A Fifty-one year old male presented 6 months after a pulmonary embolism involving the main pulmonary artery, with progressive exertional dyspnoea and decline in exercise tolerance, NYHA (New York Heart Association) functional class 3b. Right heart catheterisation revealed a mean pulmonary artery pressure of 60mmHg with a calculated pulmonary vascular resistance (PVR) of 14.7 woods units (1176 dynes). Radiological imaging confirmed a diagnosis of chronic thromboembolic pulmonary hypertension (CTEPH). The high PVR was deemed to be disproportionate to the radiologically evidenced disease burden and it was felt that the risk benefit ratio was against surgical intervention.

The patient was commenced on Sildenafil with initial clinical improvement. However he remained in NYHA class 3 with significant limitation to physical activity.

With increased experience it has become our policy to re-evaluate all CTEPH patients with distal disease to explore potential operability. After 5 years on Sildenafil 50mg TDS, NYHA class 4, a repeat right heart catheter revealed a mean pulmonary artery pressure of 65mmHg and PVR of 7.5 woods units (600 dynes). Disease distribution on imaging remained unchanged. Surgery was deemed appropriate.

The patient underwent Pulmonary Endarterectomy (PEA). Intra-operative findings included type 1 disease on the right with excellent clearance from all lobes and type 2 disease on the left with good clearance from the left lower lobe but no effective clearance from the left upper lobe and lingular. The post-operative course was unremarkable.

On routine follow up 3 months post PEA the patient was clinically well, in NYHA class 1 with a mean pulmonary artery pressure on right heart catheterisation of 25mmHg and PVR of 2.28 woods units (182 dynes).

Surgical management of CTEPH is potentially curative with improvement in clinical condition and restoration of normal or near normal haemodynamics. Over 5 years since the initial patient presentation the centre’s experience of PEA has grown from 250 to 615 cases. Experience in operative technique and patient selection have improved the outcome following PEA. This case demonstrates the importance of re-evaluation of CTEPH patients initially deemed to be inoperable as institutional experience grows.