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Bridging Specialties™: Timely Diagnosis for ILD Patients

Announcer Introduction:
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 the 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 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:
Affecting around 400,000 people in the United States, interstitial lung diseases, or ILDs as we'll be referring to them today, are a group of respiratory disorders that cause lung inflammation and may cause permanent scarring. The most common interstitial lung disease, idiopathic pulmonary fibrosis, or IPF for short, is a rare disease that causes irreversible scarring of the lung tissue. Approximately 30,000 to 40,000 people are diagnosed with idiopathic pulmonary fibrosis each year, the majority of whom lose their lives to the disease just a few years later.

Welcome to *Deep Breaths: Updates from CHEST* on ReachMD. I'm Dr. David Schulman on behalf of the American College of Chest Physicians and the Three Lakes Foundation where we are working together in an effort to reduce the time it takes to make a diagnosis of ILD. Joining me today on an initial focus on pulmonary fibrosis are Drs. Tim Hernandez and Andrew Limper.

Dr. Hernandez is a Clinical Professor of Medicine at the University of Texas Health Sciences Center in San Antonio. Dr. Hernandez, welcome to the program.

Dr. Hernandez:
Thank you.

Dr. Schulman:
And Dr. Limper is a Professor of Medicine and Director of Thoracic Research in the Pulmonary and Critical Care Division at the Mayo Clinic in Rochester, Minnesota. Dr. Limper, it's also great to have you here with us today.

Dr. Limper:
It's great to be here with you.

Dr. Schulman:
It's a nice pleasure to have a chance to talk to folks like yourselves about this incredibly important issue. And I'd like to start with you, Andy, if it's okay. What can you tell us about the epidemiology and pathophysiology of IPF? And is there a difference between pulmonary fibrosis and IPF?

Dr. Limper:
Well, certainly, that's a great question. And then there are differences between general pulmonary fibrosis and idiopathic pulmonary fibrosis. It's our job that we look at the various causes of lung fibrosis and that might be related to arthritis, autoimmune disease, hypersensitivity, or even chemotherapy for cancer or radiation. Those are diseases that have specified causes for fibrosis. When we go through that whole list, and there's well over 260 causes of fibrosis, and we come up with none of those, then what we're left with is idiopathic pulmonary fibrosis. That's pulmonary fibrosis for which there is no known cause.

Now we're learning a lot about IPF, idiopathic pulmonary fibrosis. First of all, it's a disease that becomes much more common as we

age, you know, very common in the 60s, 70s, 80s, or beyond. If we see patients in their 50s, we actually think that it might well not be IPF. Traditionally, it's been thought of as a disease that is predominant in men compared to women. And that's probably not true and evolving. When we look at Medicare populations, there's pretty equal impact of the disease upon men and women.

As far as the pathophysiologic mechanisms, those are evolving. But basically, the bottom line is scar forms in the lungs, there's areas of lung injury with scarring; that causes the lungs to be stiff, and that causes the gas exchange to drop. And unfortunately, over time, the disease progresses and leads to loss of good gas exchange, the need to use oxygen, and ultimately, it can prove to be fatal.

Dr. Schulman:
I want to hear from Tim a little bit on this issue. Given that dyspnea, Tim, is a common complaint in clinical practice for multiple reasons, and it's often misdiagnosed at least at first pass, are there specific clues in a given patient's history that might make you consider IPF in your differential diagnosis fairly early? And if so, what are the triggers that might cause you to refer your patient to a pulmonary specialist earlier rather than later?

Dr. Hernandez:
The usual clues that I find in patients is of course, as you mentioned earlier, when the patients are older usually will happen in a patient that has progressive, gradually worsening, shortness of breath, and in a patient that probably has smoked in the past. That makes me worry more about IPF. A lot of these patients will also have a chronic cough. So, whenever I have a patient that has these things that a chest x-ray isn't showing anything and physical findings suggest that they might have IPF, that's when I start beginning the workup for it.

