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Case: Using the 2022 ESC/ERS Treatment Algorithm in Older Male Patient with Cardiopulmonary Comorbidities

Announcer:

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Dr. Channick:

Hello, today I'll present a case study. Today we have a 66-year-old man with a history of methamphetamine use, none in 15 years, hypertension, diabetes, who noted the onset of dyspnea on exertion 7 months ago. He actually had to stop working, and he's currently getting short of breath and fatigued, even with just working around his house. He was a past smoker, a pack a day for 30 years and he quit 10 years ago. There were no other risk factors identified for pulmonary hypertension. On examination, the patient had a normal blood pressure and heart rate, oxygen saturation was borderline 91% on room air, and he was obese with a BMI of 35. The remainder of his exam showed some crackles at the lung bases, regular heart, but a loud second heart sound and a 2/6 murmur at the lower left sternal border systolic. He had trace pedal edema.

Initial workup for his symptoms included a 6-minute walk test, which was 320 meters, a ventilation perfusion scan that was normal, and pulmonary function tests that actually showed normal spirometry, but a reduced diffusing capacity. And he had an NT-proBNP of 500. On echocardiogram, the patient had significant moderate to severe right ventricular enlargement, and decreased right ventricular function with a TAPSE index at 1.6 and a right ventricular systolic pressure estimated at 64 plus right atrial pressure, so significantly abnormal echo suggesting pulmonary hypertension.

He then went on to a right heart catheterization, which is shown here. His right atrial pressure was 8, pulmonary artery pressure was 70/45 with a mean of 53, a wedge pressure of 8, and the cardiac output a 4.5 liters per minute, which came out to a cardiac index of 2.8 with a calculated pulmonary vascular resistance of 10 Wood units. He had a minimal response to acute testing with inhaled nitric oxide with a PVR dropping to 8 and PA pressure dropping from 53 to 48. So we'd call that a pretty unremarkable vasoreactivity test. So then going over the hemodynamics, I think you'll agree, this is a patient who has significant pre-capillary pulmonary hypertension, adequate cardiac output, and a very high pulmonary vascular resistance.

So if I was to ask you, what risk category do you put this patient in? Functional class III, that's the walk distance, NT-proBNP levels, and you can see the right heart catheterization results. This patient, I think, would fall strongly into intermediate, right in the intermediate category. With that in mind then, we have, again to summarize, a patient who has some cardiac issues, but also a history of methamphetamine use, a longtime smoker as well, who now has significant pre-capillary pulmonary arterial hypertension, intermediate-risk category, what do you do? What's your initial therapy for this patient in 2023? ERA monotherapy? Monotherapy with either a PDE5 inhibitor or soluble guanylate cyclase? A prostacyclin pathway drug, either an analogue or receptor agonist monotherapy? Or combination therapy? What would you say?

Now, the challenge is that, and this is - we're going to talk about the ERS/ESC guidelines, which recently came out, is that they really separated patients with comorbidities from those without comorbidities in the initial treatment algorithm. Typically, when we've talked

about initial treatment for pulmonary arterial hypertension in the intermediate risk, we're usually talking about dual oral therapy. And there's a wealth of data supporting that approach.

The recent guidelines suggested that in patients who had comorbidities, either cardiovascular or cardiopulmonary comorbidities, those patients might be treated differently. And in fact, the suggestion of using monotherapy in those patients with cardiopulmonary or cardiovascular comorbidities. So what do we do? What do we do for a patient like this who has those comorbidities but who has very significant pulmonary hypertension? I think my message here and my teaching point, pretty strongly, is this recommendation that you see here should did not necessarily apply to all patients. The term comorbidities is a very general term. And we really need to look at each patient individually. A patient who has severe pulmonary arterial hypertension who happens to have, let's say, well-controlled diabetes and hypertension, that patient should likely be treated just like any other patient with PAH who had no comorbidities. At the other end of the spectrum, a patient with clear superimposed heart failure preserved ejection fraction, even when they have pre-capillary pulmonary hypertension, elderly patients, patients who may have some underlying lung disease, those patients probably should be treated more cautiously and consideration as this algorithm shows for monotherapy. So I think that's really the important teaching point, is that a patient like I just presented to you should be treated likely aggressively with oral combination therapy, even though they may have some comorbidities.

And that's sort of summarized here, group 1 patient, intermediate risk, poor RV function, cardiovascular comorbidities are not really prominent, and probably not driving this patient's disease. Therefore, I would treat this patient the same as other PAH patients who had no comorbidities with combination therapy.

Thank you.

Announcer:
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