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ReachMD
www.reachmd.com
info@reachmd.com
(866) 423-7849

How Can We Make It Easier To Develop That “Index of Suspicion” for PAH?

Announcer:
Welcome to CME on ReachMD. This episode is part of our MinuteCME curriculum and is titled “How Can We Make It Easier To Develop That “Index of Suspicion” for PAH?”.

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Dr. Preston:
So how can we make it easier for our general practitioners and community physicians and healthcare providers to be aware and develop that index of suspicion for PAH? We will talk about the roles of the community physician and the PAH center in reaching the correct diagnosis and promoting referral in a timely manner.

So let's build a clinical suspicion for PAH. The symptoms are very non-specific. They're short of breath, usually with exertion. They have weakness. Sometimes they have chest pain and lightheadedness, but only when the disease is very advanced. And less frequent, they have a cough. In more advanced disease, one may also observe signs of right heart side failure, the swelling of the legs, ascites, abdominal distension. A few patients have hemoptysis, and very rare, they have the hoarseness, or Ortner's syndrome, as well as arrhythmias. The physical findings are also not very overt; you have to look for them. In the early stages, there's only a loud second sound of the heart, so a P2 component that's slightly louder, and that's it. Sometimes, in more advanced, you have a right ventricular lift, jugular venous distension, hepatojugular reflux, ascites, hepatomegaly, sometimes splenomegaly, definitely edema in more advanced condition, and murmurs of tricuspid regurgitation. In some patients, you can hear an S3 gallop that's right sided. So, if they don't have overt signs of right heart failure, the physical exam is very subtle, so you have to think about pulmonary hypertension when you put the stethoscope.

In the current era, what clinical data are needed for assessment of a patient with suspected PAH? So let's look at the battery of tests that patients suspected for PAH need to undergo. On the left-hand side, pulmonary function tests, biochemical markers, and clinical assessment, as well as the non-invasive exercise tests, such as six-minute walk distance, and the echocardiographic evaluation. All these can and should be done at the community level: the primary care practitioner or the primary rheumatologist or the primary pulmonologist or cardiologist who suspects pulmonary hypertension. All these uninvasive tests should be done initially. Now, the more complex tests that require more expertise, these are the VQ scan and the hemodynamic evaluations, the right heart catheterization with or without exercise. These can be completed at the PAH center. So, where the initial testing, including all non-invasive tests, can be performed very well close to the patient's home at their physicians, the finish of the workup and confirmation of the PAH diagnosis should be done at the PAH specialty center. That's how a strong collaboration between the community healthcare providers and PAH center can be developed to help these patients be diagnosed early and properly.

So what information should we get from PFTs? Always obtain full PFTs in the PAH workup, not only spirometry. We need lung volumes and DLco because in certain diseases, DLco is associated with the presence of pulmonary hypertension, and I'm talking about patients with scleroderma. The presence of markedly abnormal PFTs with severe obstruction and/or restriction then prompts us to think about pulmonary hypertension associated with lung disorders, with hypoxemia, and this is a group three type of pulmonary hypertension,

which has a different treatment.

So, the only serologic biomarker for pulmonary hypertension is BNP or it's pro molecule NT-proBNP, and they're very well established in the PAH world. They are a sign that the right ventricle is overworked and overstretched, and it has been associated with prognosis, and it's part of the parameters that we assess the risk scores. So, it's a very simple test, a lot of hospitals use it as a point of care, and we strongly recommend in the workup of patients with pulmonary hypertension. The guidelines that show the importance of NT-proBNP or BNP are showed below and highlight the importance and its role in assessing the prognosis of these patients.

Announcer:
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