1, 2, 3, and 4 years follow-up. Over this period were managed according to clinical need and NCG guidelines. N=124. Targeted therapy defined as Prostacyclin, endothelin receptor antagonists, and phosphodiesterase 5 inhibitors.

Results Changes at 1st and 2nd time points (median 3 months and 1 year) show statistically significant improvements in patient reported outcomes and objective measures. The NTproBNP improvement peaks at 3 years and begins to tail off at 4 years. The 6MWD improvement plateaus out between 2 and 3 years. The CAMPHOR symptom score shows significant improvement including year 3. The 4-year 6MWD does not achieve statistical significance compared to baseline. Further investigation revealed that between the third and fourth year 22 subject's follow-up was not performed; 3 died, 1 transferred to another centre, 18 are awaiting their 4-year follow-up.

Conclusion Objective and patient reported measures of change over time show improvements for CTEPH patients up to 3 years. After 3 years it becomes more difficult to determine a categorical change. More 6MW and CAMPHOR data for the 4 year time point is needed to determine if initial improvements are sustained after 3 years in this patient group, or if the improvement seen really has begun to tail off as the results here may suggest. The 4-year follow-up data for the remaining 18 patients will be available later this year.

S22 IMPROVED SYMPTOMS AND QUALITY OF LIFE AFTER PULMONARY ENDARTERECTOMY (PEA) IN PATIENTS WITH CHRONIC THROMBOEMBOLIC DISEASE (CTED) AND BORDERLINE PULMONARY HYPERTENSION (PH)

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Background Chronic thromboembolic pulmonary hypertension (CTEPH) is defined by a mean pulmonary artery pressure (mPAP) =25 mm Hg at right heart catheterisation in patients with multiple chronic/organised occlusive thrombi in the pulmonary arteries. PEA is potentially curable and is the treatment of choice for patients with CTEPH. There is a small group of patients with CTED and exertional dyspnoea but with a resting mPAP that does not fulfil criteria for PH in whom PEA may be considered. The outcome of PEA in these patients has not been previously assessed.

Objective To assess the post PEA functional and haemodynamic outcomes in patients with symptomatic CTED and borderline PH.

Methods Retrospective data were collected on patients with baseline mPAP of =25 mm Hg who underwent PEA at the UK and Ireland national referral centre between 2002 and 2010. Patients were reassessed 3 and 12 months after surgery. Right heart catheterisation was performed at baseline and 3 months post PEA.

Abstract S22 Table 1

	Baseline (n=16)	3 months post PEA (n=16)	12 months post PEA (n=11)
WHO class I/II/III/IV (%)	0/50/50/0	50/50/0/0	60/40/0/0
6MWT distance (m)	403±113	417±111	438±88*
6MWT min SatO ₂ (%)	88±7	91±8*	93±4*
mPAP (mm Hg)	22±3	16±3*	
Cardiac Index (l/m/m ²)	2±0.5	2.5±0.4	
PVR (dynes.s.cm ⁻⁵)	239±116	126±33*	
CAMPOR symptoms	10	2*	1*
Activity	6	3	0*
Quality of life	9	3*	1*

*p<0.05.

Results 558 patients underwent PEA at Papworth Hospital between 2002 and 2010. From those, 16 had symptomatic operable CTED and mPAP of =25 mm Hg. Mean age was 45±17 and 69% were female. All survived surgery and are currently alive at follow-up. The median length of stay was 13 days (IQR 6). Results are displayed in the Abstract S22 table 1 as mean ± SD. WHO class data are expressed as percentages. CAMPHOR (Cambridge Pulmonary Hypertension Outcome Review) is a disease specific quality of life questionnaire and scores are expressed as median.

Conclusion In this small series of patients with CTED and borderline PH, there was a significant symptomatic, haemodynamic and functional benefit from PEA at 1-year. Further research is required to assess the prognostic benefit in this population. We would like to acknowledge the national pulmonary hypertension centres in the UK and Ireland, and support by the Cambridge NIHR Comprehensive Biomedical Research Centre.

S23 ACCURACY OF CONTRAST ENHANCED MR LUNG PERFUSION COMPARED TO PERFUSION SCINTIGRAPHY IN DIAGNOSING CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Purpose The purpose of our study was to assess the diagnostic accuracy and reliability of contrast enhanced MR lung perfusion compared to perfusion scintigraphy in patients with CTEPH.

Methods and Materials Retrospective analysis of patients investigated for CTEPH who had MR lung perfusion, perfusion scintigraphy and CTPA performed within a time interval of 5 days. The MR images were acquired using a time resolved 3D spoiled gradient echo sequence. The sequence parameters: TE 1.1 ms, TR 2.5 ms, flip angle of 30°, FOV=48 cm², slice thickness of 5 mm, average of 32 slices and frame rate was 2 acquisitions per second. This was a breathhold sequence obtained after 0.05 ml/kg of Gadovist injection at 5 ml/s followed by a 20 ml saline flush. The subtraction images and positive enhancement dataset were analysed in the coronal plane. Four static views perfusion scintigraphy views were obtained. Final diagnosis of CTEPH or non-CTEPH was made at a multi-disciplinary meeting following detailed multi-modality assessment.

