Abstract P27 Table 1 Baseline characteristics and results of invasive and non-invasive investigations.

Gender M/F	11 (69)/5 (31)
Age [years] Median, IQR	53, 46.6–61.5
WHO class I/II/III	2/9/5
NT-proBNP [pg/ml] Mean $\pm$ SD	76.87 $\pm$ 81
6MWT Median, IQR	444, 366–521
• Distance [m]	1.5, 1–3.75
• Δ SpO ₂	
mPAP [mmHg] Median, IQR	20.5, 18–23
PVR [dyn·s·cm ⁻⁵] Median, IQR	158, 112–195.7
PAWP [mmHg] Median, IQR	10.5, 8–12
CO [L/min] Median, IQR	5.35, 4.1–5.8

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P28

CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: LONG TERM OUTCOMES IN SURGICAL AND NON-SURGICAL PATIENTS

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Introduction Chronic thromboembolic pulmonary hypertension (CTEPH) is commonly associated with a history of venous thromboembolism. Pulmonary endarterectomy (PEA) offers a potential cure in surgically accessible disease. However, a significant proportion of patients with CTEPH may not undergo surgery due to various reasons including disease distribution, comorbidities and patient choice. This group of patients have previously been considered to have a poor outcome although an international registry has recently reported on improved medium term outcomes in this patient population.

Aims and objectives To compare long term survival of patients with CTEPH undergoing pulmonary endarterectomy (CTEPH-surgical-operated), surgically accessible disease not undergoing pulmonary endarterectomy (CTEPH-surgical-not-operated), surgically inaccessible disease (CTEPH-non-surgical).

Methods Data was retrieved from hospital records and departmental database for consecutive, treatment-naïve patients with CTEPH diagnosed between 1st January 2001 and 30th November 2014 and followed up till 30th November 2015 at the Sheffield Pulmonary Vascular Disease Unit and collected in the ASPIRE registry. Patients with suspected CTEPH undergo systematic evaluation but formal pulmonary angiography is only performed when other imaging modalities such as CTPA, MR imaging and nuclear medicine imaging are non-diagnostic.

Results 592 patients, mean age ($\pm$ standard deviation), 65 $\pm$ 22 years, mean pulmonary arterial pressure 48 $\pm$ 13 mmHg and median pulmonary vascular resistance 480 $\pm$ 463 dynes/cm⁵ were identified and followed for 4.3 $\pm$ 3.2 years. 5 year survival was significantly ($p < 0.001$) better in CTEPH-surgical-operated ($n = 279$) at 82.9 $\pm$ 3.1% compared to CTEPH-surgical-not-

operated ($n = 206$) at 44.4 $\pm$ 5% (66.7 $\pm$ 9.1% patient choice, 39.4 $\pm$ 6% comorbidities) and 53.4 $\pm$ 5.8% in CTEPH-non-surgical ($n = 107$). Only 4% of the patients in our study were investigated with conventional pulmonary angiography. The median time to PEA surgery from diagnosis was 10.2 months and did not affect long term survival ($p = 0.52$).

Conclusions For operable patients with CTEPH pulmonary endarterectomy is associated with an excellent long term outcome, the long-term survival of patients with surgical disease who decline surgery is significantly better than historically reported and that a non-invasive multimodality imaging approach can be used to assess patients with suspected CTEPH. Furthermore there is no time from diagnosis to surgery which predicts outcome.

Surgical Operated	279	272	228	196	151	122
Surgical Nonoperated	206	166	110	79	57	44
Non surgical	107	96	84	63	49	39
Overall	592	534	422	338	257	205

Abstract P28 Figure 1 Kaplan-Meier estimates survival from date of diagnosis in CTEPH, surgical, operated, CTEPH, surgical, not-operated and CTEPH, non-surgical patients, log-rank test, $p < 0.001$

P29

EXERCISE INTOLERANCE IN CHRONIC THROMBOEMBOLIC DISEASE: EVALUATION, UNDERLYING MECHANISMS AND CLINICAL IMPLICATIONS

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Introduction Patients with chronic thromboembolic disease (CTED) without pulmonary hypertension commonly present with dyspnoea and fatigue. These symptoms limit physical function and impair quality of life. As resting haemodynamics in these patients are normal or near normal, stress testing may be a useful investigation to clarify mechanisms of functional impairment.

Methods We prospectively evaluated baseline characteristics of patients with CTED in a single referral centre between January 2015 and June 2016. Newly referred patients with suspected CTED underwent a standard assessment as delineated in international guidelines¹ with a minimum of 2 imaging modalities, resting and exercise right heart catheterisation (RHC) and additionally incremental cardiopulmonary exercise testing (CPET). All patients were assessed in a pulmonary endarterectomy MDT.

