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Time needed to complete: 1h 51m

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How Do We Screen For PAH in Systemic Sclerosis Patients?

Announcer:

Welcome to CME on ReachMD. This episode is part of our MinuteCME curriculum.

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Dr. Khanna:

Hi everyone, my name is Dinesh Khanna. I'm a Rheumatologist and Director of the Scleroderma Program at University of Michigan. Today, I will briefly talk about how to screen for pulmonary arterial hypertension in a systemic sclerosis patient.

So let's talk about the importance of early detection of pulmonary hypertension in scleroderma. Pulmonary arterial hypertension is common and serious complication. It affects 5 to 12% of patients during lifetime. And screening is really focused on detecting patients who are asymptomatic, such as doing a mammogram in a patient who doesn't have symptoms or signs of breast cancer. Due to higher level of misdiagnosis using echocardiography alone, the 2022 European Society of Cardiology and Respiratory Society guidelines now recommend using DETECT as an option to screen for pulmonary hypertension in patients with scleroderma. DETECT is a multi-dimensional tool, and has also been endorsed by the 6th World Symposium on Pulmonary Hypertension. And I must emphasize regardless of the method of screening that you use, both organizations recommend annual screening for pulmonary hypertension in this patient population.

So here's the data on the echocardiography that refutes that echocardiogram is the best screening tool. This data is from our DETECT study, where we took patients with systemic sclerosis, a diffusion capacity of less than 60%, and a disease duration of greater than 3 years. In this patient population, the patients had echocardiogram, other parameters, and right heart cath on the same day. What you see out here is that if you take a TR velocity of less than 2.5 meter per second, or an estimated RVSP of less than 30 millimeter of mercury, 20% of these patients indeed had pulmonary arterial hypertension. If you increase the cut-off to 36 millimeter of mercury, 1 in 3 patients had pulmonary arterial hypertension.

A multi-dimensional or a multimodal approach for pulmonary arterial hypertension is warranted. On the left is the DETECT algorithm that I will detail, and on the right is an Australian Scleroderma Interest Group, or ASIG, that again combines a forced vital capacity and a DLCO and/or NT-proBNP that will determine the patient to get other modalities including echocardiogram and then a right heart catheterization.

So, one thing I can emphasize with the slide with the 2022 ESC/ERS recommendations is a multimodal approach, whether you use the DETECT algorithm or whether you use ASIG as part of your annual screening in patients with systemic sclerosis.

So DETECT is a 2-step process. Step 1 are the non-echocardiographic variables that are listed out here. And they really include physiological tests such as pulmonary function test, physical examination on presence of telangiectasia, auto antibody, especially anticentromere antibody, two biomarkers, a NT-proBNP and uric acid, and right axis deviation. And the step 2 is echocardiographic variables that include right atrial area and tricuspid velocity. DETECT is available as an app. I download it on my iPhone, and I do it every day after I've seen the patient and when the results are available. The step 1, as we discussed, you can enter the data here, it validates the data. And in the step 1 calculation on the right side of the slide, it tells you whether an echocardiogram is needed for this





year or not as part of the screening process.

If echocardiogram is indeed needed, you go to the step 2, where you enter your TR velocity along with the right atrial area, and it determines whether the patient should be referred for a right heart cath. What is the advantage of doing a DETECT algorithm? It takes away our clinical bias out of the picture. It is a standardized way to screen a patient on an annual basis who has a diagnosis or classification of systemic sclerosis. The value of DETECT is that it is highly sensitive. And this is a data internal validation in the DETECT cohort. And the thing that I want you to really pay attention are the blue bars. If you use the DETECT algorithm, we missed 4% of the patients who had pulmonary arterial hypertension. When you look at the 2009 ESC/ERS guidelines on the right part and the blue bar, you missed 29%, or 1 in 3 patients. So that always has been my teaching. Echocardiogram. Whatever parameters you use for early screening and detection will miss 1 in 3 patients who have pulmonary arterial hypertension.

And we validated this data using University of Michigan cohort. Again, the DETECT was highly sensitive. We did not miss any patients with pulmonary arterial hypertension; whereas when we used echocardiography with the 2015 ESC/ERS recommendations, we missed anywhere from 20 to 33% of the patients who had pulmonary arterial hypertension.

A final thought. I think it's important to have these patients seen in referral centers to confirm pulmonary hypertension, and the guidelines that will be discussed quite a bit and the treatment management in patients without cardiopulmonary comorbidities, and patients with cardiopulmonary comorbidities and how the risk stratification should be done, including upfront ERA and PDE5 combination, or escalation of therapy.

Thank you for your attention today.

Announcer:

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