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Phenotyping the ILD-PH Patient To Determine Treatment Strategy

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Dr. Shlobin:
Hi, I'm Oksana Shlobin. I am Medical Director of Pulmonary Hypertension Program at Inova Fairfax Hospital. Joining me today to discuss pulmonary hypertension in interstitial lung disease is Sudar Rajagopal from Duke University, and Raj Saggar from UCLA. Welcome.

So this is an exciting time for the field of pulmonary hypertension in interstitial lung disease. We now have an approved therapy of inhaled treprostinil to treat these patients. And it's a huge advancement because we've dealt with these patients with no approved therapies for years. These patients have horrible quality of life and very high mortality. So I'm excited to talk to you about this topic.

So, Raj, let's start with you. Tell me how you phenotype those patients. There is this sort of big controversy, some - whether someone has Group 1 versus Group 3, tell us a little bit more about how you approach to phenotyping?

Dr. Saggar:
Yeah, Oksana, it's a great question. And like you said, it is a controversy in the sense that I'm not sure it's a perfect art just yet, so but we do have some markers for sort of how to distinguish the two groups. One of the key differences, of course, between Group 3 and Group 1 is that you have to have some form of lung disease to be Group 3. And so, if you look at the, you know, the study that got the inhaled treprostinil approved, they asked for diffuse parenchymal lung disease on the HRCT of the chest. So I think having enough lung disease, and as you know, in a lot of our Group 1 PAH studies, we allowed for mild interstitial lung disease or fibrosis as part of that phenotype. So basically, anything more than mild, diffuse parenchymal lung disease, interstitial lung disease, or fibrosis - and/or fibrosis would qualify, so moderate or more, I think would cover that base.

And then I think, in general, the hemodynamics, you know, we tend to ask, or tend to find patients with sort of more advanced hemodynamics. It's not necessarily necessary, but the more pulmonary hypertension they have, the more likely they are to have a pulmonary hypertension phenotype, and in the setting of that lung disease be more likely to have a Group 3 phenotype as opposed to a Group 1. We obviously use spirometry. Spirometry, as we all know, preservation of the forced vital capacity, the more that's out of proportion, that preservation, to a decline in the diffusing capacity; hence, that FVC/DLCO ratio being more elevated is more supportive, perhaps of a phenotype that supports a Group 3 PAH.

And then what's fun about this whole thing is also connective tissue disease being a risk factor, really, for both types of pulmonary hypertension. So that throws a wrench into the whole thing as well. And what you really don't want to miss, right, is the opportunity to treat the patient. And now with the approval of inhaled treprostinil, there at least is an option, which I think, given the mortality of these patients, we may want to consider the use of that medication, at least as a trial, even when there may be a little bit of a muddy, gray area perhaps, between how you want to label that patient Group 1 versus Group 3.

Dr. Shlobin:
Thank you for that. That was a great explanation, and a guide, perhaps, to phenotyping.

Sudar, I wanted to explore this just a little bit more. These patients tend to be older, they tend to have cardiac comorbidities, so hypertension, coronary artery disease, sometimes A-fib, sometimes sleep apnea. And we do know that some pulmonary vasodilators actually can be detrimental in patients who have had that, for example, or pulmonary venous vasculopathy. Tell me a little bit more how you tease out this cardiac phenotype in the setting of parenchymal lung disease, or I guess left heart disease phenotype?

Dr. Rajagopal:
That's a great question. It's a challenge I think we all run into. As you stated, you know, a lot of these folks with ILD, especially the IPF patient. When you think about your typical IPF patient, you're talking about an older man and who's had a smoking history. So when you're thinking about those patients, they have all the same risk factors for heart disease, whether it's coronary artery disease, A-fib, or, and of course, HFpEF. And with age, of course, in all of us, there is some diastolic stiffening and you're going to have some type of, you know, abnormal diastology that could contribute to their presentation.

So really, it's just like Rajan was talking about, when we phenotype these patients, it's a really in a very personalized way, we have to figure out what we think is contributing to their dyspnea and what their degree of precapillary pulmonary hypertension is. Although even that term precapillary pulmonary hypertension can be a bit misleading because you can have a high PVR due to pulmonary venous remodeling as well. So it's really trying to figure out whether we think that they have significant pulmonary arteriopathy there that's related to their underlying lung disease.

And so definitely on the right heart cath, when we look at that, yes, for a typical IPH patient, we're looking for that PVR for diagnosis greater than 2, but historically, for treatment greater than 3 Wood units. If you look at the INCREASE trial, they did treat patients, you know, for entry, the cut-off was a PVR greater than or equal to 3 Wood units. But the benefit in their sort of subgroup analysis on the forest plot, they saw more of the benefit in that greater than 4 Wood units population. So definitely, if you're seeing that in there in their hemodynamics, they have a higher PVR, if on their echocardiogram, they show more evidence of right heart failure, ideally, they don't have a ton of left atrial enlargement, obviously severe left atrial enlargement is always a red flag in all of these patients, those are the times now immediately, you're thinking okay, maybe these are patients who could benefit from that.

And then there are other approaches too, right, we can volume load the patient to see how their wedge responds to that volume load. And you can do things like invasive cardiopulmonary exercise testing, or just regular cardiopulmonary exercise testing, and determine whether they're more respiratory or cardiac limited. If they're more cardiac limited. That definitely gets me more concerned about right heart failure being the main issue with their presentation.

Dr. Shlobin:
Well, thank you so much. I really appreciate that both of you joined me here today to discuss this important topic of phenotyping patients with pulmonary hypertension in the setting of interstitial lung diseases.

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