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Treatment Approaches to ILD-PH

Announcer:
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Dr. Shlobin:
Hi, I'm Oksana Shlobin from Inova Fairfax Hospital. Joining me today to speak about treatment of pulmonary hypertension in the setting of interstitial lung diseases is Sudar Rajagopal from Duke and Raj Saggar from UCLA.

I would love to talk to you guys about treatment of PH in patients who have interstitial lung disease. We finally have a drug that was approved in the United States, inhaled treprostinil, to treat these patients the first time we have an option, a treatment option, which is FDA approved. So Raj, let's start with you. How do you holistically approach these patients? Because obviously, they don't only have pulmonary hypertension, they also have parenchymal lung disease and maybe some other comorbidities.

Dr. Saggar:
Yeah, that's actually the main point is that, in fact, they often do have existing comorbidities. They certainly have an underlying lung disease, which comes in so many different flavors. It's quite heterogeneous. Sometimes we just have a smoking-related combined pulmonary fibrosis emphysema phenotype, sometimes we have a chronic hypersensitivity phenotype, or an autoimmune disease. So all of those perhaps are being treated differently.

So the first thing we do is make sure we also address their parenchymal lung disease. And obviously, if they're smoking and those kinds of things, we want to smoking cessation to be part of this. But in terms of addressing their parenchymal lung disease, as you all know well, we have, you know, two approved anti-fibrotic therapies, nintedanib and pirfenidone, which can be used as well. I think one of the things we struggle with is some of the side effect profiles of, for instance, the pulmonary hypertension therapy like inhaled treprostinil, an anti-fibrotic, sometimes there's some GI-related issues with all of these medications. So the idea of starting everything at the same time, perhaps staggering in medications if someone is suffering more from a pulmonary hypertension issue, and more stable lung disease, perhaps the PH therapy goes on first. Or it may - they may come to you already with an anti-fibrotic on board and they're already having GI issues so you have to sort of temper that if you're going to treat their pulmonary vascular disease. So those things can become an issue. But those are some of the finer details. The point is you have to think about treating their parenchymal lung disease.

In addition to that, the comorbidities, obviously easy to say, but hard to treat, and I think Sudar will get into that. But the other aspects from a pulmonary standpoint is making sure they're oxygenated. I think we oxygenate them well at rest, but sometimes we don't actually often make sure that they're getting an adequate oxygenation during exertion. So making sure they have that at home, and also when they leave the house, adequate oxygenation is probably, you know, Pulmonary 101 in this kind of setting.

If they have sleep disordered breathing, we tend to sort of look into that and help them out with getting adequate sleep; that can contribute to their symptoms like fatigue during the day or exertional limitation, functional limitations, a lot of that can be driven by poor sleep hygiene and sleep disordered breathing. And of course, you know, when the time is right, we have to address palliation. And so there's a multimodal type of approach that we try to always think about when we're treating such patients.

So it's easy to you know, prescribe the medications, but you've got to think about all the other issues that are sort of in play for a given patient.

Dr. Shlobin:
Great. That was excellent, excellent overview. Sudar, a question for you. So we do have this therapy. It was FDA approved for pulmonary hypertension and interstitial lung disease in the United States. But recently, we have the guidelines from Europe, our European colleagues that came out that sort of put a little bit of a different spin on the subject. So in their recommendation, they do not advise treating pulmonary hypertension in setting of interstitial lung disease unless it is severe, with the caveat that inhaled treprostinil may be considered in all of the patients with PH due to ILD. For our U.S. doctors who treat patients, how would you interpret it?

Dr. Rajagopal:
Yeah, I think it's important to remember that inhaled treprostinil for ILD is only approved in the United States. And then if you look at practice patterns with inhaled treprostinil, it's really largely used in the United States. Most people in Europe don't have availability to it. So they're, really I think the European guidelines are coming from a bit of a different perspective in terms of practice pattern. If you look at the guidelines, it's a 2B recommendation. So as you said that you may consider use of inhaled treprostinil in PAH ILD based on that. Of course, if you look at the trial, the trial was extremely positive, and showed a significant benefit. And the cutoff for entry into the trial was a PVR of greater than or equal to 3 Wood units. And then if you look at the subgroup, the subgroup that seemed to get the most benefit was anyone above greater than 4 Wood units in terms of PVR, so that would suggest that data is a bit discordant with what we see in the ESC/ERS guidelines.

I do think they do have an important point in that we really don't have good data for other PAH-specific therapies in PH ILD. And there is evidence of harm with some of them, such as riociguat in the RISE-IIP trial, which was stopped early due to harm. So definitely, from that perspective, I agree with, you know, the approach or the guidelines there and that the one that you should consider is inhaled treprostinil for these patients.

But I think looking back and getting that 10,000-foot view, we really have to personalize treatments for our patients. You have to really see how they respond to the therapy even just if you just say 'Oh, inhaled treprostinil is approved, and I'm going to use it.' You also have to be careful in how you use it and how you up-titrate the medicine. You know, some patients do better with a nebulized therapy than they do with a dry powder inhaler. And it's going to vary from patient to patient.

So I think the important thing is to remember to personalize your care, look at the patient in front of you, and see how they're responding to that therapy you're using.

Dr. Shlobin:
Great, great, thank you so much. So phenotype and personalize the care and have a holistic approach to treatment. That was very, very useful.

So thank you very much for joining me today as we discussed treatment of pulmonary hypertension in interstitial lung diseases. Thank you, guys.

Announcer:
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