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Case Study: Group 3 PH Pulmonary Hypertension Due to Chronic Lung Disease

Announcer:
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Dr. Boucekkine:
So, our case is that of a 67-year-old male who was referred to our PH clinic after an echo revealed an elevated pulmonary artery systolic pressure of 80. Backstory is that over 10 years ago, he had developed dyspnea on exertion and a chronic nonproductive cough. He was evaluated in pulmonary clinic. A CT chest was notable for architectural distortion, reticulation, bronchiectasis. He was sent for a surgical lung biopsy, and wedge biopsies revealed cellular nonspecific interstitial pneumonia. He was started on azathioprine and was actually stable for many years, both from a symptomatic standpoint and with serial pulmonary function tests. Unfortunately, azathioprine had to be discontinued due to elevated serum amylase and lipase; and several months later, he started to develop increased dyspnea on exertion and increased fatigue.

So it was at this point that repeat CT chest was obtained. Let you take a look through it. But basically, you can see some evidence of architectural distortion bilaterally, some traction bronchiectasis, reticulations with a little bit of subpleural sparing, suggestive of bilateral pulmonary fibrosis with a lower lung predominance.

He had repeat pulmonary function tests done, which showed a reduction in his FVC, FEV1, and diffusion impairment as evidenced by the quite low DLCO. Unfortunately, lung volumes were not obtained but based on these numbers, we can ascertain that he likely had a restrictive ventilatory defect which goes along with what we're seeing on the CT. One year prior, his FEV1 and DLCO were slightly better, now slightly worse.

He had an echo performed, and the second video will start shortly. But this echo showed moderate enlargement of his RV as well as a moderate reduction in systolic function. His RV to LV ratio is calculated is greater than 1, TAPSE was 16 mm.

There was also some systolic and diastolic septal flattening, suggestive of RV pressure and volume overload. Interestingly, he had an echo 4 years prior which showed normal RV systolic function and normal RV size as well.

He was then referred for a right heart cath. I have the numbers up here for you to see. But his PA pressures were elevated, systolic was 59 with a mean PA of 38, wedge was 9, cardiac output of 5.5, and his PVR was elevated at 5.27. INO was administered without any really significant hemodynamic response. His mean PA didn't really budge; it was at 36. And his wedge didn't budge either. So this was suggestive of a precapillary pulmonary hypertension.

So it was at this point that the patient was started on subcutaneous treprostinil. And on the table on the middle column, you'll see his repeat right heart cath about 8 months later on subcutaneous treprostinil. And there is some improvement in his PA pressures. His PA systolic went down to 48. His mean went down 27. His PVR improved to 3.83. And also clinically and symptomatically, he reported improved dyspnea and improved fatigue. Unfortunately had multiple side effects from the medication, including a lot of site infections.

And so, unfortunately, at that point, they transitioned to oral treprostinil.

He continued to develop progression in his symptoms and so he was ultimately referred for a lung transplant. His repeat right heart cath shortly before his transplant, and this was purely on oral treprostinil, largely stable, maybe slight worsening in his mean PA pressures and in his PVR; and about 8 months after referral, he actually underwent a single left orthotopic lung transplant and has been doing well from respiratory standpoint.

Announcer:
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