A great variety of pulmonary vascular lesions have been described in congenital and acquired heart disease. Many clinicians and pathologists are not too familiar with these alterations nor with the fact that these lesions may form certain patterns, more or less characteristic for the underlying hemodynamic changes.

A schematic representation (Fig 1) of a classification of five patterns, each corresponding with a certain cardiovascular disease or group of diseases, is presented. I hope that this may aid in recognizing relevant lesions in the pulmonary vasculature and in establishing a correct diagnosis when these are observed in autopsy material or in a lung biopsy.

Group 1 is formed by cases of vasoconstrictive pulmonary hypertension, whether due to congenital cardiac defects or to primary pulmonary hypertension. In these instances there is hypertrophy of the media (red) and proliferation and fibrosis of the intima (yellow). The intimal fibrosis is of a concentric and laminar type. In addition, particularly in more severe pulmonary hypertension, there may be fibrinoid necrosis (pink) of the media with or without arteritis and the so-called plexiform lesions and dilatation lesions.

In the plexiform lesions the media is usually damaged, probably due to medial necrosis, while organization and recanalization of a fibrin clot in the lumen leads to a plexus of small channels, opening up in the dilated distal portion of the artery.

The pattern is not necessarily complete, particularly not in young children. In infants, the changes are usually limited to medial hypertrophy, and then the picture may be inconclusive. Concentric laminar intimal fibrosis and plexiform lesions are diagnostic for vasoconstrictive pulmonary hypertension.

Group 2 is represented by cases of pulmonary hypertension due to chronic pulmonary embolism. Here the media is usually normal or only mildly hypertrophic. Patchy eccentric intimal fibrosis and irregular intraluminal fibrous septa are the sequela of organized or recanalized emboli. The pattern is totally different from that in primary pulmonary hypertension (group 1) with which it may clinically be confused.

In group 3, comprising cases with increased pulmonary venous pressure, as in mitral valvular disease, both arteries and veins are affected. In the arteries, medial hypertrophy is usually severe and so is intimal fibrosis which is either concentric or eccentric but not laminar. Arteritis is uncommon. The media of the veins (blue) may be thickened. More characteristic is arteriolization of the venous media which structurally resembles that of an artery. Fibrosis of the venous intima is regularly present.

Group 4 comprises cases of pulmonary hypertension in chronic hypoxia, whether due to parenchymal disease of the lungs, living at high altitudes, or otherwise. The media of the arteries is generally normal or mildly hypertrophied. Only the smallest arterioles may show more marked medial hypertrophy. Patchy, eccentric intimal fibrosis is often present but may well be due to the underlying pulmonary disease. The occurrence of longitudinal smooth muscle bundles in the intima is pathognomonic, particularly of the small pulmonary arteries. The latter changes are also often present in cases of mitral valvular disease, especially when there is interstitial fibrosis of the lungs.

Group 5 does not refer to patients with pulmonary hypertension but to those with diminished pulmonary flow, as in tetralogy of Fallot. Here the pulmonary arteries are wider than normal, with a thin, atrophic media. Eccentric intimal fibrosis and delicate intraluminal fibrous septa, as an expression of organization and recanalization of autochthonous
thrombi, are common.

A classification of this type is, by necessity, a simplification. Therefore, some rules should be observed. An adequate number of vessels should be studied so as to get a representative picture. An elastic stain, in addition to the routine hematoxylin stain, is indispensable. It should be kept in mind that in some instances components of more than one pattern may be present in the same lung. Left cardiac failure, for instance, may add pulmonary venous alterations to the pattern of vasoconstrictive pulmonary hypertension, and chronic embolism may complicate a case of mitral stenosis. When this is realized, this classification may serve its purpose.

Figure 1. This is a simplified schematic representation of pulmonary vessels with characteristic lesions in five groups of pulmonary vascular disease. The media of muscular pulmonary arteries is depicted in red; that of a pulmonary vein in blue; intimal changes are shown in yellow. Each of the five groups is represented by its own pattern of lesions (described in text). (1) Vasoconstrictive pulmonary hypertension: medial hypertrophy; concentric laminar intimal fibrosis; fibrinoid necrosis (pink) without or with arteritis; plexiform lesion. (2) Chronic thromboembolism: mild medial hypertrophy; eccentric intimal fibrosis; intraluminal fibrous septa. (3) Pulmonary venous hypertension: severe medial hypertrophy; eccentric or concentric but nonlaminar intimal fibrosis; arterialization and intimal fibrosis of pulmonary veins. (4) Chronic hypoxic pulmonary hypertension: normal media (although often medial hypertrophy of small arterioles); eccentric intimal fibrosis; longitudinal muscle bundles in intima. (5) Decreased pulmonary flow: wide arteries with medial atrophy; eccentric intimal fibrosis; intraluminal fibrous septa.