wouldn't mind pressing the raise your hand icon if you had an oral pathologist involved in your care. As we're waiting for people to raise their hands, perhaps. Dr. Gibson, you'd like to start by just explaining what an oral pathologist is for our listeners.

Dr. Gibson: Sure, an oral pathologist. Well first oral pathology is one of the specialties of dentistry and as the oral pathologist we are tasked with basically diagnosing and managing disease processes of the head and neck area. Most often, the layperson would associate us with diagnosing oral cancer and we do that but we also do a lot of other things as far as our practice is concerned. We diagnose and manage people with

chronic mucocutaneous disorders such as pemphigus and pemphigoid as well as other conditions. As far as during diagnosis in the laboratory, we look at the specimens under a microscope for a diagnosis, and if necessary will perform additional testing. For our clinical practice we see and manage patients that have a variety of oral diseases.

Becky: Great. It looks like there were only a few people here who had an oral pathologist as part of their health care team. So that's really great. Do you often consult with other specialists Dr. Nasser to care for pemphigus and pemphigoid patients? And how do you normally do that?

Dr. Said-Al-Naief: Yes, ma'am. That's a great question. Actually, I managed the clinic just geared toward the bullous diseases including pemphigus and pemphigoid, a big portion of it and I worked within a team that included dermatologists, autoimmune specialists as well as physiologists as well believe it or not. We were a very integrated team that each did have an integral part in the management of the patients and the only way to really effectively manage these situations is to have an involvement of the team rather than one person.

Becky: Great, absolutely. So we're going to jump right into some really good questions submitted by our patients. And one of them is a common question that we received prior to our call and this is do you recommend for patients with either membranous pemphigoid or oral pemphigus vulgaris to have our teeth cleaned and what are some things that can be done to reduce pain when we do have lesions and need our teeth cleaned?

Dr. Gibson: Okay, so it's really hard to separate your dental health from your oral health in regards to your pemphigus and pemphigoid. So you can't sacrifice one in order to sort of preserve the other so it's very important that you still do your routine dental care. It may have been augmented slightly so that it’s not as uncomfortable for the patient to have their dental work or dental hygiene work performed.

Becky: Great. Dr. Nasserm Rhonda asks would you recommend that patients on immunosuppressive medications have a route of antibiotics before undergoing any

dental cleaning? And how often should I have my teeth cleaned? And what is the right provider to do this?

Dr. Said-Al-Naief: There is a plethora of evidence in the American Medical Association American Dental Association speaks for the lack of any correlation between premedication and the absence of septicemia, i.e., it is an extra step and it's not needed. Now having said that a lot of clinicians still do premedicate immunosuppressed patients and the dental office pre prophylaxis cleaning, injections etc. But I assure you there is very little evidence supporting the validity and help of pre medicating dental patients and in those types of settings.

Becky: Yeah, and I would imagine with antibiotic resistance on the rise, it would contribute to that as well. Correct?

Dr. Said-Al-Naief: Correct, antibiotics in immunosuppressed patients can have significant downsides instead of helping they can really create a lot of unusual changes in the system that may be negatively affecting the outcomes rather than positivity of outcomes. Now we have to respect different opinions of course and they're still clinicians that feel that patients when they come in and they are immune suppressed they better premedicate the patient ahead of time. I'll give you an example. I managed a lot of Lupus patients, all manifestations. Lupus patients tend to have a heart valve lesion that's similar to the bacterial endocarditis that people premedicate for and even that heart valve which is ultimately related, the literature showed no merit or no help from pre-medicating the patients and in a dental setting. So lupus is the best example of autoimmune disease, you know, similar to pemphigus and pemphigoid with the detrimental aspects. But to make it short there is really no merit or no evidence of help.

Becky: Great. Thank you. Dr. Tanya, Connie asks, am I using the appropriate medication for PV in my mouth? Currently I'm taking a triamcinolone acetonide, dental paste USP 0.1% and clobetasol, gabapentin and lidocaine (mucolox)? Is this the right thing to be using?

Dr. Gibson: My question would be whether or not you're getting any relief from using those medications. I'm not familiar with the gabapentin lidocaine, the Mucolox. I've never used that before or the dental paste, I'm not sure what's in that one. I do like

Clobetasol I use that often. Triamcinolone, that's not one of my favorites to use. I like to use fluocinonide. I think fluocinonide and Clobetasol out there are stronger steroids and patients tend to get more benefit from those in my experience.

