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CTD-Associated PAH – Perspectives for Rheumatologists and Pulmonologists: Working Together To Build Clinical Suspicion

**Announcer:**
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**Dr. Krasuksi:**
Connective tissue disease-associated pulmonary arterial hypertension: perspectives for rheumatologists and pulmonologists working together to build clinical suspicion. So let's briefly discuss the epidemiology of systemic sclerosis and pulmonary arterial hypertension. Connective tissue diseases are commonly associated with PAH. Systemic sclerosis is the most frequent connective tissue disease that's complicated by PAH. About 8 to 12% of all of these patients develop PAH and this accounts for 75% of CTD PAH cases. It's the leading cause of death in systemic sclerosis, and it's associated with a worse prognosis than patients with idiopathic disease. PAH can be detected in 1 to 5% of patients with SLE and 3 to 4% of patients with mixed connective tissue disease. Data on the prevalence of PAH in connective tissue diseases other than systemic sclerosis are less reliable, owing to the lack of screening. And it's not surprising, since screening is only recommended at this time for systemic sclerosis and also the lack of right heart catheterization in these patients.

So how do we build a clinical suspicion of pulmonary arterial hypertension? First of all, we have general symptoms. These are non-specific things such as dyspnea, weakness chest pain, lightheadedness, or syncope or cough which is in fact, a little less frequent. Signs and symptoms in advanced disease may include progressive right heart failure, edema, ascites, and abdominal distension, hemoptysis, Ortner's syndrome, which is hoarseness related to unilateral vocal cord paralysis, is in fact very rare but occasionally can be seen, and arrhythmias. On physical examination, we look for an augmented second heart sound, a large P2 component, a right ventricular lift may be present, jugular venous distension is often present, hepatojugular reflux. Which remember you push on the liver, you wait about two or three seconds, if it remains elevated, that's elevated hepatojugular reflux and jugular venous distension. Ascites, hepatomegaly, and/or splenomegaly, edema, tricuspid regurgitation, or pulmonary regurgitant murmurs, and finally a right sided S3 gallop.

Successful identification and management of pulmonary arterial hypertension requires collaboration. There are many specialists that are involved in patients with connective tissue disease care. These can include rheumatologists and community specialists that can include internal medicine or primary care, cardiologists, community and PH specialty care, and also pulmonologists, community and PH specialty care. And finally, at the center of all of this hub is the pulmonary hypertension center. And this communication back and forth is vitally important to make this all work.

The community physician in the PH center: disease identification and treatment must be a collaborative approach. For community generalists and specialists, they are involved in the initial assessment. They're identifying at-risk patient populations, they are focusing with the primary provider for care, they're identifying early disease, they're screening at risk patients, such as patients with systemic sclerosis, and they're providing routine medical care after diagnosis. But they're also collaborating, communicating with the PH specialty center where the confirmation of diagnosis occurs when there is uncertainty. There they have PH specialty physicians. They have

advanced diagnostics available. They're experienced in advanced therapies, including prostacyclins and IP receptor agonists. They're involved in trials of PH therapy. They can provide extra services, such as lung transplantation assessment. Presence of support groups are also at these centers. Nursing expertise and support teams are available. And finally, there's advanced patient education programs. So this collaboration and communication back and forth is critically important for success in this.

**Announcer:**
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