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**Title:** Modern age pathology of pulmonary arterial hypertension

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**Body:** Introduction: The impact of modern treatments of pulmonary arterial hypertension (PAH) on pulmonary vascular pathology remains unknown. The last series reporting the pathology of severe pulmonary hypertension date back two decades, well before usage of current therapies for the disease. Methods: Assessment of pulmonary vascular remodelling and inflammation in 62 PAH and 28 control explanted lungs systematically sampled, with matched clinical data. The tissue was obtained by the Pulmonary Hypertension Breakthrough Initiative. Results: Total wall, intima, and media fractional thicknesses of pulmonary arteries were increased in the PAH group versus the controls, and correlated with pulmonary hemodynamics. Despite a high variability of morphological measurements within a given PAH lung and among all PAH lungs, pathological subphenotypes were detected in cohorts of PAH lungs. This included a subset of lungs lacking intima or media remodeling, which had similar numbers of profiles of plexiform lesions as those in lungs with more pronounced remodeling. Perivascular inflammation was present in a high number of PAH lungs and correlated with mean pulmonary arterial pressure (mPAP) as well as intima and total wall thickness. The number of profiles of plexiform lesions was significantly lower in lungs of patients who were never treated with prostacyclin or its analogues. Conclusions: Our results indicate that multiple features of pulmonary vascular remodeling are present in patients treated with modern PAH regimens. Perivascular inflammation may have an important role in the processes of vascular remodeling, all of which may ultimately lead to increased pulmonary artery pressure.