Becky: Great, along the same lines another patient says that I'm using the triamcinolone orumucosal paste and also Tacrolimus oral gel but the blisters keep returning after she does get some relief. The blisters seem to form first the gums and then they form on the tongue. Is this a normal progression?

Dr. Gibson: It doesn't surprise me that you have a spread of your oral lesions. That's not surprising to me. What was the beginning about tacrolimus you asked?

Becky: Yeah, they take the triamcinolone oral mucosal paste and then Tacrolimus oral gel 0.1%

Dr. Gibson: Yeah, I've used Tacrolimus before and I've used those in patients and I didn't get a good response from using either fluocinonide or Clobetasol and those patients tend to respond to Tacrolimus.

Becky: Okay. Dr. Nasser, why do some patients only get blisters in their mouth and some on their bodies and some of us are lucky enough to get both?

Dr. Said-Al-Naief: This is a great question. It's the nature of the disease. Pemphigus lesions come first in the mouth and they are last to go from the mouth. And the mouth is a gradual step in the start of the skin lesions. The skin is tougher than the oral cavity. Skin has a very thick keratin layer. It has more cohesive cells. The oral cavity has a more fragile tissue. So really, in essence everything is attacked for example in pemphigus vulgaris, but the oral tissue is more friable or more prone to have these ulcers because it's a weaker type of tissue and this is why it probably starts there. Pemphigoid is a different story. Pemphigoid has a whole different, other additional thing that makes it limited to the mouth predominantly, the cicatricial pemphigoid. But it is a problem primarily due to the fragility or the weak epithelium compared to the strong tough cutaneous epidermis.

Becky: Great. Thank you. Our next question comes from Aster and Dr. Tanya, maybe you want to answer this one. Aster says that she started to get lesions that used to be on her body then she started getting some lesions on her gums and in her mouth. Her medication was reduced to 2.5 milligrams of Prednisone every other day about two months ago. And the doctor has recently bumped up her dose to about 15 milligrams every other day. Are there foods that could have triggered this flare or is this just the nature of the disease that it comes and goes?

Dr. Gibson: It's just the nature of the disease that it comes and goes.

Becky: Great. Thank you. Dr. Nasser, Sally says that she's had periodontal disease for 50 years, long before she was diagnosed with MMP. If I'm not able to keep my scheduled hygienist appointment in May. What can I do to keep my mouth healthy?

Dr. Said-Al-Naief: This is a very very good question. And honestly the oral hygiene has a significant effect on how good we can control the vesicular bullous diseases, pemphigus and pemphigoid and keeping the plaque and calculus to the minimum level has a great effect in keeping the eruptions sequences as well as keeping the control of the disease. I would try to go to the dentist. I used to schedule my patients three times a year for prophylactic fine scaling not with instruments, just with a scalar very gentle. I would stay away from mouthwashes that have alcohol. Try to use chlorhexidine, not the alcohol one, the formula that comes from Canada, which is alcohol-free that keeps the bacteria and plaque the minimum compared to the alcohol one, which will sting. This one doesn't sting but you have a diligent brushing, soft foods with the keeping in mind that you should not have sugary foods that will lead to caries. Once the tissue is stable then you can have better more aggressive treatment with the periodontitis trying to do for example, grafting, soft tissue grafts to make sure that you keep your teeth in good health. But the short of that answers which I took longer than I should I'm sorry is to keep things in check and try to visit the dentist in the times where you have better control, then they can go more aggressive in the treatment even though they have to be also considerate to make sure that they do not slough the soft tissue tissue, which is really fragile. Does that answer the question?

Becky: Yeah. I think so. Sally's follow-up question is that her dermatologist says that she can use a dexamethasone rinse when she has a flare but is it possible or okay to use as preventative measures?