Abstract P29 Table 1 Exercise intolerance in chronic thromboembolic disease

Gender M/F number(%)	11 (69)/5 (31)
Age [years] Median, IQR	53, 46–61.5
Camphor score Median, IQR	9.5, 5–12.3
• Symptoms	3, 1.8–6.8
• Activity	5, 0.8–12
• QoL	
mPAP [mmHg] Median, IQR	20.5, 18–23
PVR [dyn·s·cm ⁻⁵] Median, IQR	158, 112–195.7
PAWP [mmHg] Median, IQR	10.5, 8–12
Cardiac Output [L/min] Median, IQR	5.35, 4.1–5.8
Cardiac Output fold increase on exercise*	2.4 ± 0.5
mPAP on exercise [mmHg]*	30, 25.8–32.8
TPR on exercise [WU]	2.6, 2.1–3.9
Peak VO ₂ [%pred.] Mean ± SD	90 ± 19.5
VE/VCO ₂ at AT Median, IQR	36, 31–44.9
Peak O ₂ pulse [% pred.] Median, IQR	84.5, 71–107

* Exercise at 40% of peak workload achieved during incremental CPET

Results Of 21 patients with confirmed CTED, 16 have completed the full assessment protocol (median age 53, 47–62). 14 (87%) were in functional class II/III. All patients had normal right ventricular function on echocardiography. Airway obstruction was present in 7 patients (44.5%). In majority of patients peak VO₂ and oxygen pulse were decreased and VE/VCO₂ at anaerobic threshold (AT) was increased (Table 1). CPET revealed 3 types of exercise limitation: combined cardiovascular and ventilatory limitation (n = 12), ventilatory limitation (n = 2) and limitation due to other reasons (n = 2). Peak oxygen consumption correlated with the symptoms domain of CAMPHOR (pulmonary hypertension specific quality of life measure) (p = 0.0242, R 0.56), cardiac output increase on exercise (p = 0.03, R 0.569) and VE/VCO₂ at anaerobic threshold (p = 0.012, R 0.608). Resting mPAP and PVR did not correlate with peak VO₂ or symptoms.

Conclusions We confirm the limited utility of resting measurements, including RHC in CTED for understanding exercise and functional limitation. CPET identified alternative causes for breathlessness and clarifies that patients with CTED are limited on exertion because of inability to increase cardiac output and hyperventilation.

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P30 AN EVALUATION OF THE USE OF QUALITY-OF-LIFE (QOL) SCORES IN PULMONARY ARTERIAL HYPERTENSION

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Introduction Pulmonary Arterial Hypertension (PAH) is a severe, progressive condition leading to increased pulmonary vascular resistance, right ventricular failure and death. PAH is associated with poor prognosis and WHO functional class (FC) is strongly predictive of mortality. Living with PAH has significant physical,

psychological and social impact on the lives of patients and carers. Symptoms of depression and anxiety are common and may contribute to poor quality of life (QOL) and social isolation.

Although QOL scores have been developed and validated in PAH, the psychological impact of living with PAH is often overlooked. Current guidelines advocate appropriate psychological and social support for patients, however, no formal recommendations exist currently to guide clinicians with regard to the timing and involvement of appropriately skilled professionals.

Methods QOL questionnaire data (Emphasis-10) was collected retrospectively from PAH patients attending routine appointments at PAH nurse and physician-led clinics over a 5 week period (June–July 2016).

Results

- 56 patients
- Median age: 62 (31–88)
- Sex: Male 17 (30%); Female 39 (70%)
- WHO FC: II: 24 (43%) III: 26 (46%) IV: 6 (11%)
- Aetiology: Familial: 2 (3%) Portal hypertension: 2 (3%) Connective tissue disease: 8 (14%) Congenital heart disease: 14 (25%) Idiopathic (IPAH): 19 (34%) CTEPH: 12 (21%)
- Emphasis 10 score (50): FC II: 15 (median), 1–35 (range). FC III: 31 (median), 2 – 50 (range). FC IV: 33 (median), 10–44 (range)

Conclusion Anxiety and depression are common in PAH and can lead to reduced physical and social functioning and poor QOL. The management of physical and psychological symptom burden is important for holistic patient care.

Patients with advanced PAH are known to have significantly impaired QOL and this is supported by our data. Whilst QOL scores are recommended in current guidelines and frequently used in clinical practice, there remains uncertainty around the identification and referral of suitable patients to colleagues skilled in psychological interventions and the role for supportive (palliative) care.

Our data shows a wide variation in QOL scores within each FC. This shows that psychological support should be considered on an individual patient basis and not only reserved for patients with a poor FC.

P31 PHA-UK LIVING WITH PULMONARY HYPERTENSION 2016 SURVEY

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Background Pulmonary Hypertension Association UK (PHA-UK) is the only charity in the UK especially for people affected by pulmonary hypertension (PH). To gain information on the patient journey and how PH impacts on daily living, surveys have been conducted by PHA-UK in 2007, 2010 and more recently in 2016. This paper reports the 2016 survey and provides comparisons with previous survey results.

Method A quantitative survey consisting of 4 sections regarding diagnosis, management, ongoing quality-of-life and treatment was available to complete online via PHA-UK's website or by hard copy sent to PHA-UK members and to patients on PH-specific targeted therapy.