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Coping With PAH in Rarer Forms of Connective Tissue Disease

Dr. McLaughlin:
Hello, today we're going to talk about an underrepresented group of patients in clinical trials, those with rheumatologic diseases. I'm Vallerie McLaughlin from the University of Michigan. And I'm joined by my friends and colleagues, Dr. Sudarshan Rajagopal from Duke, Ioana Preston from Tufts, and Rich Krasuski from Duke. Welcome, everyone.

So Rich, let's start with the group of rheumatologic diseases that we know the most about those patients with scleroderma. Do you want to summarize that group for us briefly?

Dr. Krasuski:
Sure. So Vall, it's a highly prevalent group of pulmonary hypertension in their disease process. Their outcomes are pretty bad if you don't treat them adequately. I think their mortality is considerably higher if they develop pulmonary hypertension and we have a decent amount of literature now, suggesting that aggressive treatment, makes a great impact in terms of their clinical outcomes. And not only that, but we actually have evidence that you can detect the disease earlier if you screen for it. And ideally, you don't want to wait for those symptoms to develop, but you have to look and for instance, do echocardiography or some sort of formal screening on a yearly basis because this is such a prevalent problem and such a bad impact in terms of how patients will do once it develops. And you don't want to wait for it to get bad, before making the diagnosis.

Dr. McLaughlin:
Right, absolutely. And so, I guess we can justify doing that in the scleroderma population, as you said the disease prevalence is pretty high, but a lot of the other types of connective tissue diseases, rheumatologic diseases, they can get pulmonary hypertension, but the prevalence isn't high enough to warrant screening, perhaps with the exception of the mixed connective tissue disease with scleroderma features we screen. But what about the others? What about the lupus patients, the rheumatoid arthritis patients? Sudar, what do we advise in those patients?

Dr. Rajagopal:
Sure. So, as you mentioned, any patient with connective tissue disease can develop pulmonary or arterial hypertension, they're at increased risk for it. But these other populations outside of scleroderma or mixed connective tissue disease with scleroderma features, they're not as at high a risk. So, we don't recommend screening those patients, but it does mean that we have to be aware and we have to have awareness of the disease, because they can develop it.

So, people seeing these patients, whether it's their primary care doctor or their rheumatologist, they have to be cognizant of the fact that these patients can develop PH. So, they need to ask them about their functional status, their exercise tolerance, because if they do develop any symptoms, then they should have an echocardiogram to assess for possible pulmonary hypertension.

Dr. McLaughlin:
Sure, an echo to assess for possible pulmonary hypertension and probably PFTs to assess for ILD. Many of these groups of patients are at risk for interstitial lung diseases as well. Correct Ioana?

Dr. Preston:

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Absolutely. The rheumatoid patients can develop ILD, they can develop pulmonary nodules, pleural effusion. So, there are other pulmonary complications and not to speak about lupus patients who can have acute alveolitis and all sorts of other acute and chronic pulmonary complications. The rheumatoid patients, the pure rheumatoid patients, the prevalence of PAH is up to 3% in some reports. So, it is still much higher than in the general population. So, although we do not recommend annual screening, like in scleroderma, we do have to have a high index of suspicion, should they develop more shortness of breath.

Dr. McLaughlin:
Yeah, absolutely. Now, some of these groups act a little differently too. We approach their PH a little bit differently, like the lupus patients for example. Do you want to expound on that Sudar?

Dr. Rajagopal:
Sure. So, lupus patients in general, tend to respond very well to PH-specific therapies. And this has been seen in the clinical trials and also when a patient has a lupus flare, they can also have a PAH flare in that setting. And their PAH can improve with treatment of their lupus flare with steroids or other immunosuppressants. So in general, that's a population which tends to do better with PAH, as opposed to say the scleroderma patients.

Dr. McLaughlin:
And they tend to be younger as well with less comorbidity. So, treatment is a little bit more straightforward.

Dr. Rajagopal:
That's true.

Dr. McLaughlin:
In them. Ioana what about some of the more complex patients, like the rheumatoid patient that are also at risk for fibrosis or the mixed connective tissue disease patients? How do you approach them?

Dr. Preston:
So, they need a very comprehensive assessment and most likely multidisciplinary assessment from the rheumatologist, from the pulmonologist, from the cardiologist to understand what's the main cause of their limitation. Is it the interstitial lung disease that may be present? Is it myopathy like these patients may develop or pulmonary vascular disease? Also, patients with lupus, can have hypercoagulable state and they develop, chronic thromboembolic pulmonary hypertension. So, which type of pulmonary vascular disease, whether it's group one, group four or group three or a component of each is very important to sort out in order to decide on the type of the treatment and care.

Dr. McLaughlin:
Yeah, absolutely. I agree. We also can see this in patients with, you know, the more rare rheumatologic diseases, you know, just isolated Sjogren, polymyositis, dermatomyositis.

So Rich, as practicing physician, I think let's talk about how to approach this. You know, the main point of our conversation today is many of our clinical trials, have not included a large number of these patients. And perhaps with the exception of the lupus patients who occasionally have a little bit more inflammation, maybe a little more responsive to steroids or immunosuppressives, the pathophysiology is pretty similar. Tell me about your approach in clinical practice, your approach to treatment of the Sjogren patients with group one PAH or some of these other more rare diseases.

Dr. Krasuski:
Sure. I think the real burden here is to make sure make the diagnosis properly. I think heart catheterization is absolutely essential in these patients to make sure that you have an increased pulmonary vascular resistance. Again, I also think vasodilator challenges and things like that are very important. Some of these disease processes, can involve the left ventricle. So, ensuring that you're identifying the patient properly.

I think a lot of what we've learned from other disease processes like scleroderma is directly applicable. I think that these patients do definitely benefit from advanced medical therapies. And so I treat them very similarly, but I think as has been mentioned, that this is a group of patients you want to make sure, you know what's going on physiologically, that you're following them very closely afterwards and when you initiate therapy, ensuring that they come back for regular follow up is very important to make sure that you're impacting positively on their disease process.

Dr. McLaughlin:
Yeah. Do either of you guys want to add anything to that?

Dr. Rajagopal:

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No, I agree. I think for the key thing like Rich mentioned is getting the diagnosis. And once you make sure that there aren't large contributors to lung disease or other processes, you can treat them like other group one patients and you can treat them aggressively and have good outcomes.

Dr. McLaughlin:
Yeah. So, I think if I could summarize, prevalence is not so high as it is in scleroderma to routinely screen. But the frontline providers who see these patients, the primary care docs, the rheumatologists, they need to be cognizant of changes in exercise tolerance, changes in dyspnea and that should trigger an evaluation for PAH. Then they should go through the usual diagnostic evaluation to look for the other comorbidities that can contribute to PAH. And if they're determined to have group one disease, we really treat them very much the way we treat the rest of the group one patients. Is that the consensus basically?

Dr. Preston:
Absolutely.

Dr. Krasuski:
Absolutely, I agree.

Dr. McLaughlin:
Okay, yeah. Well, I think it's an important population. We see them all the time. So Sudar, Ioana, Rich, thank you for joining me. And thank you for joining us on this roundtable discussion on patients with connective tissue disease and PAH.

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