Dr. Said-Al-Naief: It is good actually, but remember when we have steroids in our mouths we're going to shift the flora i.e. we are going to kill good bacteria that keeps our mouth in check. If you shift the balance of the microorganisms in the mouth, you'll lead to a shift opposite way a fungus will grow further. Fungus, we don't have fungus in our mouth. We have the candida in our normal flora as a matter of fact candida is part of the plaque and calculus that accumulates on the teeth. But they don't cause disease and until you are immunosuppressed or the immunity lowered. Remember, almost all if not all of our patients are on immunosuppressant medications. So if you keep rinsing with the steroids rinse, you'll reduce the immunity and you shift the flora toward fungus growing and you'll have a higher incidence of candidiasis or fungus infection in the mouth. Also steroids will thin the mucosa, that's known so you have easier time to ulcerate that mucosa with foods and harsh foods for example potato chips or something similar. Steroids of course, even if they're rinsed and expectorate meaning you rinse and spit, you also have some absorption and when you have absorption you will have additional side effects. Long-term, diabetes, suppression of the adrenal normal corticosteroids of the patients, etc. It's not recommended to keep it. If you have a flare up use it, if you don't have a flare up hold it on the side.

Becky: Okay. I'm going to ask Dr. Nasser a question that came in during the call through our text box. This is from Teresa. And she says that she has had oral thrush for two months ever since she began prednisone to treat her PV. While her pemphigus symptoms have largely cleared up after two Rituxan treatments, she just can't get rid of the thrush. She's currently on 10 milligrams of Prednisone daily and hopes to decrease to 5 milligrams soon. Do you have any suggestions to help the thrush in her mouth?

Dr. Said-Al-Naief: Yes. There are three ways of doing this. Number one. She should eat yogurt with culture. Yogurt with culture balances is the flora to the maximum so she will have less incidents of eruptions of the candida. Number 2, there are few medications we use but they're better, one is Nystatin and Clotrimazole Troche. They are better and they have sugar to make them taste good so that the patient can tolerate them. And again sugar with fungus, fungus is gonna live on sugar so it's the kind of double sword. What I like to give the patient's is believe it or not and in males it

is difficult to convince, but in females it is easier to convince. I use vaginal suppositories and I make the patient dissolve them in their mouth and these are the least expensive the best because they coat the whole mouth and believe it or not because they coat the whole mouth and they're direct contact they are very effective in controlling the fungus eruptions.

Becky: Wow, thank you. We're going to just switch topics here a little bit. And Dr. Tanya, is there any information regarding implants in PV patients? Odette says she's interested in getting implants but wants to know if PV patients are an okay candidate to have them?

Dr. Gibson: In general. Yes. It's okay to have implants where. I would caution you at is if you have a flare up, I would wait until your remission before you undergo the surgical procedure because we want to ensure that not only we get healed within the bone but within the soft tissue above the bone and if you have a outbreak that's going to impair your healing of the soft tissues.

Becky: Great. Thank you. Augustine is asking what is the prognosis for a PV patient who relapses with lesions even though his antibody levels one in three are within normal ranges? Dr. Nasser if you'd like to answer you can.

Dr. Said-Al-Naief: Sure. Relapsing pemphigus there is no I mean as far as the only downside is and relapsing is the sloughing and potential bacteremia and patients can have a kind of introduction into the bacteria in today's system. And of course the pain and the bleeding and the tissue fragility etc. In a patient who is relapsing and not controlled. I was in the process of leaving my institution and the previous institution using Rituxan with the help of the rheumatologist. Rituximab has anti cd20 it has been proven to be a great controller of pemphigus. Actually patients have ulcers free, vesicles free for months at a time as long as they stay on the medication. And the reason we like to work with physicians, we had them part of our team is because Rituxan does have some side effects of the heart and other organs. So it's not immense but it's a great medication to think.But in the latter of five six years it's been almost like an put part of the regiment of the treatment of pemphigus. So I would explore with the young lady's physician to see if he would be willing to try her through her doctor, Rituximab. I think she will benefit from it.

Becky: Great. Thank you. Dr. Tanya, you kind of alluded to this earlier. But what topical treatments have you found most effective for patients with pemphigus and then I'm going to follow that with and also for pemphigoid?

Dr. Gibson: Actually for both of them, I've used Clobetasol, Fluocinonide, the topical gel formulations for those and I've also used a mouthwash Prednisolone and received good results.

Becky: Great. Thank you. Dr. Tanya if you don't mind answering our next question as well. Robin says that my gums are irritated and tender below the gum line. The skin doesn't shear off but they are spongy and she's concerned about her oral health. Do you have any suggestions to help her?

