PH categories listed here sequentially as groups 1–5: PAH; PH due to left heart disease; PH due to chronic lung disease or sleep-disordered breathing; CTEPH; and a group of miscellaneous diseases that rarely (or inconsistently) cause PH.

Pulmonary Arterial Hypertension WHO group 1 PH, or PAH, involves marked pulmonary arterial precapillary remodeling, including intimal fibrosis, increased medial thickness, pulmonary arteriolar occlusion, and classic plexiform lesions. The hemodynamic criteria for PAH are sustained elevation in resting mPAP >20 mmHg, PVR ≥3.0 WU, and PAWP or LVEDP of ≤15 mmHg based on RHC. Idiopathic PAH (IPAH) is a progressive disease that leads to right heart failure and early mortality. From the original National Institutes of Health registry on IPAH in 1987, the average age at diagnosis was 36 years, with only 9% of patients with IPAH over the age of 60. However, contemporary data now inclusive of numerous international registries suggest a different clinical profile. The mean age of PAH patients is reported to be 54–68 years old across studies. This reflects, in part, rising awareness of this disease in the elderly. The prevalence of IPAH favors women to men by ~3.1-fold; however, the hemodynamics at diagnosis are more severe, and the prognosis is less favorable in men compared to women.

Diseases Associated with PAH Other forms of PAH that deserve specific consideration are those associated with congenital heart disease with intracardiac shunt, connective tissue disease, portal hypertension, and HIV.

CONGENITAL HEART DISEASE PAH in the setting of congenital heart disease is important to recognize since surgical correction may be indicated and when successful is associated with favorable prognosis. This is particular salient today, as more congenital heart disease patients live to adulthood and populate general medical practices. Still, referral to adult congenital heart disease centers should be considered for patients with suspected PAH, which in this population is subclassified into four groups: Eisenmenger's syndrome, systemic-to-pulmonary shunts, coincidental or small