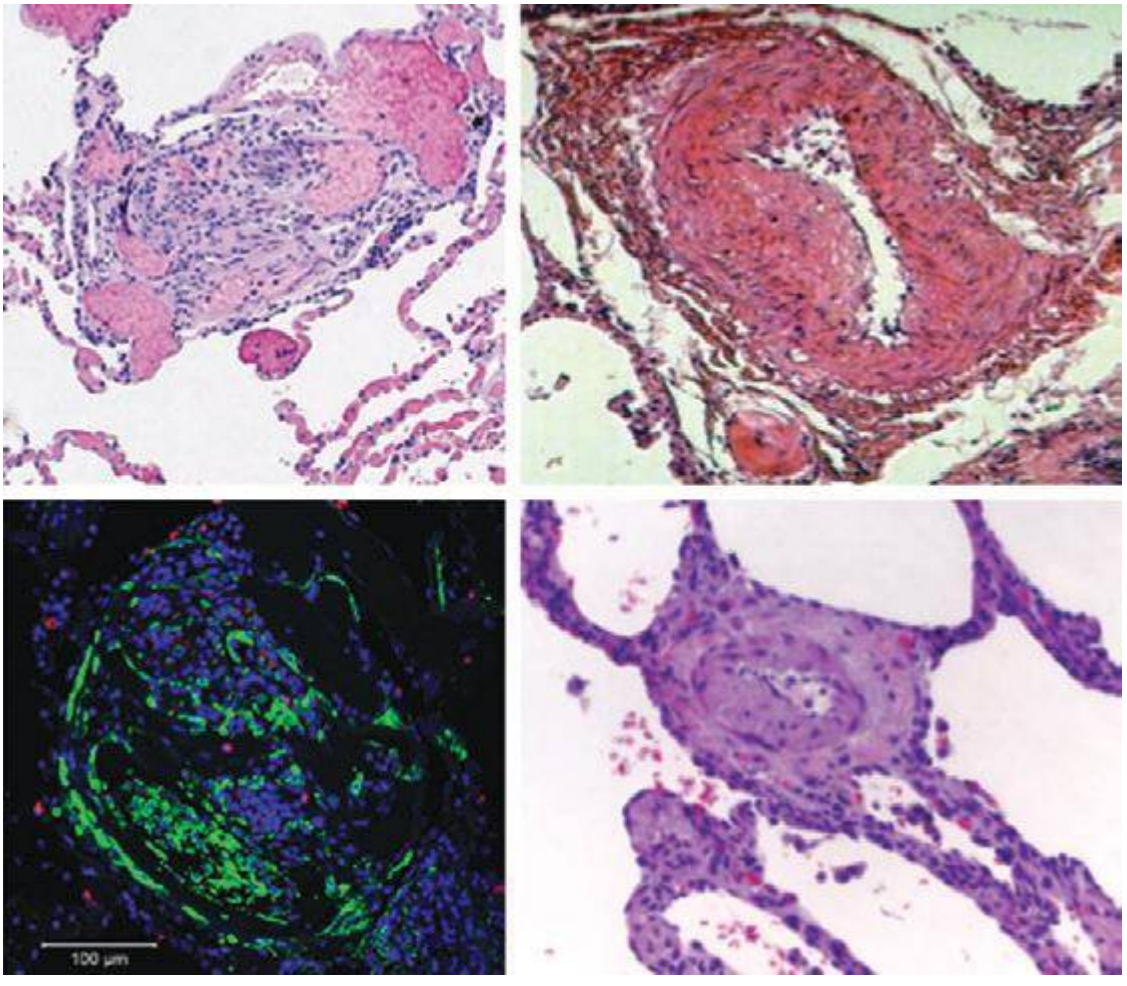


**FIGURE 304-1 The *left panels* show examples of plexogenic pulmonary arteriopathy.** These are obstructive and proliferative lesions of the small muscular pulmonary arteries, composed primarily of endothelial cells with intermixed inflammatory cells, myofibroblasts, and connective tissue components. The *lower left panel* demonstrates proliferating cells (red PCNA stained cells). *Panels on the right* demonstrate medial hypertrophy of muscular pulmonary arteries. *(Photographs on the left are courtesy of Dr. Stephen Archer, Queen's University School of Medicine, Kingston, Ontario, Canada.)*

dilated right ventricle (Fig. 304-2) with elevated estimated pulmonary artery systolic pressure. Important additional information can be gleaned about specific etiologies of PH such as valvular disease, left ventricular systolic and diastolic function, intracardiac shunts, and other cardiac diseases.

Although the accuracy of Doppler echocardiography is often debated, a high-quality echocardiogram that is absolutely normal may obviate the need for further evaluation for PH. An echocardiogram is a screening test, whereas invasive hemodynamic monitoring is the gold standard for diagnosis and assessment of disease severity. With a normal echocardiogram, there may still be some concern for PH; this is particularly true if there is unexplained dyspnea or hypoxemia. In this setting, it is reasonable to proceed to right heart catheterization for definitive diagnosis. Alternatively, if the patient has a reasonable functional capacity, a cardiopulmonary exercise test may help to identify a true physiologic limitation as well as differentiate between cardiac and pulmonary causes of dyspnea. If this test is normal, there is no indication for a right heart catheterization. If a cardiovascular limitation to exercise is found, a right heart catheterization should be pursued.

If the echocardiogram or cardiopulmonary exercise test (CPET) suggests PH and the diagnosis is confirmed by catheterization, a reasonable effort must be made to establish the etiology because this will largely determine the therapeutic approach. A stepwise approach to evaluation is outlined below.

Chest imaging and lung function tests are essential because lung disease is an important cause of PH. A sign of PH that may be evident on chest x-ray include enlargement of the central pulmonary arteries associated with "vascular pruning," a relative paucity of peripheral vessels (Fig. 304-3). Cardiomegaly, with specific evidence of right atrial and ventricular enlargement, can often be observed. The chest x-ray may also demonstrate significant interstitial lung disease or suggest hyperinflation from obstructive lung disease, which may be the underlying cause or contributor to the development of PH. High-resolution computed tomography (CT) may provide additional useful information. Classic findings of PH on CT include those found on chest x-ray: enlarged pulmonary arteries (Fig. 304-4), peripheral pruning of the small vessels, and enlarged right ventricle and atrium. However, high-resolution CT may also reveal signs of venous congestion including centrilobular ground-glass infiltrate and thickened septal lines. In the absence of left heart disease, these findings suggest pulmonary veno-occlusive disease, a rare cause of PAH that can be quite challenging to diagnose.

CT angiograms are commonly used to evaluate acute thromboembolic disease and have demonstrated excellent sensitivity and specificity for that purpose. Ventilation-perfusion ($\dot{V}/\dot{Q}$) scanning has traditionally been used for screening because of its high sensitivity and its role in qualifying patients for surgical intervention. The role of CT angiograms in the diagnosis of chronic thromboembolic pulmonary hypertension (CTEPH) remains controversial, even with the advent of spiral CT. Although a negative $\dot{V}/\dot{Q}$ virtually rules out CTEPH, some cases may be missed through the use of CT angiograms.

Pulmonary function tests are an important component of the evaluation. Although an isolated reduction in $DL_{CO}$ is the classic finding in PAH, results of pulmonary function tests may also suggest restrictive or obstructive lung diseases as the cause of dyspnea or PH. The 6-minute walk test