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Time needed to complete: 4h 49m

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Panel Discussion: Real-World Issues in CTD-PAH Screening, Diagnosis, and Management

Announcer:
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Dr. Cuttica:
I'll start off with a question then. So when you decide to treat a patient with ILD-associated PH and your go-to is either sildenafil or Tyvaso, how do you approach the patient? What do you talk to the patient about what their expectation is, or what the expectation is for the patient? Because oftentimes these patients come in, and their major complaint is, 'I'm on oxygen, and I hate being on oxygen, and all I want to do is get off oxygen.' So as they get referred to the pulmonary hypertension clinic as their comorbid condition, how do you approach the patient and have that conversation?

Dr. Moles:
Mike, that is a great question. And it's something that we try to address on the first visit. You know, one of the things that I disclosed with patients who come and they have significant interstitial lung disease, is that there's not a single reason why they're short of breath or they're symptomatic. And we're going to try to find ways to make them feel better. If they have significant pulmonary hypertension, we can address that. Medications for pulmonary hypertension tend to improve pulmonary hypertension. But at least as far as we know, now, they don't improve their underlying interstitial lung disease. So there's always going to be a ceiling effect on the pulmonary hypertension therapy that we give to them. And I tell them, you may experience some improvement in their - in your symptoms as we go. But there may be a threshold where even though we're increasing your PH therapy, you may not feel as better as you were feeling before. And that may be that ceiling effect that it's imposed by their interstitial lung disease.

The other thing that I disclose very early on is PH medical therapy is not intended to get you off oxygen. And in I never, never promise that; it's happened very occasionally. But actually, I give them the reality that pulmonary hypertension medication can potentially worsen their V/Q mismatch and make them need more oxygen. So that's something that is disclosed even before we get to the right heart catheterization on their initial visit.

Dr. Cuttica:
Yes, Val?

Dr. McLaughlin:
My question is about the scleroderma patients or the CTD patients, can you both talk a little bit about some of the things that influence your choice of therapies for them? You know, because they tend to have a lot of comorbidities, they tend to have a lot of GI issues, and many of our PH therapies have GI issues. You know, sometimes their hands are very affected. And it can, you know, can pose challenges for parenteral issues. So, you know, these are common problems that we face every day. So I thought maybe you can summarize that.

Dr. Cuttica:
Yeah, it's a great, great question. They are complicated patients, right? And whether it's the all the comorbid non lung and heart

conditions, like their GI issues going on, or like your case that you presented, the patient that has a little bit of ILD but has a lot of PAH, and trying to figure out what's the right drug going forward. So I think what, at least in our clinical practice, I tend to follow the – you know, we tend to follow the standard AMBITION protocol. Like I tend to, my first go-to as an ERA and a PDE5 for these patients. We do worry about fluid retention in them if they have concomitant underlying heart issues. And then I would say that, when we get to the point of discussing prostacyclin for them, I would probably be more toward – leaning more towards inhaled prostacyclin or parenteral, rather than the oral agents because of the GI issues that our scleroderma patients tend to have.

Dr. Moles:
And I agree with everything you said, Mike. I would add the fact that, you know, I am more concerned about fluid retention with scleroderma patients in particular. There is a high prevalence of diastolic dysfunction. And there was a very interesting study that was published a couple of years ago, data from Denmark, showing that actually that the presence of diastolic dysfunction on an echo predicts mortality, even a little bit better than a pulmonary hypertension does, probably because we don't have very good therapy diastolic dysfunction as we have for pulmonary hypertension. So sometimes what I consider when starting an ERA, I may give them a prescription for a diuretic, and if they start having swelling, you know, they will start that diuretic right away, so we will be prepared to face that. Pump discussion sometimes can be very, very difficult because some patients do need parenteral prostacyclin, but their fingers may have a lot of sclerodactyly. There may be a lot of retractions. Some of the newer pumps that are coming from IV or sub-q administration have devices that are a little bit too technological, even for elder patients. So it's a very individualized discussion. And I don't think that I have a prescription for everybody who has connective tissue disorders.

Dr. McLaughlin:
Yeah. So I agree they're a challenge. And similarly, like, you know, these patients in the REVEAL registry made up a quarter, I think it's probably a larger proportion than that at our place, just because of the big scleroderma program, maybe at Northwestern as well, Mike. But you know, taking that treatment discussion one step further, when we do risk stratification, sometimes there's, you know, there's more challenges with the scleroderma patients as well because they have other problems, other comorbidities, other musculoskeletal issues that can affect things like their hall walk, like their functional class, they also have a lot of other symptoms. So can you just tell us a little bit about, you know, as you go through not just the first treatment choice, but as you go through the reassessment and the risk stratification, some of the other things that you take into consideration as you treat a scleroderma patient?

