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CTD-PAH: What Is the Focus in 2023?

Announcer:
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Dr. Channick:
So hello, my name is Richard Channick from UCLA Medical Center. And today we're going to be talking specifically about the connective tissue disease population. And some of the sort of new recommendations and guidelines for these patients. Delighted to be joined by my colleague, Rajan Saggar, also from UCLA. Welcome, Rajan. And Dr. Dinesh Khanna from University of Michigan.

Dr. Khanna:
Thank you.

Dr. Channick:
Our rheumatologist.

So let's start with sort of the beginning for these patients, which is finding them or detecting them. What's new there? And maybe it's not new, but what would you emphasize?

Dr. Khanna:
Yeah, I think one thing that's very clear, Rich, is that people who have scleroderma or scleroderma spectrum, you know, they need to be screened annually. I think what people miss in rheumatology community is that they feel screening is same as diagnosis. And I want to emphasize screening is in patients who are asymptomatic, whereas diagnostic algorithm is for patients who are symptomatic, whether the symptoms may or may not be suggestive of pulmonary hypertension.

The 2022 ERS/ESC guidelines recommended annual screening for scleroderma patients. But what is new is that they recommend it in a patient with scleroderma who's breathless for unexplained reasons, a right heart cath may be recommended, which I thought was a very important recommendation this time.

Dr. Channick:
And then maybe I'll just follow up really quickly with Raj. I mean, we see a lot of these patients with unexplained dyspnea, what do you think of that recommendation? And where does cath fit in? Or even exercise testing?

Dr. Saggar:
Yeah, so I think, you know, the recommendation there for the right heart cath is really to express the point that you really don't want to miss occult pulmonary hypertension, I shouldn't say occult - manifest pulmonary hypertension in a patient who clearly is symptomatic that you have no other explanation for. But to your point, there's other ways to potentially get at that. And maybe, as you know, most of these patients are breathless on exertion, so we - there's a growing literature for exercise echocardiography, we have the cardiopulmonary exercise testing that you can do, which is - can be invasive, non-invasive, it may involve a concurrent right heart catheterization at some centers where we can do actually do the CPET with the right heart catheterization. But I think the majority of

centers are offering just simple cardiopulmonary exercise testing. And I think all of those modalities - and of course, you could throw in cardiac MRI there as well, at least at rest, all of those modalities, I think, particularly the exercise, the provocative ones, will actually - can actually provide some information there.

Dr. Channick:
Yeah. Because I mean, it's - these people can be short of breath for so many different reasons, whether it's their heart, their lungs, their peripheral muscles. And so it's – or just general illness. And so, it makes it, you know, quite complicated, I think, to figure out why a patient is having these symptoms. I think it also, you know, ties into the aspect of how we make a diagnosis in terms of treatment, specifically as it relates to pulmonary hypertension, we've talked a lot about comorbidities, and this - the new recommendations related to maybe more cautious treatment in patients with comorbidities. How does that apply? And it clearly does to the scleroderma or CTD population in general?

Dr. Khanna:
Yeah, thank you for the question. I think it is a difficult situation. On one hand, you have scleroderma or rheumatoid arthritis, patients who have increased cardiovascular disease due to inflammation who have more diastolic dysfunction, we tend to give them quite a bit of glucocorticosteroids, so we are causing diabetes. On the other hand, we know that scleroderma patients have a high mortality due to progressive pulmonary arterial hypertension. In fact, it's probably the most associated with the highest mortality for all the connective tissue diseases.

So I think from a rheumatologist and a cardiologist or pulmonologist, it becomes an important conundrum whether, do you treat them aggressively? Or whether you don't treat them aggressively? And that's a question actually, for both of you. How do you approach a patient with scleroderma who has diabetes or diastolic dysfunction? And how do you treat them knowing that this will likely be the cause of their death in the next 5 to 10 years?

Dr. Saggar:
That's an issue that I think we deal with all the time. But, and to Rich's point earlier, since you can develop pulmonary pretension in the setting of scleroderma or systemic sclerosis, that spectrum of disease, or frankly any CTD from several different sort of mechanisms, right? I mean, you can come at it from a left heart mechanism, a thromboembolic mechanism, you know, a precapillary mechanism. And there may be other associated conditions as well, right? So you really are - you're dealing with such a heterogeneous potential for the type of pulmonary hypertension. But what you don't want to miss is Group 1 PAH. And because of its mortality.

Dr. Channick:
Let me dig in there a little bit. We have a couple of minutes. I mean, you're pulmonologists, you're world's expert in interstitial lung disease and pulmonary hypertension. How do you make that distinction? I mean, these people have a little ILD, they've got some PAH, how do you actually, in real life, determine what's causing what and how you should treat it?

Dr. Saggar:
Right. So it just - it goes back to this whole thing where I feel like if the hemodynamics are significant, and I think we all can agree on a significant set of, you know, severe hemodynamics, for instance, and we're talking about someone with comorbidities for left heart disease, let's give them three comorbidities. Okay? And/or significant interstitial lung disease, either spectrum, either the cardiopulmonary phenotype, if you will, or the left heart phenotype with significant precapillary pulmonary hypertension. My approach there is I think that's a very mortal condition. And for me, I think the, you know, the idea is that I would offer that patient pretty much because I know that their mortality is high, regardless, and I risk not treating Group 1 PAH, I will offer them an aggressive trial of therapy, unless they have a side effect or they fail or there's some other adverse reaction profile. I am pretty aggressive in that setting. You agree with that?

Dr. Khanna:
I would agree. I think the challenge and what the 2022 ERS guidelines has challenged us is to really keep in mind the comorbidities, acknowledge it, but also sort the difference between pulmonary arterial hypertension, whether versus PH, ILD, or left heart disease. But once you have done that, you really have to treat them aggressively.

Dr. Channick:
Yeah, it sounds like we're in agreement. And that's a really important message and your particular patient population with the CTD.

Well, that's all the time we have. Thank you, guys, for joining us. And thank you all for listening.

Announcer:
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