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Why Should We Screen for PAH in Systemic Sclerosis Patients?

Announcer:
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Dr. Khanna:
So hello everyone, my name is Dinesh Khanna. I'm a Rheumatologist and Director of the Scleroderma Program at University of Michigan. I'll be talking about why should we screen for pulmonary arterial hypertension in patients with systemic sclerosis or scleroderma?

We know that pulmonary arterial hypertension is common in patients with systemic sclerosis. In fact, 5 to 12% of the patients will develop pulmonary arterial hypertension during the course of their life. It's a serious complication and it is a leading cause of mortality in systemic sclerosis. Unfortunately, many patients present with advanced symptoms, significant functional impairment, and severe hemodynamics. at diagnosis.

WHO, or the World Health Organization, defines screening as a presumptive identification of unrecognized disease in an asymptomatic patient population. Data that I will share with you shows that timely diagnosis via systematic screening of asymptomatic patients may help support early intervention efforts to improve outcomes in pulmonary arterial hypertension associated with systemic sclerosis. DETECT algorithm, which is a multimodal multi-dimensional, non-invasive tool is recommended both by the 6th World Symposium on Pulmonary Hypertension, and the 2022 European Society of Cardiology and Respiratory Society guidelines. Whatever methodology that you use for screening, it is critical to highlight that both organizations recommend annual screening for pulmonary hypertension in patients with systemic sclerosis.

So I want to dwell a little bit more into the screening. You know, I get this question all the time, a patient comes to you with shortness of breath, dyspnea on exertion, getting disliked when they climb stairs, syncopal episodes, cough, all those are symptoms that should lead to your diagnostic workup. In fact, based on the WHO definition, screening is not the same as early diagnosis. Screening invites people such as scleroderma patients who do not have symptoms suggestive of pulmonary hypertension, to undergo testing. Early diagnosis is intended to detect conditions in patients who are symptomatic.

The purpose of screening is to identify people who are at high risk of health problems such as pulmonary hypertension so that an earlier treatment or intervention can be offered and lead to better outcomes in this patient.

So this is really depicted in this slide by Marc Humbert and colleagues. These are our colleagues in France, where they had two different patient populations. On the left are patients who came in routine practice, 16 patients, they were symptomatic, as you can see majority of the patients were in functional class 3 and 4. And in the middle are patients who were systematically screened with annual echocardiogram that was done at each center. And based on a predefined algorithm, they were referred for right heart cath. And as you see in the middle slide, that 50% of the patients came in functional class 1 and 2. The question becomes, does it really matter? And in fact, it does. On the right part of the slide, you see the 8-year survival after the diagnosis of scleroderma or pulmonary arterial hypertension, in orange are the patients who came as part of screening algorithm, and you see a much better survival compared to the

patients who came through routine practice. All these patients were treated with pulmonary arterial hypertension drugs with approximately 20% on the combined pulmonary hypertension medications.

Now, the 2022 ESC/ERS guidance on pulmonary hypertension has made some very important recommendations, and I would like to highlight a few of them. The first one, every patient with systemic sclerosis should be screened on an annual basis. Number two, echocardiogram by itself is not an appropriate screening tool. You have to really think about a multimodality tool, such as DETECT, or a combination of echocardiogram biomarkers and pulmonary function test. They also recommended that in patients with systemic sclerosis, who are short of breath but it is undiagnosed, it is unexplained why they're having these symptoms, a right heart cath is recommended to exclude pulmonary arterial hypertension. And finally, in patients who are symptomatic, so we talked about screening versus diagnosis, CPET, echocardiographic with exercise, or cardiac MRI may be considered to aid the decision to perform right heart cath.

So in summary, I want to again highlight that pulmonary arterial hypertension is common; 5 to 12% of scleroderma patients will develop pulmonary hypertension during the course of their lifetime, and it is a leading cause of mortality in patients with PAH-CTD. Many patients with scleroderma unfortunately experienced long delays in diagnosis of PAH and worsening prognosis. There are certain demographics and clinical features that are associated with increased risk of pulmonary hypertension in systemic sclerosis. Proactive screening can help detect pulmonary arterial hypertension at early disease stages and improve patient outcomes. As I highlighted, the DETECT algorithm has been endorsed both by the 6th World Symposium on Pulmonary Hypertension, and also by 2022 ESC/ERS guidelines. And due to the higher levels of misdiagnosis using echo alone, the guidelines now recommend use of DETECT as an option to screen for pulmonary hypertension associated with systemic sclerosis.

Thank you very much for attending this session.

Announcer:
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