right heart failure. The 3-year survival of SSc patients with untreated PAH is <50%. Risk factors include limited cutaneous disease, older age at disease onset, high numbers of cutaneous telangiectasia, and antibodies to centromere, U3-RNP (fibrillarin), and B23. Mutations in the *BMPR2* gene implicated in idiopathic PAH are not associated with SSc-PAH.

In SSc, PAH is often asymptomatic in early stages. Patients present with exertional dyspnea and reduced exercise capacity. With progression, angina, near-syncope, and symptoms and signs of right-sided heart failure appear. Physical examination may show tachypnea, a loud pulmonic component of the S_2 heart sound, pulmonic/tricuspid regurgitation murmur, palpable right ventricular heave, elevated jugular venous pressure, and dependent edema. Doppler echocardiography is a noninvasive screening method for estimating the pulmonary arterial pressure. In light of the poor prognosis of untreated PAH and better therapeutic response in patients with early diagnosis, SSc patients should be screened for PAH at initial evaluation and annually thereafter. Estimated pulmonary artery systolic pressure >40 mmHg at rest or tricuspid regurgitation jet velocities >3 m/s suggest PAH. Pulmonary function testing may show a reduced D_{LCO} in isolation or out of proportion with the severity of restriction. Because echocardiography can over- or underestimate pulmonary artery pressures, cardiac catheterization is required to confirm the diagnosis of suspected PAH; assess its severity, including the degree of right heart dysfunction; rule out veno-occlusive disease and other cardiac (postcapillary) causes of pulmonary hypertension; and provide prognostic parameters. Distinguishing PAH in SSc patients from pulmonary hypertension secondary to pulmonary fibrosis and hypoxia can be difficult. Serum levels of N-terminal pro-brain natriuretic peptide (NT-proBNP) correlate with the presence and severity of PAH, as well as survival. While NT-proBNP can be useful in PAH screening and in monitoring the response to treatment, elevated levels are not specific for PAH and also occur in other forms of right and left heart disease. Despite more favorable hemodynamics of SSc-associated PAH, its prognosis is worse and treatment response poorer than for idiopathic PAH.