Dr. Gibson: Is she talking about the control of her gum lesions oral hygiene regimen as far as keeping her teeth clean?

Becky: Probably both.

Dr. Gibson: So sometimes patients have persistent gingival lesions. I may introduce a medication stent and with the medication stent we can get a closer proximity of the medication to the affected tissues and it kind of stays in place as opposed to manually putting it on there. It gives you just a little bit more contact with the medication. When trying to figure out a dental hygiene regiment, you want to use a very soft toothbrush and sometimes even the soft toothbrushes are even too hard for a patient to use so to soften up even more I tell my patients to run on under hot water and that will soften up the bristles a little bit more for them and then to brush their teeth in a circular motion at a 45 degree angle. And if you use any type of mouthwash, make sure you use something that is alcohol-free.

Becky: Great, are there any kind of toothbrushes that you usually have patients use or toothpaste?

Dr. Gibson: As far as toothpaste, I recommend something that's fluorinated and it doesn't have a lot of bells and whistles. So nothing that's tartar control. Nothing that has whitening. Anything that's extra besides fluoride in the toothpaste I don't

recommend patients use those things. And I even recommended kids toothpaste products. If you can stand the berry flavor, they tend not to be as caustic as adult toothpaste. They thing that works with cleaning your teeth is having that toothpaste as a sort of like an extra soap just so and doing the circular motion at the 45 degree angle.

Becky: Perfect. So I myself used the children's toothpaste and I thought that was a big move for me. And I liked it because it wasn't mint flavored. Mint with my oral lesions created a lot of burning. So I'm sure they'll be...

Dr. Said-Al-Naief: The mint burns.

Becky: Yeah. Absolutely. Where can patients buy the toothbrushes, like the very soft toothbrushes? Are they sold online or can they get them at their local pharmacy or drugstore or do they come from the dentist?

Dr. Said-Al-Naief:I think they're available everywhere. They can get the softest toothbrushes anywhere like Walgreen etcetera, but I also used to use sponges. It's a stick with a sponge on the end and it's available over the internet and some companies carry these on the internet, they sell them in bulk. You can brush the surfaces of the teeth with a very soft sponge and some people soak them in hydrogen peroxide and then clean the surfaces some. Some will use it with water, some will use it with the children's toothpaste. And again, I love children's toothpaste. I believe in it 100%. I tell all of my pemphigus and pemphigoid patients to use children's toothpaste, no exception. So I think patients may find this useful.

Becky: Great. Thank you. Nancy says that she's had PV for 2 years inside of her mouth and nose and she tends to develop a thick film on the basis of some of her teeth that doesn't come off during brushing. What is this and what can be done to avoid thick film for me?

Dr. Said-Al-Naief: The thick film is actually a mixture of edema with fluid coming from the areas where they ulcerate as well as some surface necrotic tissue. These, often cleaned by the clinicians, I mean scraped away to let the tissue breathe. They're nothing to be alarmed about; it's part of the consequences of having those blisters and ulcers in the mouth. How to get rid of them? The only thing the patient can do is brush

with the soft and spongy soft brushes and sponges. I would not scrape them. I would not try to peel that that tissue away. I would let it take its role and it peels automatically with eating and with the process of brushing but not intentionally just to make sure they don't slough tissue excessively.

Becky: Great. Our next question asks, is it recommended for me to numb my mouth before I brush because it is so painful that I don't even want to brush and if I can what products are recommended, where can I get them? Dr. Tanya?

Dr. Gibson: Yeah, I'm not a huge fan of numbing your mouth. I'm afraid that if you numb your mouth you're going to be a little too vigorous with your oral hygiene regimen and cause more damage. I would use mild toothpaste products and do that gentle circular motion at a 45 degree angle and not do that really rough side to side motion that a lot of people do when they're brushing their teeth.

Becky: Great. Thank you. Dr. Nasser, how often is lichen planus associated with pemphigoid? This man says that my dad has oral pemphigoid, but his skin has both pemphigoid and lichen planus.

