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PULMONARY HYPERTENSION CAUSED BY PERIPHERAL PULMONARY ARTERY STENOSIS

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Objective: Introduction
Peripheral pulmonary artery stenosis (PPAS) is well described in children but is often underrecognized in the adult population. It may be present as an isolated congenital problem or as a part of Williams-Beuren syndrome (WBS), Alagille syndrome, Takayasu arteritis, Behçet disease or even develop after surgery for congenital heart disease involving pulmonary artery reconstruction.

Case report
We report the case of a 58-year-old woman who was admitted in our hospital because of progressive dyspnea lasting for three years. On examination her blood pressure was 130/80 mmHg, pulse rate 80/min, peripheral oxygen saturation of 95%, loud pulmonary component (P2) over pulmonary artery was present and a continuous murmur in the right paravertebral line. The patient walked 350 meters in the 6-minute walk test. Pulmonary function tests showed mild disorder of carbon monoxide diffusing capacity (DLCO). The concentration of brain natriuretic peptide was 328 ng/L. The erythrocyte sedimentation rate, hematologic and biochemical tests, antinuclear-, antiphospholipid- and vasculitis-antibodies were all within the physiologic range. Transthoracic echocardiography showed enlarged right ventricle with a normal left ventricular function and an estimated right

ventricular systolic peak pressure of 87 mm Hg. The ventilation/perfusion scan showed mismatch with a severe hypoperfusion of the whole right lung. The CT pulmonary angiography (CTPA) revealed dilated main pulmonary artery (4.5 cm in diameter) and proximal part of the right pulmonary artery (3 cm in diameter). It also showed focal narrowing of distal part of the right pulmonary artery (0.3 cm in diameter). Right heart catheterization showed severe pulmonary arterial hypertension with a mean pulmonary artery pressure (mPAP) of 53 mm Hg, pulmonary artery wedge pressure of 10mm Hg and pulmonary vascular resistance (PVR) of 8 WU. Percutaneous pulmonary artery stenting and balloon angioplasty were performed to relieve stenosis and alleviate pulmonary hypertension. The procedure was followed by improved vessel diameter (distal part of the right pulmonary artery was 9 mm in diameter afterwards), decreased mPAP up to 31 mmHg, decreased PVR up to 3.97 WU and symptomatic improvement.

Conclusion
In the adult population, PPAS may commonly be misdiagnosed as idiopathic PAH or chronic thromboembolic pulmonary hypertension (CTEPH). It is important to recognise patients with PPAS since pulmonary balloon angioplasty or stent angioplasty present the optimal treatment option which may decrease pulmonary artery pressure, pulmonary vascular resistance and lead to symptomatic improvement.