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Clinical Perspectives on Idiopathic Pulmonary Fibrosis

Announcer:
You're listening to *Deep Breaths: Updates from CHEST* on ReachMD. This is a non-promotional, non-CME disease state educational podcast produced in partnership with the American College of Chest Physicians and is supported by Three Lakes Foundation, aimed at improving timely recognition and diagnosis of interstitial lung diseases like pulmonary fibrosis.

Your host is Dr. David Schulman, the President of the American College of Chest Physicians and is a Professor of Medicine in the Division of Pulmonary, Allergy, Critical Care, and Sleep Medicine at Emory University in Atlanta. Here's Dr. Schulman now.

Dr. Schulman:
Welcome to *Deep Breaths: Updates from CHEST* on ReachMD. I'm Dr. David Schulman, and joining me to share clinical perspectives on idiopathic pulmonary fibrosis are Dr. William Lago and Dr. Tejaswini Kulkarni.

Dr. Lago is a Clinical Assistant Professor of Family Medicine at the Heritage College of Medicine at Ohio University, and also the Medical Director of Cleveland Clinic's COVID reCOVer Clinic in Independence, Ohio. Dr. Lago, welcome to the program.

Dr. Lago:
Thank you very much for having me. It's a pleasure to be here.

Dr. Schulman:
And Dr. Kulkarni is the Director of the Interstitial Lung Disease Program at University of Alabama at Birmingham. Dr. Kulkarni, thank you for being here with us today.

Dr. Kulkarni:
Thank you for the introduction, Dr. Schulman, and it's a pleasure being here.

Dr. Schulman:
I appreciate both your time, and I'm looking forward to a very robust discussion. Let's start with Dr. Lago. Why do you think it's so challenging for primary care physicians to diagnose idiopathic pulmonary fibrosis?

Dr. Lago:
I think diagnosing idiopathic pulmonary fibrosis is tough for primary care physicians because of a number of reasons. If you think about it, symptoms tend to be very vague. A lot of people just present with things like shortness of breath or cough that can mimic as a lot of other issues. Primary care physicians I think also don't have quite as much understanding of the different treatments and the different outcomes that can occur with pulmonary fibrosis.

The other thing I think that makes it tough is misdiagnosis. Oftentimes, we may simply want to adjust a medication for a patient that already has a pulmonary disease, or we may focus on symptoms that tend to be more deadly for the patient initially, such as cardiac issues.

Dr. Schulman:
One quick follow-up. What role, if any, do you think challenges with navigating procedures and other things in the healthcare system plays?

Dr. Lago:
I think that certainly navigating the healthcare system can be tough. You know, depending on where a patient is from, obviously getting

to appropriate settings can be difficult if they're in a rural setting, such as travel, finding people that they can see, for instance, if they're trying to see someone who's in a pulmonary fibrosis clinic, or if we want to have them perform, for instance, a high resolution CAT scan that is not always readily available for certain folks. And in addition, obviously referrals getting things pre-approved through insurance can often be a very, very difficult thing for both patients and physicians.

Dr. Schulman:
Thank you. On the topic of delays, I'd like to turn to Dr. Kulkarni. Dr. Kulkarni, how did delays like the ones that Dr. Lago just referred to impact prognosis of patients with idiopathic pulmonary fibrosis?

Dr. Kulkarni:
Both nintedanib and pirfenidone medications that have been approved for treatment of idiopathic pulmonary fibrosis have some degree of anti-inflammatory but mostly anti fibrotic activity. They slowed down the rate of the decline in lung function, and there's some data that they could lengthen survival. However, there is no cure for idiopathic pulmonary fibrosis. Essentially, time lost is lung lost.

But unfortunately, like Dr. Lago mentioned, there are several levels from the patient presenting with symptoms, to delays in healthcare system, to finally getting a diagnosis and starting treatment. The delays in the diagnosis is really common in this patient population and thus there's ultimately delay in patients receiving the appropriate treatment. So it's very important that we focus on trying to identify these patients early, thus trying to impact the prognosis in patients with IPF.

Dr. Schulman:
For those just tuning in, you're listening to *Deep Breaths: Updates from CHEST* on ReachMD. I'm Dr. David Schulman, and I'm speaking with doctors William Lago and Tejaswini Kulkarni about clinical findings in idiopathic pulmonary fibrosis, or IPF for short.

Dr. Lago, I want to circle back to you a little bit and maybe switch gears for a minute. Primary care physicians have a really tough job. They evaluate folks with a wide range of symptoms, and even when somebody comes in with coughing, shortness of breath, it could represent a wide range of things, idiopathic pulmonary fibrosis, other interstitial lung diseases, or something else entirely. What can we do to make primary care physicians more aware of the possibility of interstitial lung diseases, and specifically IPF, in folks presenting with these early symptoms?