Dr. Said-Al-Naief: That’s a great question. There is a very rare condition called lichen planus pemphigoides. It's a mixture. Even in the immune fluorescent we see both potential patterns of pemphigus and pemphigoid. They act closer to pemphigoid but that is a proven condition. I had diagnosed 3 total cases in my life so far, no more and it's very rare. You have a pattern of both pemphigus and pemphigoid. And they really honestly they're controlled the same way with the same medications, both. Lichen planus can be controlled with additional medications. But yes this possibility exists. It's not a myth.

Becky: Great. Thank you.

Dr. Said-Al-Naief: May I make a comment about the immune?

Becky: Sure

Dr. Said-Al-Naief: There are patients who have difficulty swallowing because especially in pemphigoid. It's kind of like to go back and throw it in the soft palate. I do use two things especially with the ones that are patients that are kind of painful to them and it's taken a toll on them personally. Either viscous lidocaine, not too long because lidocaine too long in the mouth can burn the tissue believe it or not an additional double sword. I also use Benadryl. People can use Benadryl sparingly. That kind of has somewhat of a numbing situation too and it helps them tolerate eating and brushing without having that sloughing or the blood coming back from the throat. Especially the throat for swallowing in the pemphigoid patients, it helps quite a bit. But as long as they understand there are some side effects of both lidocaine and Benadryl. So we need to be very sparing and make sure you don't swallow. Just rinse with it. Keep it a few minutes at a time and then throw it away. And when you feel better just spit it out

Becky: Great. Thank you.

Dr. Gibson: I want to add one more thing to that because you made me think of something with your comment. I like to use a mixture of Maalox and Benadryl. I mix that together. I have the patients mix that together and use that as a swish and spit and that kind of coats the mouth and it gives the patient a little relief when it comes to performing oral hygiene care regimen and eating.

Becky: Great. Thank you. Our next question is, is it a good idea to get Rituximab if you only have pemphigus in your mouth and how is it determined what the best treatment for me really is?

Dr. Said-Al-Naief: How control is the patient is the issue. If you're very controlled with simple medications, topical steroids or even low doses systemic steroids among other immunosuppressants like triamcinolone for example, you should stick with that regimen. If you have flare-ups, refractory disease, very difficult to control disease, the Rituximab again clears the disease totally in weeks. On the downside you have to be on the medicine for the period of the disease to keep controlling it. Once you stop the medication a lot of studies show mixed results means if he stopped the medication they go back and flare up and some of them get lesser disease when they stop the medication. So it really needs a very nice thorough discussion with the clinician with your physician and then go from there, but I fully believe in the drug. Rituxan is a great

drug to control pemphigus. It's actually now part of the regimens of treating pemphigus vulgaris.

Becky: Absolutely. Dr. Tanya, we've gotten a few questions in about how to slow or stop gum loss due to MMP and there seems to be a lot of questions and postings on a lot of community discussion boards. How common is tooth loss and gum loss and what can be done to stop that?

Dr. Gibson: I think that the tooth loss and the subsequent bone loss and gum loss, I think that's more of a result of not having adequate oral hygiene care and you're losing that due to secondary to periodontal disease. So if we can get the patient's lesions under control that would make the performance of the or care regimen easier for the patient more tolerable for the patient and thereby hopefully slowing down the progression of tooth and bone loss.

Becky: Great. Our next question comes from Clair and she says that she's also having bone loss and says her doctors have advised her to continue with steroids. Is that recommended with oral bone loss?

Dr. Said-Al-Naief: The higher doses of steroids actually have a negative effect on the bone. So if you are on a high dose, it's not recommended. It's probably better to use other medications for suppression like that triamcinolone and even Methotrexate, Plaquenil have to be introduced or Imuran and try to control like Dr. Gibson said hygiene in the best shape if possible. Eventually if and when the patient is controlled, they should have some active treatment like grafting, soft tissue and bone grafting to make sure that the teeth are maintained.

Becky: Along those same questions, Dr. Nasser, Sarah asked should gums be worked on with active disease or should they be left alone? And how does one handle dental work whether it's fillings for cavities or a root canal or even implants when they have active disease?

