"I can't catch my breath!"

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A 39-year-old woman presents with a seven-month history of exertional dyspnea. The dyspnea has now progressed to a level where she experienced it on moderate activity (Medical Research Council rating III). She is a non-smoker, is not taking any medications, and denies the use of recreational drugs. She is married with two children, holds an office job in an accounting firm, and denies any environmental exposures.

Her physical exam reveals:

- Blood pressure: 118/68 mmHg
- Heart rate: 78 beats per minute
- No evidence of respiratory distress at rest
- Oxygen saturation: 96% on room air
- Normal breath sounds
- Absence of any clubbing
- A loud S_2, with no other findings of heart failure

The patient has an athletic build, and the remainder of the physical exam is essentially normal.

Her blood work, chest X-ray, electrocardiogram, and pulmonary function tests (PFTs) are unremarkable.

Her cardiopulmonary exercise test (CPET) is shown in Figure 1.

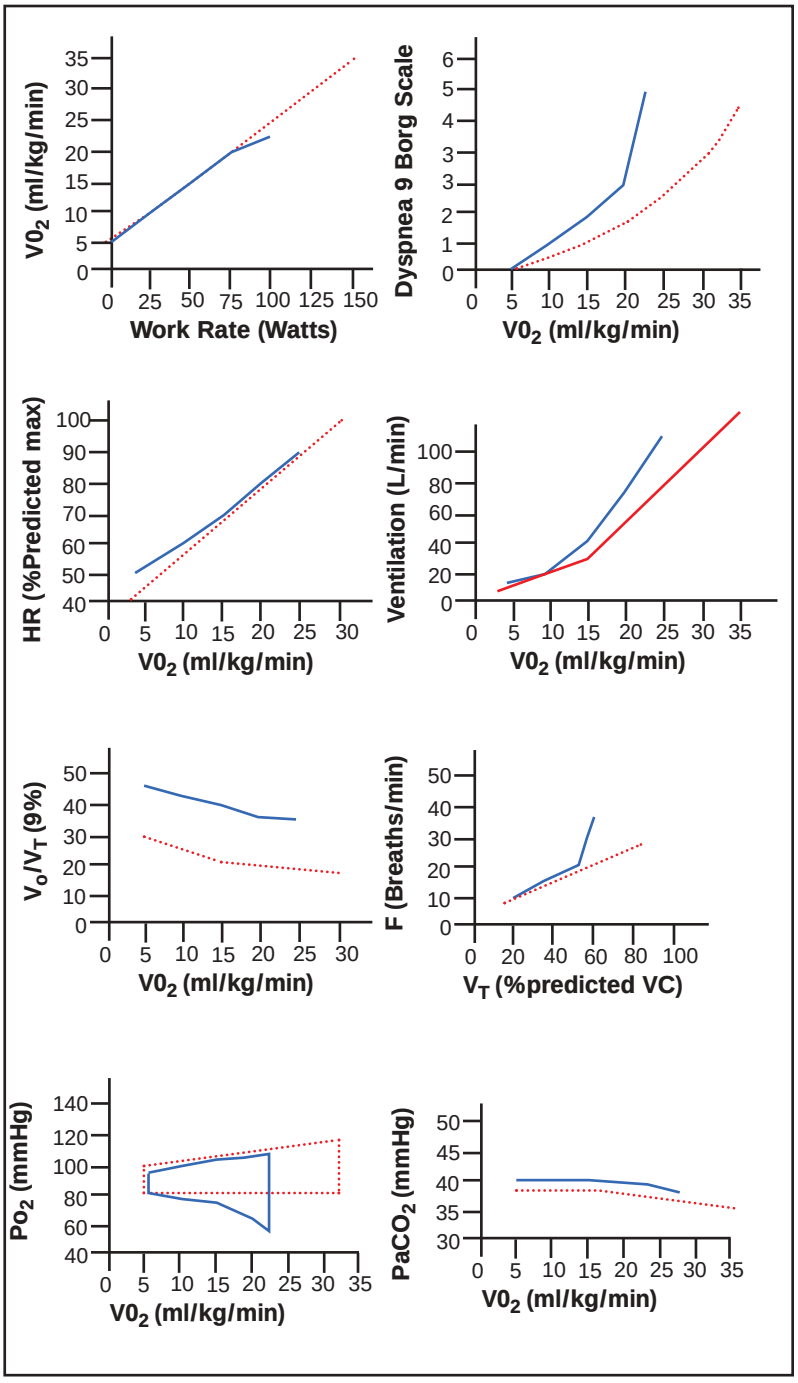

Figure 1. Results of the patient's CPET.

What's your diagnosis?

a) Early interstitial lung disease (ILD)
b) Heart failure
c) Recurrent pulmonary emboli
d) Deconditioning

Answer on page 38 ⟶

Answer:
Recurrent pulmonary emboli

Given the CPET findings (Table 1), the differential diagnosis would include chronic obstructive pulmonary disease, ILD, and pulmonary hypertension (from any cause). Since the chest radiograph and PFTs are normal, the most likely diagnosis would be pulmonary hypertension.

An echocardiogram is done, which rules out any cardiac cause for the symptoms, but fails to demonstrate elevated right ventricular systolic pressures. Therefore, pulmonary angiogram is required as a definitive test.

The angiogram demonstrates elevated pulmonary arterial pressures and multiple, bilateral filling defects consistent with the presence of acute and subacute pulmonary emboli.

Table 1
Results of the CPET

- Exercise limitation secondary to severe breathlessness
- Normal oxygen saturation throughout the exercise
- Reduced symptom limited peak of oxygen utilization (VO_2 max)
- Hyperventilation during submaximal exercise
- High dead space (wasted ventilation), suggesting a perfusion defect
- Blunted fall in dead space during exercise, with an eventual plateau
- Reduced P_{AO2} and widened A-a gradient

CPET: Cardiopulmonary exercise test
P_{AO2}: Partial pressure of oxygen in alveolar gas gradient
A-a: Partial pressure of oxygen gradient between alveolar gas and arterial blood

More on CPET

CPET is being increasingly used in clinical medicine, as it can offer objective assessment of patients' cardiac or respiratory reserves. It has a wide spectrum of clinical applications, but is mainly used for the evaluation of impairment or disability, in pre-operative evaluation for lung resection, for evaluation of unexplained dyspnea, and for prediction of survival and response to therapeutic interventions.

Making the diagnosis of early or mild pulmonary hypertension through tests performed on a patient at rest can be extremely difficult. Such patients often present with dyspnea or fatigue of unknown origin, which are classic indications for CPET. By the time patients are symptomatic, their maximum oxygen uptake is generally decreased due to depressed cardiac output and arterial oxygen desaturation. Ventilation is often excessive and inefficient. These patients do not have the normal pulmonary vascular distension and recruitment during exercise, leading to increased dead space and ventilatory requirements during activity. The exaggerated widening of the partial pressure of oxygen gradient between alveolar gas and arterial blood with exercise is a gas exchange feature that helps distinguish pulmonary arterial from venous hypertension.

CPET is a safe, useful, and relatively inexpensive investigation that provides objective information regarding patients' functional limitations. It can also provide important mechanistic insight into the origin of exertional symptoms. Dx

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