COR PULMONALE

The most frequent cause of death of patients with PAH is right ventricular failure, also called *cor pulmonale.* Prolonged increased afterload causes the right ventricle to hypertrophy and then dilate. The interventricular septum shifts to the left, and filling of the left ventricle is decreased, with subsequent decreased cardiac output. Dilation of the right atrium causes atrial tachyarrhythmias and further decreased cardiac output.

Treatment of cor pulmonale is directed at the underlying cause of pulmonary hypertension. The existence of cor pulmonale is a bad prognostic sign in group 1 pulmonary hypertension (level 1 evidence).

PULMONARY THROMBOEMBOLISM

Definition and Epidemiology

Pulmonary thromboembolism refers to the passage of a clot from the venous system or the right ventricle that lodges in a pulmonary artery. Other materials, such as tumor or injected foreign bodies (e.g., talc) can also lodge in the lung circulation, but pulmonary thromboembolism is a complication of venous thrombosis.

Pulmonary thromboembolic disease is a relatively common entity, with an incidence ranging from 400,000 to 650,000 cases per year in the United States. The deep veins of the femoral and popliteal systems of the lower extremities are most often involved, but right atrial, right ventricular, and upper extremity thromboses can also embolize to the lung. The predisposing factors for pulmonary embolism are the same as those for venous thrombosis and include venous stasis, hypercoagulability, and endothelial injury. Congenital or acquired procoagulant disorders (e.g., activated protein C deficiency, factor V Leiden) are also considered predisposing factors.

For a deeper discussion on this topic, please see Chapter 68, "Pulmonary Hypertension," in Goldman-Cecil Medicine, *25th Edition.*

Pathology

After a clot dislodges from the lower extremity circulation, it travels to the pulmonary circulation, where it can obstruct a branch of the pulmonary artery. The affected lung segment develops an increased $\dot{V}/\dot{Q}$ ratio. This increases overall dead space ventilation, which can lead to inefficient excretion of carbon dioxide, potentially raising the partial pressure of carbon dioxide in arterial blood ($Paco_2$). Blood flow is shifted from the obstructed site to other areas, which may include areas with a low $\dot{V}/\dot{Q}$ ratio, leading to shunting and hypoxemia.

Infarction of the lung distal to the occlusion is rare because of the redundancy of the pulmonary circulation and because of oxygenation of lung parenchyma by bronchial arteries and by alveolar oxygen. However, pulmonary thromboembolism causes deficiency or inactivation of surfactant and commonly causes collapse of alveolar units perfused by the obstructed vessel.

Clinical Presentation

The most common symptom of pulmonary thromboembolism is shortness of breath (level 1 evidence). Other, less common symptoms include chest pain, hemoptysis, and syncope. A careful history is paramount when evaluating patients for thromboembolic disease to determine risk factors, such as recent immobilization or surgery, malignancy, or a history of pulmonary embolus or deep vein thrombosis. Using a validated clinical scoring system to assess the pretest probability of pulmonary embolism, such as calculation of the Wells or Geneva score, is helpful in the integration of subsequent laboratory and radiologic testing results.

The most common physical examination findings are tachycardia and tachypnea (level 1 evidence). The physical examination may be normal or may reveal isolated crackles or even diffuse wheezing. Pleural effusions are identified by areas of dullness to percussion. Edema of the extremities, especially if the edema is asymmetrical, may point to venous thrombosis. In deep vein thrombosis, dorsiflexion of the foot may cause calf pain as a result of stretching the calf muscles and deep veins (Homan's sign). Signs of pulmonary hypertension and right ventricular strain, such as increased pulmonary component of the second heart sound or right ventricular heave, are not usually appreciated unless there is a massive pulmonary embolus or preexisting heart or lung disease.

Arterial blood gas determinations usually reveal respiratory alkalosis with a normal Pao_2 value (level 1 evidence). However, when the alveolar-arterial oxygen gradient is calculated, it is frequently widened. However, a normal alveolar-arterial gradient in the partial pressure of oxygen does not exclude acute pulmonary embolism. A normal $Paco_2$ in a patient with tachypnea suggests increased dead space and, in the appropriate setting, may point to the diagnosis. In severe cases, arterial blood gas measurement may show acidemia, hypoxemia, and hypercapnia.

An elevated level of lactate dehydrogenase (LDH) may be the result of tissue infarction, but this test is insensitive and nonspecific. An elevated level of B-type natriuretic peptide (BNP) is useful in assessing the severity and likelihood of complicating right ventricular failure. If the highly sensitive D-dimer level is normal, it effectively excludes pulmonary thromboembolism if the pretest probability is intermediate or low. However, the D-dimer level may be elevated in patients with unrelated medical conditions, such as congestive heart failure, chronic illness, and connective tissue disorders.

The electrocardiogram may show atrial tachyarrhythmias or evidence of right heart strain as shown by a new right bundle branch block, right ventricular strain pattern, and the $S_I\ Q_{III}\ T_{III}$ pattern (i.e., S wave in lead I, Q wave in lead III, T wave inversion in lead III) that mimics inferior myocardial infarction. The chest radiograph is often normal but may show atelectasis, isolated infiltrates, or a small pleural effusion. Oligemia (Westermark's sign), an abrupt cutoff of pulmonary vessels or enlarged central pulmonary arteries (Fleischner's sign), and a pleural-based area of increased opacity (Hampton's hump) may be seen on the radiograph. In most cases, chest radiographs are not sufficiently sensitive to diagnose a pulmonary embolism.

Three major diagnostic methods are used for the diagnosis of pulmonary embolism: the $\dot{V}/\dot{Q}$ scan, chest CT, and pulmonary arteriography (Fig. 18-1). CT angiography provides a noninvasive and sensitive way to detect pulmonary emboli (E-Fig. 18-5). The Prospective Investigation of Pulmonary Embolism Diagnosis II (PIOPED II) study demonstrated that the best approach is a combination of clinical suspicion, D-dimer determination, CT