Dr. Said-Al-Naief: In my humble opinion the patient will not tolerate dental work during active flare-ups, it’s just not possible. So it should be done during the times of control when the disease is controlled when the patient has lesser outbreaks. When the disease has been mediated, then I would go to the dentist. And again the dentist has

to be soft and gentle with the tissues because even if they look good they still have antibody deposits in them believe it or not. So even if the clinical looks good the tissue is still fragile. So no, I would not go when it's flare up when there's a flare up. Similarly you should not floss when there's flare up. You should not do any flossing. Just gentle brushing, kids toothpaste, sponges, all gentle techniques. Rinses that are not alcohol too and then go to your dentist during the times when you have no exacerbation.

Becky: Great. Thank you. Dr. Gibson. Rossana says that her lips are sticking together at night. And what is the best way to prevent this and then to pull them apart in the morning to loosen the damage? She also states that the lining of her mouth is peeling and sticking to food when she eats and she's looking for recommendations for how to stop that as well.

Dr. Gibson: Yeah that's tough. You can use a wax-based lip balm and hopefully that will help. Also, you want to keep your mouth moist. You can also try using the Maalox and Benadryl mixture that we spoke about earlier to help with the eating and the oral hygiene regimen.

Becky: Great. Thank you. Seymour says, Dr. Nasser said that he has pemphigoid and while his blisters are gone his lips are still swollen and he still has a bad oral taste. What can be done to help this?

Dr. Said-Al-Naief: The lips swollen from ulcers? Does he have active ulcers?

Becky: Well, he says the blisters are gone however, his lips are swollen.

Dr. Said-Al-Naief: It's pretty potentially lymphadenitis after the lymphoid tissue within the lips or just simple inflammation. So it really should be looked at as what's the cause of the swelling of the lips. Is it just inflammation, edema, or is there something else going on with the inflamation. There are certain conditions that are characteristically seen with lip swelling. But if it's due to the pemphigus itself or pemphigoid, probably inflammation and edema remains in the tissue even after the healing. That's my best bet without knowing more details about what's the nature of the swell.

Becky: Okay. Thank you. Dr. Tanya you mentioned about dental floss just a little bit ago are all dental floss is the same or is there a type or brand that's better than others and even is a water flosser recommended for patients with pemphigus and pemphigoid?

Dr. Gibson: You want to be careful of anything that's too rough because you don't want to traumatize fragile tissue. So even though flossing is really good it can be kind of rough on the fragile tissues. So I would probably recommend it if you have to be really gentle but maybe a wax-based floss may be helpful or if you're gentle with it some type of water flosser may be helpful. But again, you have to be really gentle and use these products even though you need to use and clean in between the teeth surfaces. You have to be gentle with them.

Becky: Great. Thank you. Ann says that after getting her Rituxan infusion, she experienced tongue inflammation and it turned brown on top. She tried to treat it Nystatin but didn't get any improvement. Is there any other mouthwash or treatment that she should be using?

Dr. Said-Al-Naief: I assume the tongue doesn't have any ulcers on top of it, but I think this is probably every tongue as a secondary to the use of the drug. Rituxan suppresses immunity and when you suppress immunity, there's overgrowth of bacteria and fungus on the tongue, dorsal tongue, which can lead to elongation of the filiform papillae and then pigmentation secondary to those elongated papillae on the surface. So brushing the tongue is a good method of following. I don't think Nystatin is going to help much but people do use it because there may be a fungus element in this. Brushing is a good technique of the surface of the tongue. There is one medication people used to get rid of this staining and the elongation of the surface, but it's a chemo, topical chemo with significant side effects. Therefore I'm not even going to mention the name. Believe me it's not good. We tried to tend to a patient that had more side effects than the Rituxan. So keep brushing the dorsal of the tongue, avoid hydrogen peroxide rinses if you are using them and drink and eat something with cultures like yogurt with culture that will balance the flora of the mouth.

Becky: Great. Thank you. Our next question says, should you see a specialist for cleanings like a periodontist or another specialist?

Dr. Gibson: I think that whether you see a periodontist or a dental hygienist, I think it's important that you see someone who's familiar with oral pemphigus or pemphigoid so that they know how to maneuver around the tissues to be as gentle as possible. So I think it's more valuable to have someone who's experienced with those diseases.

Becky: Great. Thank you. This is kind of on the same line with this next question. My dentist made a recommendation for me to see an oral maxillofacial surgeon for a biopsy. From there the surgeon recommended I see an oral pathologist or an oral medicine specialist. The question is do I need to continue to see all these oral health providers and my dermatologist or only some of them? And who do I decide who I still need to see?

