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If Unexplained Dyspnea Is Not Fully Explored, What Are the Implications…Especially if It Is PH?

Announcer:
Welcome to CME on ReachMD. This episode is part of our MinuteCME curriculum and is titled “If Unexplained Dyspnea Is Not Fully Explored, What Are the Implications…Especially if It Is PH?”.

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Dr. Preston:
So, if unexplained dyspnea is not fully explored, what are the implications? Especially if it is Pulmonary Hypertension.

So let's think about common pitfalls in PAH patient care. It is easy to dismiss dyspnea as something else. And many of my patients were referred to me after being told that they're anxious and depressed, and that's why they feel short breath, especially the young ones. So, there is lack of screening of at risk populations. We have to recognize these at risk patients, who have underlying conditions such as Scleroderma, or Mixed connective tissue disease, or those who have a familial history of PAH. Another pitfall that is very common is relying on only systolic PA pressure estimates of measurements from the echocardiogram imaging instead of obtaining the evidence, for structural signs of RV dysfunction in combination with the right heart catheterization. So, a common pitfall is to focus on the systemic pressures, but not to look at the RV function on echo. The most common and the most dangerous I should say, pitfall is not confirming the diagnosis with the right heart catheterization. Because certain medications are indicated in certain types of PAH, and are not indicated in other types. So, we really need to sort out what the group of pulmonary hypertension we're dealing with. And the last one, that comes to my mind is lack of risk assessment leading to delay incorrect therapy and escalation. And we know, that for the past 20 years the average time from an onset of symptom to diagnosis of PAH is over two years.

So we have to reduce that, and be proactive in working it out. And why is that? Two years is a long time, because PAH is a Progressive Structural Disease. And if you look at this very famous diagram. The top graph shows pathology of an early pulmonary vessel in early PAH phase, and then in the middle as more symptomatic, more severe remodeling. And in the last phase, actually there are lungs patients who underwent lung transplant, and shows very severe pulmonary vasculopathy. And these changes, are accompanied by hemodynamic effects, where in the facing of an increased vascular resistance in the pulmonary vasculature the PA pressure increases to overcome this resistance. And in the beginning cardiac output, the force with which the right ventricle pushes blood through the lungs is maintained by the RV being hypertrophied, but in time it if the compensates and cardiac output drops, but also the pulmonary artery pressure drops because the RV cannot mount that much of a pressure and patients develop right heart failure. And this is their most common cause of death.

So, what factors influence time to definitive PAH diagnosis? Some of the, you know, factors that we should take into consideration are time between patient reported onset of symptoms and the definitive PAH diagnosis. This is consistently delayed and we have to work on that. The delay study out of Australia retrospectively examined factors contributing to diagnostic delays and the time for definitive PAH diagnosis. So, in 32 patients and the majority were females they were reporting exertional dyspnea. The meantime from symptom onset to diagnosis was 47 months, on an average. Patients reported to over five general practitioner visits. And an average of three specialists were reviewed before being seen at the PAH center. So there was a lot of back and forth for these patients. So that's where we have to

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get better. Factors significantly associated with delay diagnosis advanced age, number of GP visits, the heart rate and the systolic blood pressure, which is very interesting. And this diagram shows the length of the journey of our patients from onset of symptoms to the final diagnosis.

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