A 72-year-old lady was referred to our Pulmonary Hypertension Unit with WHO Class IV dyspnoea. Previous computed tomography pulmonary angiography had revealed chronic thrombo-embolism. The degree and distribution of disease was confirmed with gadolinium-enhanced cardiac magnetic resonance (CMR) pulmonary angiography (Panel A, see also Supplementary material online, Movie 1). The right ventricle was severely dilated with an ejection fraction of 32% by CMR. Functional testing demonstrated reduced 6 min walk distance (6MWD) of 225 m. The estimated right ventricular systolic pressure by echocardiography was 72 mmHg. Surgical pulmonary thrombo-endarterectomy (PTE) was performed. Eighteen months post-operation, both her right ventricular function and pulmonary pressures had returned to normal. The 6MWD had improved to 470 m (P < 0.01). Repeat CMR revealed normalization of right ventricular size with substantial improvements in pulmonary blood flow (Panel B, see also Supplementary material online, Movie 2).

Chronic thrombo-embolic pulmonary hypertension (CTEPH) is an increasingly recognized condition, and is the only cause of severe pulmonary hypertension which is potentially curable without the need to resort to lung transplantation. Once considered a rare condition, it is now recognized that up to 4% of patients with acute pulmonary embolus may develop CTEPH. Pulmonary thrombo-endarterectomy is the treatment of choice when performed in major centres by experienced surgeons and offers a surgical remedy to this severely disabling and often fatal disease process. Long-term outcomes following PTE are generally excellent with most patients experiencing significant improvements in haemodynamics and functional capacity, as illustrated by this case. Cardiac MR imaging and MR angiography are emerging as key investigations in quantification of right ventricular function and the assessment of pulmonary vascular anatomy.

Panel A. Pre-operative magnetic resonance pulmonary angiogram demonstrating vascular obstruction, webs, and stenoses and poor tissue perfusion due to chronic pulmonary thrombo-embolism.

Panel B. Follow-up magnetic resonance pulmonary angiogram showing substantial improvement of the pulmonary vasculature. CMR also showed normalization of right ventricular volume and function (not shown).

Supplementary material is available at European Heart Journal online.