Dr. Lago:
Sure. Thinking about it as you had discussed, you know, as a primary care doctor, we're faced with complex patients with multiple problems. And so oftentimes, we have to separate out what do we worry about the most right at that instance? You know, what's going to kill them tomorrow? So, a lot of times, things like ILD get put on the back burner as we pursue other things. So, I think anything we can do to bring that awareness, at least on board for us as primary care doctors, will help with that. So, if we can get tools that we can use easily, tools that we can utilize while the patients are in the office, that will help us to better diagnose and better understand that, I think that will make a difference. I think any way that we can educate us as primary care doctors to things that are coming down the pipeline as far as treatment and as far as testing to help us diagnose that will help also.

Dr. Schulman:
Thank you, Dr. Lago. Turning now to Dr. Kulkarni, one of the things we've just heard about are tools that should be made available to primary care physicians to help them make an early diagnosis. There was a recent white paper looking into this, there is the ongoing Bridging Specialties Initiative. How do you foresee these tools helping to address the gaps that we've just identified?

Dr. Kulkarni:
Absolutely. So the Bridging Specialties Initiative was created by CHEST and Three Lakes Foundation, really to look at understanding the gaps that exist in, that lead to a delay in diagnosis of ILD, and then work towards bridging that gap.

So the survey results, you know that we got from sending out surveys to primary care physicians and community pulmonologists started to give us an understanding of some of these gaps in knowledge as well as clinical behavior that contribute to significant delays in diagnosis, limitations of the treatment options, as well as, which ultimately lead to a worsening quality of life for some of our patients.

With these data, we've created a toolkit and the components include a patient questionnaire, eLearning module, which is geared towards early ILD detection, decision-making tool, which is based on patient case scenarios, resources that include radiology modules, infographics, and some educational games. For examples, the reported items that were most likely included when taking a patient's history did not include early symptoms that could give you a clue for ILD. For example, various household exposures or review of autoimmune conditions or medications that can cause interstitial lung diseases. So the updated patient questionnaire allows for all this data to be collected before even the patient sees the primary care physician. That's really helping us with addressing the issue where time is really short for each of these visits.

Another thing that we learned from the survey results are that almost half of the primary care physicians refer patients for cardiac workup, and that is the leading differential diagnosis when it comes to shortness of breath. And that is followed by shortness of breath and cough with COPD and asthma, which are much more prevalent, much more common than interstitial lung diseases. Only a minority of the respondents said that spirometry would be a part of their initial workup, and an even lesser CT scans.

And so the eLearning module was created to help understand how to diagnose ILD early; what are the signs to look out for; and, what is the initial workup to do? And then the decision-making tool includes patient case scenarios to work through these steps of asking them the right questions and history, physical exam, and then spirometry, chest x-rays, and CT scans. So as the toolkit is updated, our next steps will really be to think about practical implementation as well as dissemination of this toolkit to primary care physicians as well as pulmonologists.

Dr. Schulman:
Thank you. As we close, I want to get a couple of final thoughts from each of you. This is a complicated issue. There are a lot of barriers to early diagnosis, but you both iterated the importance of that diagnosis in terms of getting focused on treatment and trying to minimize the irreversible components of the disease. So I'd like to get just a couple of thoughts from each of you as we close. Dr. Kulkarni, do you have any final takeaways that you'd like to leave with our audience?

Dr. Kulkarni:
I think what I want to really focus on is, you know, we have to work together in the healthcare system starting from patient's initial symptoms where they present to primary care physicians to them doing the initial workup or referring to either community pulmonologists and/or from there, to an interstitial lung disease center, really to work together to not just start on therapy, either antifibrotic and or anti-inflammatory therapy depending on the type of ILD but to also include a more comprehensive management. Ultimately, early diagnosis leads to early treatment. And that is what we really want to focus on. Finally, just really want to emphasize time lost is lung lost. So, the earlier we can diagnose our patients, the better our outcomes will be.

Dr. Schulman:
Well said. And Dr. Lago, I'll give you the final word.

Dr. Lago:
Thank you. The big thing I think that we need to do, particularly as primary care physicians, is try to put this on our radar. If we're not thinking about ILD as a potential diagnosis, we're going to potentially delay people's treatments, we're going to delay their overall outcome and impair their quality of life. So by working together with our pulmonologists, by getting better knowledge of the disease process, and the tools that we need to do in order to diagnose it, I think the only thing we can do is certainly help our patients in the long run. And so obviously, that's our challenge, I think with this initiative.

Dr. Schulman:
With those key takeaways in mind, I want to thank my guests, Dr. William Lago and Dr. Tejaswini Kulkarni, for joining me today and sharing their insights on idiopathic pulmonary fibrosis.

Dr. Lago, Dr. Kulkarni again, great having you both on the program.

Dr. Kulkarni:
Thank you for having us.

Dr. Lago:
Yeah, thank you very much.

Dr. Schulman:
I'm Dr. David Schulman. Thanks for listening.

Announcer:
This episode of *Deep Breaths: Updates from CHEST* was produced in partnership with the American College of Chest Physicians in partnership with The Three Lakes Foundation. To access other episodes of this series, visit ReachMD.com/CHEST, where you can Be Part of the Knowledge.