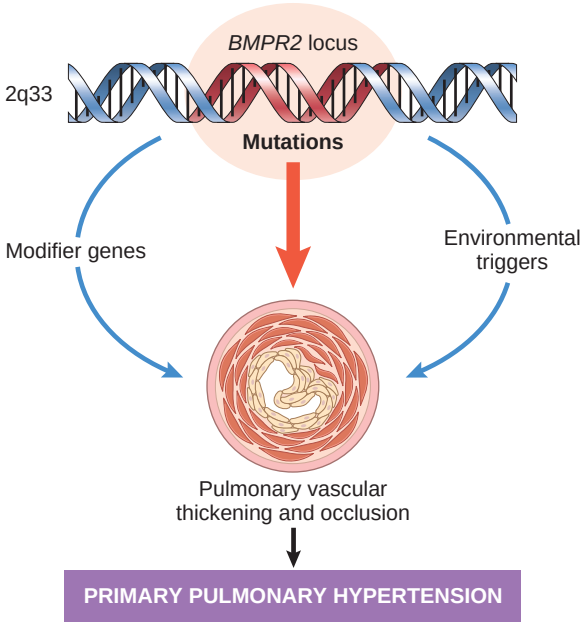

Figure 15-29 Pathogenesis of primary (idiopathic) pulmonary hypertension. See text for details.

MORPHOLOGY

Regardless of their etiology, all forms of pulmonary hypertension are associated with **medial hypertrophy of the pulmonary muscular and elastic arteries, pulmonary arterial atherosclerosis, and right ventricular hypertrophy**. The presence of many organizing or recanalized thrombi favors recurrent pulmonary emboli as the cause, and the coexistence of diffuse pulmonary fibrosis, or severe emphysema and chronic bronchitis, points to chronic hypoxia as the initiating event. The vessel changes can involve the entire arterial tree, from the main pulmonary arteries down to the arterioles (Fig. 15-30). In the most severe cases, atheromatous deposits form in the pulmonary artery and its major branches, resembling (but lesser in degree than) systemic atherosclerosis. The arterioles and small arteries (40 to 300 μm in diameter) are most prominently affected by striking medial hypertrophy and intimal fibrosis, sometimes narrowing the lumens to pinpoint channels. One extreme in the spectrum of pathologic changes is the plexiform lesion, so called because a tuft of capillary formations is present, producing a network, or web, that spans the lumens of dilated thin-walled, small arteries and may extend outside the vessel. Plexiform lesions are most prominent in idiopathic and familial pulmonary hypertension (group 1), unrepaired congenital heart disease with left-to-right shunts (group 2), and pulmonary hypertension associated with human immunodeficiency virus (HIV) infection and drugs (also group 1).

Clinical Course. Idiopathic pulmonary hypertension is most common in women who are 20 to 40 years of age and is also seen occasionally in young children. Clinical signs and symptoms in all forms of pulmonary hypertension become evident only in advanced disease. In cases of idiopathic disease, the presenting features are usually dyspnea and fatigue, but some patients have chest pain of the anginal type. Over time, severe respiratory distress, cyanosis, and right ventricular hypertrophy occur, and death from decompensated cor pulmonale, often with superimposed thromboembolism and pneumonia, usually ensues within 2 to 5 years in 80% of patients.

Treatment choices depend on the underlying cause. For those with secondary disease, therapy is directed at the trigger (e.g, thromboembolic disease or hypoxemia). A variety of vasodilators have been used with varying success in those with group 1 or refractory disease belonging to other groups. Lung transplantation provides definitive treatment for selected patients.

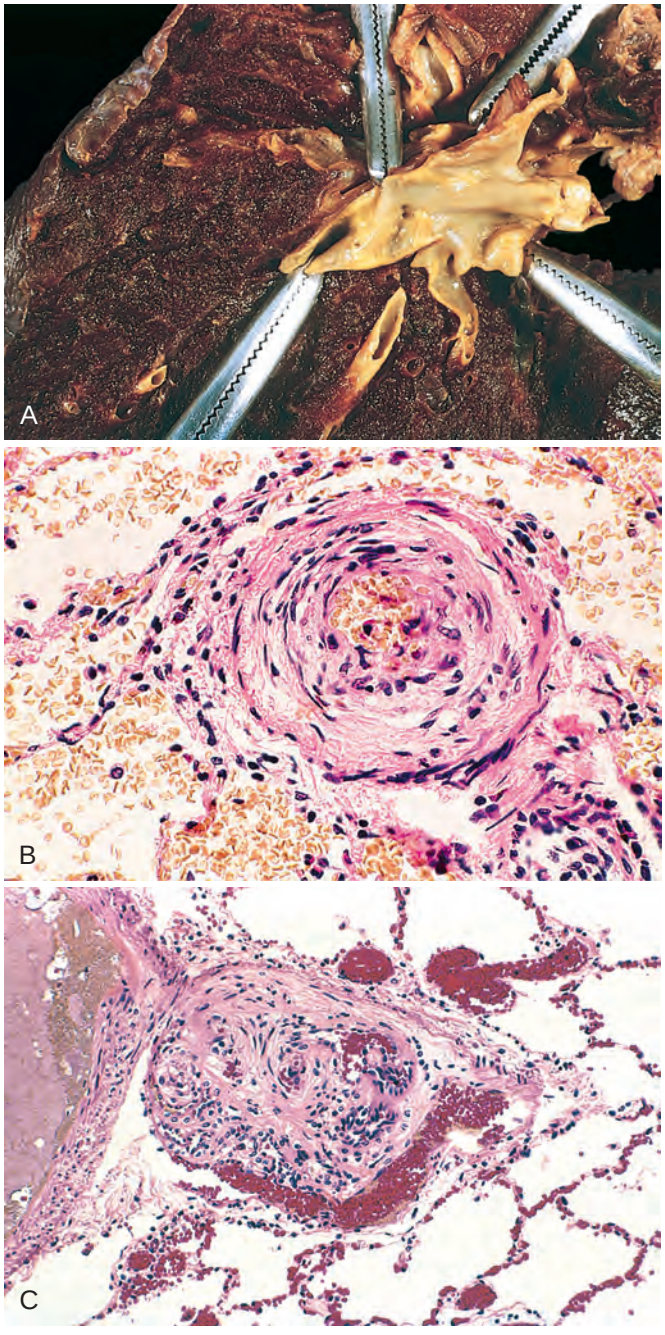

Figure 15-30 Vascular changes in pulmonary arterial hypertension. **A,** Atheroma formation, a finding usually limited to large vessels. **B,** Marked medial hypertrophy. **C,** Plexiform lesion of small arteries that is characteristic of advanced pulmonary hypertension.