Dr. Schulman:
Andy, one of the things that we see fairly frequently in this diagnosis of IPF is a relatively late diagnosis because the symptoms are sort of indolent, they don't, rapidly fluctuate certainly not early on, in most cases. In patients with longstanding shortness of breath, cough, fatigue, and these fairly vague, indolent symptoms, what are the indicators that you as a pulmonary specialist might use to determine this is somebody on whom I want to get CAT scanning, a full set of pulmonary function tests, and maybe additional things that might help you better work up folks for idiopathic pulmonary fibrosis?

Dr. Limper:
So, when somebody has chronic dyspnea, or cough or fatigue, I think some things that tend to not be attended to is common things are common. So, folks will think, ‘Oh, the chest x-ray’s a bit abnormal, is this a pneumonia?’ bilateral shadows, they'll give antibiotics, they may give inhalers, they may try corticosteroids. But the reality is if those early interventions don't change the symptoms of dyspnea, and I would say cough, and some people present with major cough, you need to think about getting testing early, just as it's been explained. And that would include complete pulmonary function tests, as well as a noncontrast, high-resolution CT scan of the chest. I will also say, oftentimes, there's clues on the plain old chest x-ray that folks have in the office. Some of these are read as dirty x-rays, increased markings in the bases, some scar in the bases. That should be a trigger that that's not a normal x-ray.

Lastly, I got to add, listen to the chest because when you hear crackles in the bases, or clubbing in the fingers, that's not what you see in COPD and it's not what you see in asthma and other causes. So those crackles are important to hear. And I've had some of my primary care docs within Northern Iowa and Southern Minnesota, call me up and just say the chest doesn't sound right, could you see the patient, and that causes a trigger of that testing that gets earlier diagnosis.

Dr. Schulman:
I want to turn next to treatment. But before we do, 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 Drs. Andrew Limper and Tim Hernandez about idiopathic pulmonary fibrosis, or IPF.

Andy, let's talk a little bit about treatment. Can you talk a little bit about the typical course of idiopathic pulmonary fibrosis and what treatment options are currently available to manage it better?

Dr. Limper:
Sure. Well, unfortunately, this disease tends to progress over time. I would say that the rate of progression varies in patients widely. Some people progress slowly over time. Some people unfortunately, have more rapid deterioration.

Now we do have treatments for this disorder. The traditional treatments of immunosuppression with prednisone, azathioprine, and other kinds of immunosuppressions really do not work in this disease, and there's some evidence they might be harmful.

Over the last four or five years, we've had antifibrotic agents, pirfenidone and nintedanib. These agents slow the course of the fibrosis in the average patient over time. And they do have benefits in that they do reduce the risk of hospitalization. And over a couple of years, they also do have benefits on mortality for patients taking the antifibrotics. I believe that it's important to consider the antifibrotics in most

patients, but there's also a whole comprehensive treatment that needs to be considered.

We need to assess oxygen needs. Oxygen replacement needs, very important in these patients, which will usually be progressive over time as well. We also need to worry about things like exercise. Patients need strong muscles to move the stiff lungs that they have from the fibrosis. And routine exercise, even walking or even perhaps pulmonary rehabilitation, can be beneficial for these patients. I also always query about reflux, because we know that reflux can be a cofactor that may accelerate the disease. I look for that and then I treat it, both with acid medications, proton pump inhibitors.

Finally, there are other diseases or disorders that are common along with IPF. And one is sleep-disordered breathing or sleep apnea. And I do screen all patients for that. It's very frequent that I do find it. And that needs to be treated as well, both because it supports the oxygenation levels at night, but also because when there's obstructive efforts to breathe against an obstructed upper airway, that actually promotes reflux and small-volume aspiration. So important things.

Dr. Schulman:
Andy, very quickly as a follow-up, are any of these therapies that a general internist or primary care doc should consider prescribing once the diagnosis is made? Or would you advise that this really be a pulmonologist routinely?

Dr. Limper:
Well, the comprehensive general medical approach that I talked about actually can be the general internist or primary care doc can be very helpful and making sure all that happens. Specifically, though, with the antifibrotics, those medicines are expensive and require preauthorization. And oftentimes, the insurance review and approval process requires that a pulmonologist prescribe them. And across the country about 97, 98% of the prescriptions are provided by pulmonologists, either in the community, which is fine, or in referral centers, which is fine. Anybody that's comfortable with that diagnosis and treatment can prescribe the antifibrotics.

