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Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. <u>Pulmonary</u> <u>veno-occlusive disease and/or pulmonary capillary haemangiomatosis</u>. ORPHA:431353

Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis (PVOD and/or PCH) constitutes a rare subgroup of pulmonary arterial hypertension (PAH; see this term) characterized by obliterative fibrosis of the small pulmonary veins and venules and/or capillary infiltration of the pulmonary interstitium leading to increased pulmonary vascular resistance and right ventricular dysfunction.