

**Abstract S50 Figure 1** CAMPHOR SCORE for individuals with CTEPH undergoing PEA. The median score for individuals is plotted at each timepoint. Groups are dichotomised by significant residual pulmonary hypertension ( $\geq 30$  mmHg) at 1 year follow up. Baseline (bl) is the pre-operative timepoint and yr1-yr5 are the post-operative 1–5 year longitudinal follow-up timepoints. Number of individuals at each time point: No residual/residual PH bl=482/255, yr1 469/245, yr2=294/174, yr3=116/62, yr4=99/40, yr5=76/28). Score ranges: Activity (0–30), Quality of Life (0–25), Symptoms (0–25).

PEA improved by 6,7 and 9 points for activity, QoL and symptoms respectively. The median difference for individuals having consecutive paired pre- and post-PEA scores also improved (median  $\pm$ IQR: activity  $4\pm 7$ ; QoL  $4\pm 8$ ; symptoms  $7\pm 8$ ). Patients were dichotomised into those with significant residual pulmonary hypertension (previously reported risk threshold of  $\geq 30$  mmHg,  $n=302$ ) and those without ( $n=569$ ). The improvement in CAMPHOR score was greater and more sustained in those without residual pulmonary hypertension (figure 1).

**Conclusion** PROs relating to activity, QoL and symptoms improve after PEA in CTEPH when evaluated by CAMPHOR score. The improvement is sustained up to 5 years in those without residual pulmonary hypertension. Ongoing work will examine the utility of PROs in addition to traditional clinical outcome measures.

**Acknowledgements** National Pulmonary Hypertension Centres UK and Ireland for referring patients considered for PEA.

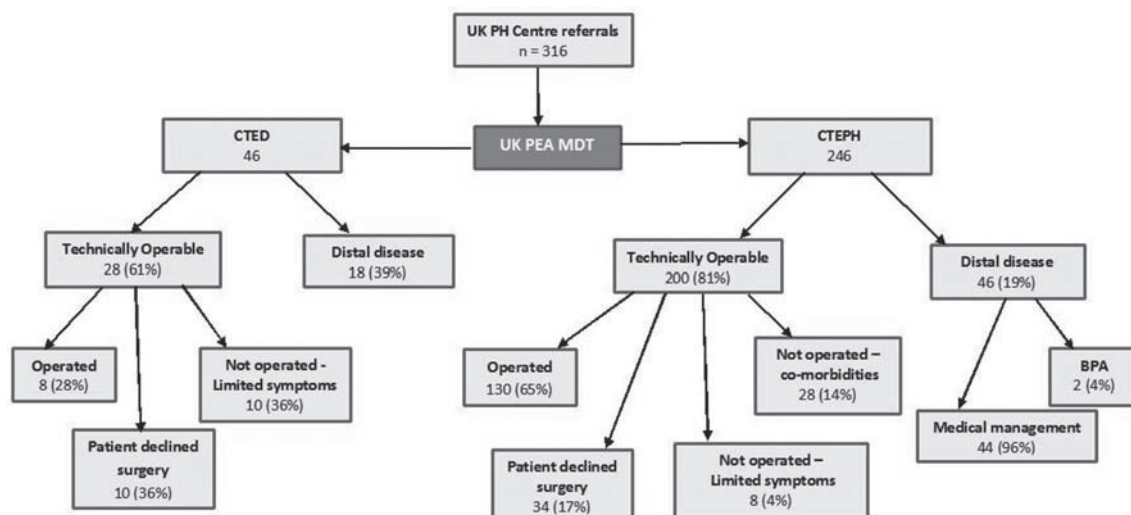
### S51 PATIENT PATHWAY MAPPING OF UK REFERRALS TO THE NATIONAL PULMONARY ENDARTERECTOMY MDT (JUNE 2015 – MAY 2016)

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**Introduction** Pulmonary endarterectomy (PEA) is the treatment of choice for Chronic Thromboembolic Pulmonary Hypertension (CTEPH) and can be offered to selected patients with Chronic Thromboembolic Disease without pulmonary hypertension (CTED). Our aim was to map the pathway of UK patients referred to the National PEA MDT and their onwards management.

**Methods** All consecutive individuals referred to the National PEA MDT at Papworth Hospital between June 2015 and May



**Abstract S51 Figure 1** Patient pathway flowchart from UK referrals to national PEA MDT.

2016 were included. Patients were divided into CTEPH/CTED groups and their pathways followed until July 2017.

**Results** Of the 316 referrals to the PEA MDT there were 246 cases of CTEPH, 46 of CTED and 24 with alternate diagnoses. Age range of referrals was 22–88 years (mean 59 years). 51% were male. The majority of CTEPH cases had technically operable disease (n=200, 81%) with 130 proceeding to PEA (65%). 53% of all CTEPH cases were operated on (figure 1). Median time from first MDT discussion to PEA was 230 days with a median surgical waiting list time of 115 days. 11 patients required ongoing targeted therapy post-operatively. Of the 70 individuals with an operable disease distribution who did not proceed to PEA, 34 patients chose to decline surgery and 36 were rejected by the surgical team due to co-morbid conditions (n=28) or limited symptoms (n=8). Two patients with distal CTEPH underwent Balloon Pulmonary Angioplasty (BPA). Remaining patients were medically managed with targeted therapy. Of CTED cases, 28 (61%) had an operable disease distribution with 8 (29%) proceeding to PEA. 17% of total CTED referrals were operated on.

