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**Title:** A case of pulmonary capillary haemangiomatosis (PCH) diagnosed by lung biopsy

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**Body:** PCH is a rare cause of pulmonary hypertension. We report histologically confirmed PCH in a 18-yr-old male student. He presented with progressive exertional dyspnoea of two years' duration. He had no significant past medical or family history, never smoked and had no occupational or environmental inhalational history. Coarse crackles were present in both lungs. Biochemistry showed a raised LDH, otherwise normal. Arterial blood gas analysis in room air showed hypoxaemia (PaO2 43.2 mmHg) and hypocapnia (PaCO2 20.9 mmHg). Serum brain-type natriuretic polypeptide was elevated at 3840 pg/mL. Pulmonary function tests revealed normal forced vital capacity (FVC), forced expiratory volume in 1 s (FEV1) and FEV1/FVC, but diffusion capacity for carbon monoxide (DLCO) was markedly reduced at 4.83 mL/mmHg /min, 39% of predicted. ECG showed cor pulmonale and incomplete right bundle branch block. Echocardiography showed a pericardial effusion, marked right atrial and ventricular enlargement with severe tricuspid regurgitation and an estimated systolic pulmonary artery pressure of 80 mmHg. Right heart catheterisation was not performed. Chest radiography and high-resolution computed tomography demonstrated marked cardiomegaly, dilated main pulmonary artery, ground-glass opacities, thickening of the septal lines, and hilar and mediastinal lymphadenopathy. A surgical biopsy showed features of PCH. Diuretics and the intravenous prostacyclin iloprost were used with caution but to no avail. Lung transplantation was considered. He died soon thereafter. PCH is a rare condition of adults and children, related to veno-occlusive disease, which may be idiopathic or complicate connective tissue diseases.