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Screening the CTD Patient For Development of PAH - Why Must We Do This?

Announcer:
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Dr. Krasuski:
Screening the CTD patient for development of pulmonary arterial hypertension. Why must we do this? Active screening identifies patients earlier, and earlier detection equals better survival. So, this is data from the French registry. And this was a program looking at systemic PH detection versus routine clinical practice. As you can see in this particular study, they identified patients earlier, provided therapy for those patients earlier, and there was a significant improvement in survival that was seen. So again, identifying the disease at its earlier stages and getting that therapy on board can potentially make a huge difference in patient survival.

Screening of high-risk patient populations for pulmonary arterial hypertension. Annual screening is recommended in asymptomatic patients with systemic sclerosis spectrum diseases, and should include a two-step approach. In this case, you look for the presence of telangiectasia. You measure the anti-centromere antibodies. You look at the pulmonary function tests and DLco measurements. You look at the electrocardiogram and cardiac biomarkers, such as NT-proBNP and uric acid. And this is the initial stage. After this, echocardiography can be performed. And then there is consideration of right heart catheterization in patients with abnormal findings. Screening should be part of a scientific protocol or registry whenever possible. Patients with systemic sclerosis and other connective tissue diseases with clinical signs and symptoms of PH should be evaluated by right heart catheterization. Scleroderma, or systemic sclerosis, and scleroderma spectrum diseases, again, annual screening is recommended, and screening for scleroderma spectrum with uncorrected DLco of less than 80% of predicted. Screening tools should include the DETECT algorithm, the 2015 ESC/ERS recommendations for echo or forced vital capacity over DLco calculation with a ratio greater than 1.6, assuming none-to-mild interstitial lung disease, and greater than 2-fold of the upper limits of normal elevation in the NT-proBNP. If any of these tests are positive, these patients should be referred for right heart catheterization. When the uncorrected DLco is greater than 80% of predicted, echo screening may be used.

So, the bottom line is if your patient has systemic sclerosis, screen them annually for development of PH. Again, how often? This should be done yearly in a patient with systemic sclerosis or mixed connective tissue disease with scleroderma features. These patients should be screened with tests, including echos, PFTs, NT-proBNP, and then the use of the DETECT protocol if the DLco is less than 80%. If these tests are abnormal, the patient should get a right heart catheterization. Other testing includes echocardiography, especially if there's new symptoms, NT-proBNP, pulmonary functions with DLco, and again, DETECT algorithm. And finally, remember that if the patient presents with other connected tissue diseases, it's not currently recommended to screen these patients due to the low prevalence of pulmonary hypertension.

Announcer:
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