Dr. Said-Al-Naief: Biopsy of vesicular bullous disease is very tricky and I've done quite a bit of them to know. There is a special instrument all surgeons as well as the dermatologist believe it or not, in all specialties. They kind of clip, they put the vesicle in place and they kind of hold it together and they go around it and they take out the vesicle as it is. And that instrument is available in the market. So if the clinician has that instrument and they're used to doing that procedure and that's that should be that clinician. Dermatologists are good, oral surgeons are good, oral pathologists are good oral medicine specialists are good. All of them are good as long as they have been doing this procedure and using that special instrument to clamp that vesicle. The danger of somebody biopsing this superficially is if you do a superficial biopsy and you just clip only the superficial part of the tissue, you will miss the immunofluorescence pattern which may be deposited in somewhere else not sampled in the biopsy. So I think the patient should ask the simple question to the clinician, have you done this procedure or do you do these procedures quite a bit? And who's going to read it? Where is this going to go? Is going to go to a general lab or is it going to go to somebody who reads those lesions. So it's the biopsy, the procedure, the tissue and who's going to read those lesions makes a difference in where they should go.

Becky: Great. Thank you. Very helpful. Yeah. I think that's very helpful. Our next question comes from Kelly and she says that she uses Curaprox toothbrushes, but they're still very painful and her gums bleed profusely. Do you have any suggestions for better at home dental hygiene?

Dr. Gibson: I am not familiar with that particular toothbrush that you mentioned. Hopefully that's something that has very soft bristles that you're using to brush with. And as I stated earlier you want to run hot water over those bristles to get them even more soft for your brushing.

Becky: Great. Thank you. Our next question comes from Mona and she says that she's currently taking a dexamethasone elixir mouthwash. However, it seems that the blisters, lesions and ulcers are always erupting. And as soon as one side of my mouth calms down the other starts, this is common? And then she also says that she's been seeing thrush on her gums, any ideas to help her? Dr. Nasser, do you want to answer that?

Dr. Said-Al-Naief: Yes, of course, I think it with this type of behavior of disease I think we need to kind of switch from the topical swishing, swishing expectorate to some type of a long-term systemic. Either Imuran or Dexamethasone. Even maybe swallow it in certain regimens instead of just spitting the medication out, but it's obvious that her disease is refractory. It's not responding to topical treatment. So, I would apply some muscle to treat it. It needs to be tackled systemically, I think the systemic medications like either low-dose prednisone for a long term, or even Tacrolimus or Imuran, or Plaquenil may help kind or quiet the eruptions and make the patient in a better situation.

Becky: Great, thank you. Dr. Tanya. Summer says that she's been using a dexamethasone oral solution whenever I feel a mucous membrane is about to have a lesion but it's been about 3 months since I've had an ulcer in my mouth. How do I know when I should stop using that oral solution?

Dr. Gibson: I would do it at the recommendation of who's managing your oral lesions. With me when I take my patients off of their oral swish and spits or oral mouthwashes, I wait until we get resolution of the oral lesions. And once you get a resolution of the oral lesions then I take them off. I caution my patients not to stop taking the medication because it may be feeling better because even though you may feel better with your mouth symptoms, you still may have lesions present that you're not aware of. Before I stop the medication I want to make sure that all the lesions are gone. So do it at the doctor's discretion.

Becky: Great advice. Thank you. Dr. Nasser, our next question asks is tooth whitening okay for patients with pemphigus or pemphigoid and what should we think about before we whiten our teeth?

Dr. Said-Al-Naief: Honestly, I wouldn't. There is a harsh chemical in it. It all depends on the patient that they can tolerate it and those trays they put to whiten the teeth or even the process does not irritate the gingiva they can try it but in my experience it's not tolerated well, it burns and makes the tissue inflamed. But if they can tolerate it they can try it.

Becky: Great. Thank you. Dr. Tanya, Our next question says have you noticed people who have gingivitis or excessive tartar often develop pemphigus or pemphigoid? Is there a connection?

Dr. Gibson: There's not a connection with tartar build-up and pemphigus and pemphigoid because they are autoimmune diseases. Now what can happen is that if you do have a lot of buildup on your teeth that can exacerbate the clinical presentation of your gingival lesions, it can make it more intense.

