Pulmonary veins stenosis

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Abstract
Pulmonary veins (PV) stenosis is an heterogeneous entity including stenosis of normally positioned individual PV and PV obstruction accompanying total anomalous pulmonary venous connection. Congenital stenosis or atresia of individual PV is a very rare anomaly encountered in about 0.5% of autopsy in children. It may manifest as an isolated lesion or associated with other cardiac defects (more than 50% of cases). In all cases of congenital and acquired stenosis of PV, the lesion is progressive and includes narrowing of one or more PV at their junction with the left atrium, or hypoplasia with narrowed intra- or extrapulmonary portions at variable distances. Most commonly, all PV of one lung are affected, causing pulmonary hypertension (PH) and consecutive pulmonary arterial hypertension (PAH). By contrast, lesions of a single PV may not lead to PH. In case of PH and PAH, severe symptoms of pulmonary edema occur in early infancy including dyspnea, tachypnea, repeated pulmonary infections, hemoptysis, and in the most severe cases cyanosis and signs of right heart failure. Etiology of congenital stenosis or atresia of individual PV is a fibrotic stenosis. Acquired stenosis of PV may be due to constrictive pericarditis, mediastinitis, tuberculosis, obstructive tumors or operative scar. Angiocardiography may allow identification of the precise anatomy of PV stenosis. Without surgical treatment, patients with significant stenosis of all or most PV die before reaching adulthood and often much sooner. Overall prognosis is bad when at least 50% of PV are stenotic.

Keywords

Included diseases
Pulmonary veins (PV) stenosis is an heterogeneous entity including stenosis of normally positioned individual PV and PV obstruction accompanying total anomalous pulmonary venous connection.

Differential diagnosis
Differential diagnosis includes pulmonary infections in case of stenosis of one or two PV and all causes of pulmonary hypertension (PH) such as mitral insufficiency, mitral stenosis, supraavalvular mitral membrane and divided left atrium and total anomalous pulmonary venous connection.

Frequency
Congenital stenosis or atresia of individual PV is a very rare anomaly encountered in about 0.5% of autopsy in children. It may manifest as an isolated lesion or be associated with other cardiac defects (more than 50% of cases).
Clinical signs
In all cases of congenital and acquired stenosis of PV, the lesion is progressive and includes narrowing of one or more PV at their junction with the left atrium, or hypoplasia with narrowed intra- or extrapulmonary portions at variable distances. Most commonly, all PV of one lung are affected, causing PH and consecutive pulmonary arterial hypertension (PAH). By contrast, lesions of a single PV may not lead to PH. In case of PH and PAH, severe symptoms of pulmonary edema occur in early infancy including dyspnea, tachypnea, repeated pulmonary infections, hemoptysis, and in the most severe cases cyanosis and signs of right heart failure.

Etiology
Etiology of congenital stenosis or atresia of individual PV is a fibrotic stenosis. Acquired stenosis of PV may be due to constrictive pericarditis, mediastinitis, tuberculosis, obstructive tumours or operative scar.

Diagnostic methods
Cardiac auscultation is non specific with an accentuated 2nd heart sound in cases with PAH. In that case, electrocardiography shows right atrial and right ventricular hypertrophy. Chest-x-ray typically demonstrates normal heart size, prominent pulmonary trunk, reticular appearance of the lungs or ground-glass opacification, and evidence of Kerley B lines in the region of the obstructed veins.

On transthoracic or transoesophageal echocardiography, examination of the point of entry of the pulmonary veins into the left atrium may show turbulent flow. Lung scans may show reduced or absent perfusion in the affected pulmonary region. Cardiac catheterization documents PAH and increased pulmonary wedge pressure but normal left atrial pressure. Angiocardiography may allow identification of the precise anatomy of PV stenosis.

Treatment
Without surgical treatment, patients with significant stenosis of all or most PV die before reaching adulthood and often much sooner. No effective treatment of long-segment atresia or severe PV hypoplasia except unilateral pneumonectomy is available. Patch grafting or balloon angioplasty has been tried in atresia or localized stenosis, but lasting success is unlikely. Overall prognosis is bad when at least 50% of PV are stenotic.

References