Dr. Moles:
Yeah. So scleroderma patients in particular are challenging because of their comorbidities. But you know, lupus patients are as well. Rheumatoid arthritis patients can have a lot of musculoskeletal issues. I would say that when I have a suspicion that musculoskeletal issues are affecting their 6-minute walk distance, which will affect their risk stratification, I try to look at other things such as the echocardiogram. If they have a normal right ventricle, and by that I mean a completely normal right ventricle that I can look at and lay eyes on, and make sure that the size the function is good, looking at that RVOT Doppler notching, if it was there before but it was it's not present right now, it makes me suggest that the PVR may be improved.

And the other thing that I find very simple and actually very rewarding is when a patient comes with a severely elevated BMP and then I can normalize that. I think that's a very objective and powerful piece of information that helps me decide that despite the fact that there may be still low-intermediate risk or high-intermediate risk, maybe the risk is overestimated because of that 6-minute walk distance.

Dr. Cuttica:
Yeah, I agree. And I don't want to say I do this more in our connective tissue disease patients but I do think especially in a scleroderma patient, the trends in the walk test, as you say, are they can't or maybe they as we look at like REVEAL risk score, where the risk scoring system is dichotomized at a distance that a patient can walk, maybe our scleroderma patients are going to walk less than 300 meters or less than 200 meters because they've got contractures and their legs are down, so maybe the trend in that individual patient over time carries a little bit more weight than the actual number that's driving the REVEAL risk score down. And again, I don't know if I want to say more, but yeah, the other parameters on the echocardiogram that we're looking at are, am I really convinced that my therapy is improving the function of the right side of the heart for that patient? Which is harder in a scleroderma, scleroderma patient. Yeah, Ruben?

Dr. Mylvaganam:
You mentioned lupus and how patients with lupus and if they're in an active flare or have lupus ongoing, treating their lupus impacts their PH and their PH overall. Is that – do you think that that in some select populations represents a population of patients in which withdrawal of PH therapy might be possible if their lupus activity is well under control, they're on their baseline immunosuppression, their RV is normalized? These are young, in most cases, female patients who have that disease, is that a population like the endarterectomized CTEPH patients where we can start to think about withdrawal therapy?

Dr. Cuttica:
Yeah. That's a great question. So if a lupus patient comes in with a lupus flare, and their PH looks terrible, and you get the lupus under control, and the PH gets better, can you withdraw PH-directed therapy? I'm going to say, one, not answer to your question, and then I'll try and answer your question. One, we have had, and I'm sure you have had over the years, many vigorous discussions with our rheumatology colleagues about the idea of could the flare of the PH or the PH itself be the evidence that the lupus is active, and therefore we need to treat the lupus? And there's always a healthy or oftentimes a healthy back-and-forth about maybe PH should be one of the criteria for lupus in there. So I think that argument is important to make sure the lupus is being treated.

But I do think in a lupus patient that gets treated and everything normalizes, I think there is a pathway. I mean, withdrawal of PH therapy is always a troubling thing and always difficult conversation. But I think there is a pathway in those patients to say, well maybe we draw back. But like any patient, you're going to draw back with close follow-up in them and say we're going to watch it really closely as we withdraw these things away and see. And, again, I think everything in medicine, but particularly PH is, you know, expectation setting with the patient is extremely important, right? And to say to a lupus patient, if you start getting short of breath, I'm going to be worried your lupus is active, and I want to make sure your rheumatologist is involved, and we're going to re-escalate therapy if you're off it.

Dr. Moles:
No, I 100% agree. I think that lupus is a little bit different than the rest of the PAH patients. And I do think that there is a component to, you know, if a patient is having an active lupus flare, I would look at them a little bit different and with more caution. If their PH is based on echo and their right ventricle looks good, I usually you know, tell them just, if they're in the hospital, let's just wait for the lupus flare to come down. Let's reevaluate you back in clinic in a couple of months with an echo and see how you're doing. Anecdotally, I have a patient who we diagnosed in the hospital, which I didn't think that was having a lupus flare. The dermatologist didn't think either. And the PH was quite severe, PVRs in the 15s, cardiac index of 1.7. So we decided to go with triple upfront combination therapy. She was an IV epoprostenol. And 6 months later, she's feeling great. We do a right heart cath. The PVR is 2, and the cardiac index is 5. And she was in a pretty low dose of IV epoprostenol, she was in 12 nanograms. So that was a patient that, you know, we had a shared decision-making and we said okay, you know, this is clearly too much. So we're going to cut down and she - we were able to successfully wean her off IV epoprostenol, and she was doing phenomenal. Actually, I saw her 2 weeks ago. So that can happen, and certainly I consider it a special population.

Dr. Cuttica:
Alright, we're going to wrap it up and break for lunch, and then we'll resume at 1 o'clock with a discussion of left heart disease-associated PH.

Announcer:
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