worse outcome than those with PH-COPD (p = 0.015), and borderline worse outcome than patients with PH-ILD (p = 0.050), **Figure**. 48 of 94 patients were diagnosed with severe PH-RESP, defined at mPAP≥40 mmHg. WHO functional class (p = 0.036), TLCO (p = 0.019), RVEF (p = 0.033) were significant independent predictors of outcome in patients with severe PH-RESP.

Conclusion Patients with severe PH-RESP have a dire clinical outcome. RVEF is an independent predictor of adverse outcome in these patients and may be a powerful biomarker for use in clinical trials of targeted therapy in patients with pulmonary hypertension associated with lung disease, particularly given the unreliable performance of echocardiography in patients with advanced lung disease.

S121 THE UTILITY OF THE INCREMENTAL SHUTTLE WALKING TEST IN PULMONARY HYPERTENSION: RESULTS FROM THE ASPIRE REGISTRY

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Introduction The Incremental Shuttle Walk Test (ISWT) has been evaluated in a number of diseases and found to be a safe, reproducible test which correlates better with VO_2 max than the Six-Minute Walk Test (6MWT). We aimed to evaluate the utility of the ISWT as a prognostic indicator in pulmonary hypertension (PH).

Methods Data was retrieved for consecutive cases of PH diagnosed in our unit from 2001–2010, a cohort previously described.[1] ISWT was performed routinely as part of baseline assessment according to a modified protocol of Singh *et al*.[2] Data was analysed in 5 Groups according to the distance achieved based on ISWT level. A p-value of was deemed statistically significant.

Results 1002 of 1,344 patients diagnosed with PH underwent baseline ISWT within 3 months of cardiac catheterization and prior to pulmonary vascular therapy. Complete baseline data was available for 998 patients.

Kaplan-Meier analysis showed that increasing level of ISWT was associated with increased survival (Figure 1), including the PAH sub-group, with no ceiling effect.

ISWT distance correlated with WHO Functional Class, right atrial pressure, pulmonary vascular resistance, cardiac index, mixed venous oxygen saturation and percent predicted carbon monoxide diffusion (DLco) (p all ≤0.01). Multivariate Cox regression survival analysis including sex, body mass index, age, haemodynamic parameters and percent predicted DLco, demonstrated that ISWT distance was an independent predictor of survival.
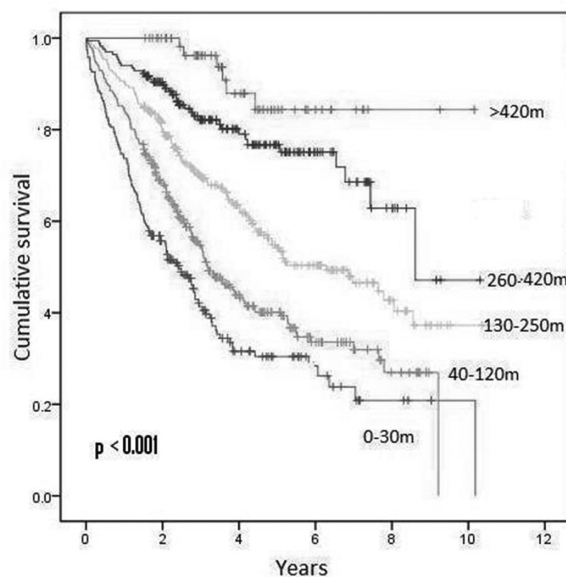
One year follow-up data was available for 397 patients. Kaplan-Meier analysis showed that ISWT level on treatment at 1 year was predictive of survival (p < 0.001). Survival was also superior in patients whose ISWT distance improved from baseline ≥30 m compared to those whose distance remained stable (-20 to +20 m) or declined by ≥30 m (p = 0.20).

Conclusion Baseline ISWT distance correlates with WHO functional class and pulmonary haemodynamics with no ceiling effect. It is an independent predictor of survival and change in ISWT predicts outcome. These features make it a viable alternative to the 6MWT in the assessment of patients with pulmonary hypertension, with a number of potential advantages.

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Figure I. Cumulative survival by Incremental Walk Test distance

0-30m	191	139	94	52	31	21	13	8	4	2
40-120m	297	251	181	119	74	51	29	18	9	1
130-250m	278	251	199	144	111	74	57	32	19	7
260-420m	169	159	134	98	69	49	29	20	9	3
>420m	63	63	63	42	29	16	10	6	2	2
Totals	998	863	671	455	314	211	138	84	43	15

Abstract S121 Figure 1 Cumulative survival by Incremental Walk Test distance

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S122 OUTCOME AFTER PULMONARY ENDARTERECTOMY (PEA): LONG TERM FOLLOW-UP OF THE UK NATIONAL COHORT

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Introduction Chronic thromboembolic pulmonary hypertension (CTEPH) is a life threatening condition that historically has a poor outcome with supportive medical treatment. Pulmonary endarterectomy (PEA) is the treatment of choice and offers the only chance of cure. Data on the predictors of long term survival after PEA are limited. We analysed the long-term data from the UK PEA cohort.

Method All patients who underwent a PEA for CTEPH at Papworth hospital between January 1997 and December 2012 were included. Pre- and post-operative data on haemodynamics, exercise capacity, functional class and targeted PAH therapies taken were obtained from databases of the UK PH centres. The NHS spine summary care record tracking system was used for survival data and causes of death from the England and Scotland General Register Offices. The causes of death were further classified into 4 groups: 1. Post operative, 2. Right ventricular failure away from operative period, 3. Related to anticoagulation, 4. Unrelated to CTEPH e.g. malignancy.