Results 27 patients had a diagnosis of CTEPH and 55 patients were diagnosed as non-CTEPH. In the CTEPH group, MRI lung perfusion diagnosed 26 patients as CTEPH and 1 patient with distal surgically inaccessible disease as non-CTEPH. In the non-CTEPH group, four patients were diagnosed as CTEPH and 48 patients were identified

Abstract S23 Figure 1

as non-CTEPH and three MRI were considered non-diagnostic. MR lung perfusion was not performed in 11 patients due to impaired renal function or patient unable to tolerate breathhold. None of the surgically accessible disease was missed with either modality. The MR lung perfusion had a sensitivity of 96%, specificity of 92%, PPV of 87%, NPV of 98% and accuracy of 94% for diagnosing CTEPH. Perfusion scintigraphy had a sensitivity of 93%, specificity of 90%, PPV of 83%, NPV of 96% and accuracy of 91%.

Conclusion Our results show that MR lung perfusion compared to 4-view static perfusion scintigraphy has an overall higher accuracy compared to perfusion scintigraphy and has a role in assessment of patients with suspected CTEPH.

S24 INCIDENCE OF PERSISTENT PERFUSION DEFECTS FOLLOWING PULMONARY EMBOLISM: IMPACT ON PULMONARY ARTERY SYSTOLIC PRESSURE AT 6 MONTHS

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Background The incidence of pulmonary hypertension (PH) after an episode of acute pulmonary embolism (PE) is thought to be up to 4%. The incidence of persistent clot without PH is less clear. We evaluated the incidence of persistent perfusion defects in patients followed up after PE and assessed the impact on pulmonary arterial pressure measured by echocardiography.

Methods The clinical pathway for outpatient follow-up of patients with PE includes a nuclear medicine ventilation-perfusion scan at 5 months post PE. When this is positive, an echocardiogram is requested to look for evidence of PH. A retrospective study of consecutive patients attending outpatient follow-up was carried out to determine the incidence of persistent perfusion defects and echocardiographic findings suggestive of PH.

Results Ninety-three patients were identified between February 2009 and July 2011 and their clinical data were studied retrospectively. 59 patients (63%) had persistent perfusion defects at 6 months. Of these 49 (83%) underwent echocardiography of which 12 had undetectable tricuspid regurgitation (TR). In the remaining 37, pulmonary artery systolic pressure (PASP) could be estimated from the TR velocity. Results are shown in Abstract S24 figure 1 and are skewed due to the presence of patients with PH. Median PASP (with IQR) was 32 mm Hg (26–39). The shaded area shows the 95% CIs for the normal PASP (28 ± 9.8 mm Hg, mean $\pm$ 2SD). Fourteen patients had estimated PASP above the normal range. Six patients were investigated with cardiac catheterisation

Distribution of Estimated PASP

Abstract S24 Figure 1 Distribution of estimated PASP.

and two were found to have chronic thromboembolic pulmonary hypertension (CTEPH). Borderline patients were kept under observation.

Conclusions Although the number of patients with CTEPH was in the expected range, the incidence of persistent perfusion defects following PE was surprisingly high in our cohort. Many of these patients fall in to a grey zone with borderline elevated pressures on echocardiography. The long-term outcomes of these patients and the physiological implications of their persistent perfusion defects are yet to be established and require further evaluation.

S25 INCIDENCE OF SURGICALLY TREATED PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION IN THE UK DURING THE LAST DECADE

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Introduction Pulmonary endarterectomy (PEA) is the treatment of choice for patients with proximal chronic thromboembolic pulmonary hypertension (CTEPH). The UK has a single centre performing this operation and the program became nationally funded since 2000. Patients are referred from seven specialist pulmonary hypertension centres.

Method All 625 patients treated with PEA from 2000 to 2010 were mapped according to their home postcode at the time of PEA surgery. Primary care trusts (PCT's) were assigned from these home postcodes. The incidence rate of each PCT and overall incidence were analysed. Mapinfo software was used to generate the referral maps.

Results The new patient incidence ranges from no referrals in 123 PCT's in 2000–2002 to 90 in 2008–2010. The most recent period shows highest referral rates for PEA. From our PEA data mapping analysis we have calculated that the incidence of operated patients was 0.4 million population in 2000 (n=22) and 2 per million population in 2010 (n=122).

Conclusion There has been a fivefold increase in PEA activity in the UK over the last decade. The analysis of our data are limited to surgical cases. The current incidence of PEA in the UK is already higher than historical estimation of the incidence of all CTEPH (0.1–0.5/million, Fedullo, *N Engl J Med* 2004). Since 30% of patients with CTEPH have distal disease distribution and some patients with proximal CTEPH do not proceed to surgery due to choice or comorbidities, the overall incidence of CTEPH is likely to be significantly higher than 2 per million/year and higher than previously suspected.

Abstract S25 Table 1 Patient Incidence/million population/period

Patient incidence in PCT/million	2000–2002 # of PCT's	2003–2004 # of PCT's	2005–2006 # of PCT's	2007–2008 # of PCT's	2008–2010 # of PCT's
0	123	136	124	113	90
0.1 to 2.4	16	13	10	18	12
2.5 to 4.9	40	29	37	37	50
5.0 to 7.4	10	9	19	15	20
7.5 to 9.9	2	4	0	3	11
10 to 35.6	3	3	4	8	11
Total incidence/ million	0.56	0.65	1	1	1.6

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