Becky: Great. Thank you. Barbara is asking, does IVIG cause gums to bleed?

Dr. Said-Al-Naief: Not necessarily, no. Immunoglobulins don't necessarily cause bleeding but depending on what may be the underlying we need to probably need to know more information about the underlying condition, why they're giving them immunoglobulin IV. Is it part of the therapy, or is it for some other condition? But there is some vessel fragility potential with immunoglobulins and that's mentioned but not necessarily like a protocol documented. It may be considered and steroids will have some potential bleeding and long term because of the thinning of the tissue. Or maybe she's thrombocytopenic if she has an additional autoimmune component, additional to the pemphigus or pemphigoid which can, by the way they can coexist. For example, pemphigus can exist with lupus, etc. So it probably should be investigated why the bleeding is happening with her physician.

Becky: Okay, great. Thank you so much. Our next question says that when I have a flare of my PV the oral lesions start, but then it moves to my throat at that point. It feels

like razor blades in my throat whenever I eat and drink anything. What is the best treatment for me?

Dr. Gibson: You will probably require some type of systemic treatment. I only use topical treatments if there are only oral lesions. If it’s oral lesions and other places you have to consider systemic treatment for that.

Becky: Okay. Thank you.

Dr. Said-Al-Naief: I'm sorry, I’m intervening also. I think she can also try Viscous Lidocaine. They should try not to use too much since it's going to affect the swelling. If it's too much, they should just barely put it in the back, in and out so that it quiets down the sensation so that she it will help her eat. Very sparingly, so you don't want to unstimulate the swallowing and you don't want to eliminate the physiologic procedure totally.

Dr. Gibson: It will make you swallow your food whole, and I can lead to choking.

Becky: Thank you.

Dr. Said-Al-Naief: That's why I mentioned that it has to be done sparingly because you don't want to suppress the function of the larynx. So it's only if it's only in the soft palate, yes. If it's laryngeal it's a whole different game. But, if it’s soft palate you can without any problems. But if it's laryngeal, it's a different story.

Becky: Okay, our next question comes from Bonnie and she says that she's had MMP blistering gums for two years. She recently had her breast implants removed, stopped her medication and her blisters are almost gone. Is there any relationship between bodily implants and MMP?

Dr. Gibson: Not that I'm aware of.

Dr. Said-Al-Naief: I've seen some studies in the past where, let me try to recall the data. I remember that implants when the tissue is mild, had no significant difference

from the regular peri-implantitis that can happen in the dental immunosuppressed patients. But with the severe autoimmune disease, pemphigus or pemphigoid its case by case based. So clinicians have to evaluate the patient, a single situation and see what's the pros or cons of having implants in the area. The fears from having peri implantitis which is inflammation and inflammation goes in the bone and then it's secondarily affecting the whole predention and the predention is already inflamed with each with the for example gingivitis or ulceration. So it has to be if it's a mild disease.

Becky: Well great. Well that was a very quick hour. I want to thank both of you for being on the call with us today and taking the time to answer a lot of our patients questions. I know I learned a lot by having you here, and I'm sure our community did as well, so thank you.

Dr. Said-Al-Naief: Thank you. Appreciate you.

Dr. Gibson: Thank you.

Becky: I'd also like to give a thank you to everyone who hung with us and listened to the call today and a big thank you Genentech and Principia Biopharma for helping to make today's call possible. Before we leave I have a few announcements. During these uncertain and stressful times we know the importance of ways to help reduce stress. Our next Patient Education Webinar will be on May 7th with Barbara Hee, certified Laughter Leader and owner of Lighten Up with Laughter for a fun presentation on how to reduce stress and laugh. 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!

I know a lot of questions have been submitted throughout our call today and we didn't have enough time to answer them. If you have a question that didn't get answered on the call, or you have additional questions. Please email me Becky Strong at becky@pemphigus.org or call 912-922-1298 extension 105 and we would be more

than happy to help. This call recording will be sent out with a survey after the call. Thank you very much everyone. Goodbye.

Dr. Said-Al-Naief: Thank you, Mrs. Strong. Appreciate it.

Dr. Gibson: Goodbye.