Results 880 patients underwent PEA over the 15 year period. The mean age was 57 (range 15–84) and 53% were male. 89% were in WHO functional class 3 or 4 before surgery with an mean mPAP of 47 mmHg and PVR of 830 dynes. Post surgery 84% of patients

Abstract S122 Table 1 First year post PEA haemodynamics and exercise capacity predict long term risk of death

Predictor value	Value	Hazard ratio	95% confidence intervals
6 minute walk distance (m)	110	3.03	1.82–5.04
	230	1.74	1.35–2.24
	350	Reference	
	470	0.57	0.45–0.74
	590	0.33	0.20–0.55
Mean pulmonary artery	15	0.67	0.55–0.83
pressure (mmHg)	25	Reference	
	35	1.49	1.21–1.83
	45	2.21	1.46–3.36
	55	3.29	1.76–6.16
Cardiac index (L/min/m^2)	1.5	1.60	1.00–2.56
	2	1.26	1.00–1.60
	2.5	Reference	
	3	0.79	0.62–1.00
	3.5	0.63	0.39–1.00
	4	0.49	0.24–1.00
Pulmonary vascular resistance	50	0.51	0.37–0.70
(Dynes/sec/cm^5)	250	Reference	
	450	1.95	1.43–2.67
	650	3.81	2.03–7.14
	850	7.44	2.90–19.08

were in WHO functional class 1 or 2 and there was a reduction in the mean mPAP to 27 ± 9 mmHg and PVR to 286 ± 198 dynes/sec/cm^5 by 12 months (p).

Conclusion There was prolonged haemodynamic improvement but targeted therapy was used in 23% of patients with a mean follow-up of 4.3 years. The 10-year survival was 72% with mortality predominantly in the peri-operative period and later due to causes unrelated to CTEPH.

Novel approaches to rehabilitation and exercise therapy in COPD

S123 DOES EXERCISING WITH DOMICILIARY NON-INVASIVE VENTILATION (NIV) IMPROVE QUALITY OF LIFE (QOL) IN PATIENTS WITH SEVERE CHRONIC OBSTRUCTIVE PULMONARY DISEASE (COPD)?

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Introduction and objectives Patients with severe COPD experience breathlessness leading to exercise limitation impacting on QoL. Pulmonary rehabilitation (PR) can improve QoL, but those with the severest disease are frequently hospitalised and cannot readily access PR. Previous studies have trialled positive pressure as a means of relieving ventilatory load, allowing more severe COPD patients to exercise.[1] Studies have assessed mixed pathology or stable COPD patients.[2,3] In this study, we have assessed patients with severe COPD admitted to hospital with Type 2 respiratory failure and acidosis treated with acute NIV.

Methods 18 patients (11 female), recruited during admission were randomised into 3 groups. Group 1 received standard hospital physiotherapy care; Group 2 exercised on NIV (Trilogy 100, Philips-Respironics) with a mean pressure support of 10 cmH$_2$O twice weekly during admission; Group 3 exercised on NIV twice weekly

Abstract S123 Table 1 Results for SGRQ and LCADL from baseline to M3

Group		1 (n = 6)	2 (n = 5)	3 (n = 4)
	Baseline	81.06 ± 9.27	74.27 ± 20.94	69.54 ± 14.35
SGRQ	M3	73.89 ± 16.20	80.11 ± 13.15	50.25 ± 24.55
Mean Change		-7.17	-5.84	-19.29
	Baseline	53.00 ± 13.07	47 ± 9.49	49.83 ± 15.69
LCADL	M3	51.67 ± 7.65	56.6 ± 5.50	40.75 ± 13.94
Mean Change		-1.33	+9.6	-9.08

during hospital admission and continued this at home for 3 months post-discharge. Exercising included weights, pedal cycling and walking. QoL was assessed using the St Georges Respiratory Questionnaire (SGRQ) and the London Chest Activities of Daily Living Questionnaire (LCADL). Mean changes in total scores for SGRQ and LCADL were compared between baseline and at 3 months (M3). Data are mean±SD or mean (range).

Results The group (n = 18) age was 66.5 years [46–97], FEV$_1$: 25% predicted [9–51%] and MRC score 3 [1–4]). 3/18 patients died during the study. The results are presented in Table 1.

Conclusion Patients exercising with NIV, in hospital and at home twice weekly (Group 3) showed the greatest improvement in QoL, compared to the other two groups. The use of NIV during exercise at home may assist patients unable to access pulmonary rehabilitation.

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S124 EFFECTS OF TWO ADAPTED PHYSICAL ACTIVITY TRAINING PROGRAMS ON PULMONARY FUNCTIONALITY AND EXERCISE CAPACITY IN PATIENTS AFFECTED BY CHRONIC OBSTRUCTIVE PULMONARY DISEASE

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Introduction It has been reported the efficacy of adapted physical activity (APA) in exercise capacity improvement.[1] Nevertheless, there is still no consensus on training modalities and intensities to be prescribed in patients affected by chronic obstructive pulmonary disease (COPD). The aim of the study was to assess the effects of two 16 weeks APA training programs (endurance vs endurance +strength) on respiratory parameters (FVC%, FEV1%, FEV1/FVC %) and exercise capacity (V'O$_2$ peak) immediately after APA training program (first follow up: FU1) and after six months (second follow up: FU2)

Methods Sixty five COPD patients were randomly assigned to endurance training (ET) or to endurance + strength training (EST). All Patients underwent 3 sessions per week. For ET, as upper intensity training limits were considered 40–50% heart rate reserve; for EST training limits were considered 40–50% heart rate reserve and 50% 1RM.[2] Before training programs, at FU1 and at FU2, all patients underwent: clinical assessment, respiratory functionality tests, maximal cardiopulmonary test. Repeated measures ANOVA