TABLE 279-4 ETIOLOGY OF CHRONIC COR PULMONALE

Diseases of the Lung Parenchyma
Chronic obstructive pulmonary disease
Emphysema
Chronic bronchitis
Cystic fibrosis
Idiopathic interstitial pneumonitis
Idiopathic pulmonary fibrosis
Nonspecific interstitial pneumonitis
Sarcoidosis
Bronchiectasis
Pulmonary Langerhans cell histiocytosis
Lymphangioleiomyomatosis
Disorders of Chronic (Alveolar) Hypoxia
Alveolar hypoventilation syndromes
Obesity hypoventilation syndrome
Central hypoventilation syndrome
Neuromuscular respiratory failure
Chest wall disorders
Kyphoscoliosis
Living at high altitude
Diseases of the Pulmonary Vasculature
Pulmonary arterial hypertension (PAH)
Idiopathic PAH
Heritable PAH
Associated PAH
Venoocclusive disease
Chronic thromboembolic pulmonary hypertension
Pulmonary tumor thrombotic microangiopathy

water homeostasis. Anatomically, the RV is a thin-walled, compliant chamber that is better suited to handle volume overload than pressure overload. Thus, the sustained pressure overload imposed by pulmonary hypertension and increased pulmonary vascular resistance eventually causes the RV to fail.

The response of the RV to pulmonary hypertension depends on the acuteness and severity of the pressure overload. Acute cor pulmonale occurs after a sudden and severe stimulus (e.g., massive pulmonary embolus), with RV dilatation and failure but no RV hypertrophy (Chap. 300). Chronic cor pulmonale, however, is associated with a more slowly evolving and progressive pulmonary hypertension that leads to initial modest RV hypertrophy and subsequent RV dilation. Acute decompensation of previously compensated chronic cor pulmonale is a common clinical occurrence. Triggers include worsening hypoxia from any cause (e.g., pneumonia), acidemia (e.g., exacerbation of COPD), acute pulmonary embolus, atrial tachyarrhythmia, hypervolemia, and mechanical ventilation that leads to compressive forces on alveolar blood vessels.

CLINICAL MANIFESTATIONS

Symptoms The symptoms of chronic cor pulmonale generally are related to the underlying pulmonary disorder. Dyspnea, the most common symptom, is usually the result of the increased work of breathing secondary to changes in elastic recoil of the lung (fibrosing lung diseases), altered respiratory mechanics (e.g., overinflation with COPD), or inefficient ventilation (e.g., primary pulmonary vascular disease). Orthopnea and PND are rarely symptoms of isolated right HF and usually point toward concurrent left heart dysfunction. Rarely, these symptoms reflect increased work of breathing in the supine position resulting from compromised diaphragmatic excursion. Abdominal pain and ascites that occur with cor pulmonale are similar to the right HF that ensues in chronic HF. Lower-extremity edema may occur secondary to neurohormonal activation, elevated RV filling pressures, or increased levels of carbon dioxide and hypoxemia, which can lead to peripheral vasodilation and edema formation.

Signs Many of the signs encountered in cor pulmonale are also present in HF patients with a depressed EF, including tachypnea, elevated jugular venous pressures, hepatomegaly, and lower-extremity edema. Patients may have prominent *v* waves in the jugular venous pulse as a result of tricuspid regurgitation. Other cardiovascular signs include an RV heave palpable along the left sternal border or in the epigastrium. The increase in intensity of the holosystolic murmur of tricuspid regurgitation with inspiration ("Carvallo's sign") may be lost eventually as RV failure worsens. Cyanosis is a late finding in cor pulmonale and is secondary to a low cardiac output with systemic vasoconstriction and ventilation-perfusion mismatches in the lung.

DIAGNOSIS

The most common cause of right HF is not pulmonary parenchymal or vascular disease but left HF. Therefore, it is important to evaluate the patient for LV systolic and diastolic dysfunction. The ECG in severe pulmonary hypertension shows P pulmonale, right axis deviation, and RV hypertrophy. Radiographic examination of the chest may show enlargement of the main central pulmonary arteries and hilar vessels. Spirometry and lung volumes can identify obstructive and/or restrictive defects indicative of parenchymal lung diseases; arterial blood gases can demonstrate hypoxemia and/or hypercapnia. Spiral computed tomography (CT) scans of the chest are useful in diagnosing acute thromboembolic disease; however, ventilation-perfusion lung scanning remains best suited for diagnosing *chronic thromboembolic disease* (Chap. 300). A high-resolution CT scan of the chest can identify interstitial lung disease.

Two-dimensional echocardiography is useful for measuring RV thickness and chamber dimensions. Location of the RV behind the sternum and its crescent shape challenge assessment of RV function by echocardiography, especially when parenchymal lung disease is present. Calculated measures of RV function (e.g., tricuspid annular plane systolic excursion [TAPSE] or the Tei Index) supplement more subjective assessments of RV function. The interventricular septum may move paradoxically during systole in the presence of pulmonary hypertension. As noted, Doppler echocardiography can be used to assess pulmonary artery pressures. MRI is also useful for assessing RV structure and function, particularly in patients who are difficult to image with 2-D echocardiography because of severe lung disease. Right-heart catheterization is useful for confirming the diagnosis of pulmonary hypertension and for excluding elevated left-heart pressures (measured as the pulmonary capillary wedge pressure) as a cause for right HF. BNP and N-terminal BNP levels are elevated in patients with cor pulmonale secondary to RV myocardial stretch and may be dramatically elevated in acute pulmonary embolism.