Dr. Schulman:
Tim, I want to turn back to you for a moment. Have you ever had any challenges, barriers, experiences that you can recount where there's been difficulty getting pulmonary function testing, CAT scanning, or formal referral to a pulmonary specialist for a further workup?

Dr. Hernandez:
I've been fortunate that I don't run into those barriers. But I can imagine that there would be certain circumstances where a primary care physician, internal medicine, or family practitioner, or one of the APPs would have trouble obtaining pulmonary function tests or a high-resolution CT. Typically, what I do is just document that the patient is continuing to have dyspnea, or the dyspnea is worsening, and that the chest x-ray may be equivocal, but that the patient still has the symptoms, and I don't have an issue obtaining those tests. However, I can imagine in some settings, people in their clinics might have simple spirometry, and things that may sort of point to a direction that something's going wrong. But I still think that complete pulmonary function testing is needed to help verify the exact diagnosis that's occurring.

And with regard to referral to a pulmonologist the way I overcome that obstacle is I try to diagnose the patient ahead of time, and not just send the pulmonologist a patient that has a cough or shortness of breath without some kind of a workup. And then I like to be collaborative with my specialists and subspecialists or superspecialists and speak to them directly. Because I think that's the most important thing. Whether you're in the hospital or in the clinic primarily, I think you need to talk to your colleagues because in modern medicine, there's a great disconnect between physicians that are in the hospital and those that are in the clinic. And I think our patients suffer because of it.

Dr. Schulman:
We're going to wrap up here, but I'd like maybe one more comment from each of you on the topic of collaborative care. Can you speak a little bit to the role of collaborative care in the early detection and rapid administration of treatment of IPF? Andy, I'll start with you, and then I'll throw it to Tim for the last word.

Dr. Limper:
Yeah, I think that getting at this diagnosis, and the appropriate treatment early is going to take a team. And at least working in tertiary referral center as somebody that specializes in pulmonary fibrosis, I want to keep those patients working with their primary care docs and their local pulmonologists. And I'm here as a resource to help with the care and also to provide things that cannot be provided locally, such as access to clinical trials. The antifibrotics do not cure this disease, they slow the disease. And the only way we're going to move to cure is to get more patients enrolled in clinical trials.

But the patient needs to have practitioners, providers close to home for when they have complications, or if they get an infection or whatever. And we can communicate so well. Many electronic records are connected, we're connected with all of the referral docs that are in this neck of the woods, and I can see the notes from the primary care offices, the local pulmonologist offices, test results, and

always just pick up the phone or email or message.

So, it is going to take a team approach to both get at an earlier diagnosis but also manage these patients.

Dr. Schulman:
Thanks. Tim, any closing thoughts?

Dr. Hernandez:
I'd like to underscore those thoughts and say that I agree. Many of us, especially maybe even our younger colleagues will like to rely on the electronic record for communication or maybe an email. But I think what Andy said is really important. If things aren't working properly for the patient, or if for whatever reason we're not sure, as a primary care physician, what we should do next; I think it's important to reach out and call your colleague and ask them what they think you should do next or ask them to see the patient. I always prefer to communicate directly if, at the very least, by phone with my colleagues. And in that way, I think we can ensure better care for the patients.

Dr. Schulman:
With all of these key takeaways in mind, I do want to thank my guests, Dr. Andrew Limper, Dr. Tim Hernandez, for joining me to discuss pulmonary care and idiopathic pulmonary fibrosis. Andy, Tim, it was great having you both on the program.

Dr. Limper:
It was great to be here.

Dr. Hernandez:
Thank you. It was a pleasure being here.

Dr. Schulman:
For those listening, I hope you'll consider following CHEST on one of our multiple social media platforms including Twitter, Instagram, and Facebook, and please consider signing up to receive updates as new resources become available to aid in the recognition and timely diagnosis of interstitial lung diseases like pulmonary fibrosis. For those interested in learning more about our toolkit, which is designed to aid clinicians in the recognition and timely diagnosis of interstitial lung diseases, please use the link in the podcast description to sign up for additional updates.

Announcer Close:
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