**Conclusion** This patient pathway mapping is the first undertaken for any national CTEPH/CTED cohort. The majority of UK CTEPH cases suitable for PEA are operated on (65%). The duration of time between MDT referral and PEA is reflective of disease complexity and decision-making process. We observe a group of highly selected individuals with Chronic Thromboembolic Disease (CTED) without pulmonary hypertension who undergo PEA after careful consideration of their symptoms and operative risk.

#### S52 COMPUTED TOMOGRAPHY IN THE DIAGNOSIS OF LEFT HEART DISEASE IN PATIENTS WITH SUSPECTED PULMONARY HYPERTENSION

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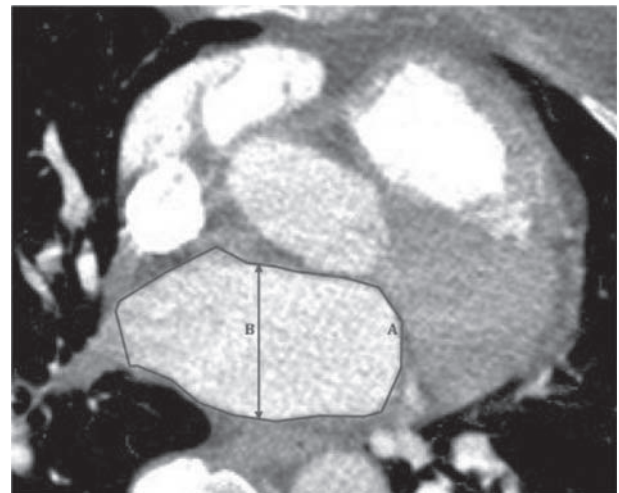
**Background** Identification of patients with left heart disease (LHD) as the cause of pulmonary hypertension is challenging, developing a tool that can identify these patients would reduce unnecessary referral for investigation at specialist centres and may reduce the burden of invasive investigations. The aim was to investigate the capability of computed tomography (CT)-derived metrics for the diagnosis LHD in a cohort of patients with suspected pulmonary hypertension.

**Methods** Patients with suspected pulmonary hypertension who underwent CT and RHC were identified. Derivation and validation cohorts were randomly constructed to derive and test a binary logistic regression model. All image analysis took place on PACS system blinded to patient's cardiac catheter data and diagnosis. CT measurements of the cardiac chambers and vessels were taken. LHD was defined by increased pulmonary arterial wedge pressure (PAWP)  $\geq 15$  mmHg. A second threshold of 18 mmHg defined more advanced LHD. Backward binary logistic regression in a derivation cohort identified a model for predicting Group 2 PH. This model was tested in the validation cohort and compared to individual CT derived variables using receiver operating characteristic curve analysis and chi-square.

**Results** The CT scans were from 66 different centres and 446 patients were identified, derivation cohort (n=235) and

validation cohort (n=211). Left atrial area was found to be most significant individual predictor of elevated PAWP, area under curve (AUC) 0.86,  $p < 0.001$ , the accuracy was higher for identification of PAWP  $\geq 18$ , AUC 0.87,  $p = 0.87$ ,  $p < 0.001$ . Derived regression models did not add diagnostic value AUC in validation cohort 0.87,  $p < 0.001$ . A limit for enlarged left atrial area was set at 27.5 cm<sup>2</sup>. This had sensitivity 65% and specificity 90% in predicting Group 2 PH using PAWP  $\geq 18$  mmHg as a threshold.

**Conclusions** CT derived left atrial area is a specific predictor of LHD in suspected pulmonary hypertension. Composite models did not increase diagnostic value. Left atrial area on CT may be a useful tool for diagnosing PH-LHD and may reduce unnecessary referrals to specialist PH centres and reduce the number of invasive investigations.



Abstract S52 Figure 1

## From diagnosis to treatment in interstitial lung disease

#### S53 MARKED SMALL AIRWAY DYSFUNCTION AND CONSEQUENT AIR-TRAPPING CHARACTERISE CHRONIC HYPERSENSITIVITY PNEUMONITIS (CHP) BUT NOT IDIOPATHIC PULMONARY FIBROSIS (IPF)

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**Introduction** CHP, the major differential diagnosis for IPF, has varied radiological features that can confound CT agreement in up to a third of cases of fibrotic interstitial lung disease presenting to multidisciplinary panels. Given that the pathological basis of CHP incorporates diffuse bronchiolitic abnormalities, we hypothesised that differences in small airway dysfunction and resultant air-trapping might help refine a multi-domain approach to distinguishing CHP from IPF.

**Methods** Analysis of lung function records of individuals with multidisciplinary diagnoses of CHP and IPF (n=118 in each group) with matched disease severity (% predicted FVC: 69.3  $\pm$  21.9 vs. 73.8  $\pm$  16.2 respectively,  $p = n/s$  and % predicted