patients with higher rates of CTEPH with less severe pulmonary haemodynamic changes.

S119 LEFT VENTRICULAR DYSFUNCTION INFLUENCES SURVIVAL IN CONNECTIVE TISSUE DISEASE ASSOCIATED PULMONARY ARTERIAL HYPERTENSION BUT NOT IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION

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Background Connective tissue disease – associated pulmonary artery hypertension (CTD-PAH) has a worse prognosis compared with idiopathic pulmonary arterial hypertension (IPAH). We investigated the prognostic significance of left and right cardiac dysfunction in IPAH and CTD-PAH.

Methods and results Between 2003 and 2011, patients with a new suspected diagnosis of pulmonary hypertension underwent diagnostic assessment including cardiac magnetic resonance (CMR) imaging and right heart catheterization (RHC). 138 patients fulfilled the criteria for pulmonary arterial hypertension, of which 74 were diagnosed with IPAH and 38 were diagnosed with CTD-PAH. At baseline, there was no significant difference in age, functional class, lung function or six-minute walk distance between the two groups. At CMR, both groups had right ventricular (RV) dilatation and impaired RV systolic function, but well preserved left ventricular (LV) ejection fraction. Patients with IPAH had greater right ventricular hypertrophy than those with CTD-PAH (VMI 1.16 v 0.99, p = 0.03). Left atrial volume, a marker of LV diastolic dysfunction, was lower in IPAH than CTD-PAH (23 v 33 ml/m^2, p < 0.0001). At RHC, mean pulmonary artery pressure was higher in IPAH than CTD-PAH (50 v 43 mmHg, p = 0.01).

There was no difference in the distribution of initial disease-targeted therapies between the groups. Survival was better in IPAH than in CTD-PAH (p = 0.03), with rates of 83% at 1 yr and 74% at 3 yrs in IPAH, but 75% at 1 yr and 53% at 3 yrs in CTD-PAH. Poor baseline right ventricular function was associated with reduced survival in both conditions. However, poor left ventricular function, as measured by left ventricular stroke volume index (LVSVI), only influenced survival in CTD-PAH (p = 0.002) and not in IPAH (p = 0.21).

Conclusions Poor LVSVI at diagnosis is associated with impaired survival in CTD-PAH but not IPAH. Intrinsic LV problems, particularly diastolic dysfunction, may contribute to the excess mortality in CTD-PAH.

Figure (A)

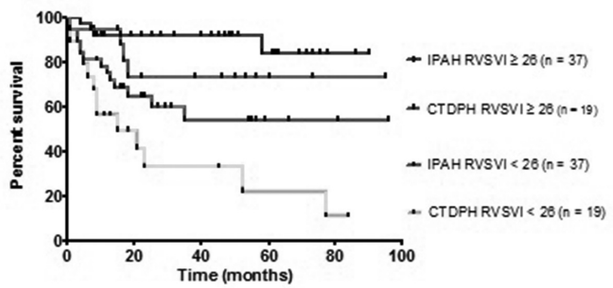
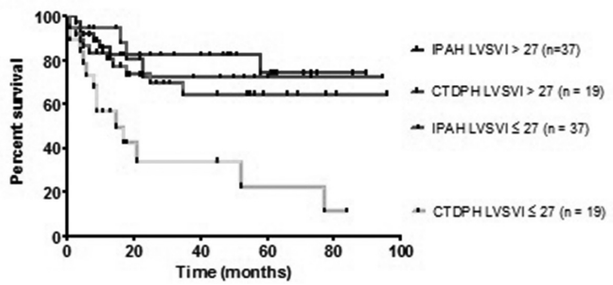


Figure (B)



Abstract S119 Figure 1 Survival rates of patients stratified according to disease, and baseline RVSVI(A) or LVSVI (B). Poorer right ventricular function (inframedian RVSVI) was associated with impaired survival in both IPAH and CID-PAH (A). Poorer left ventricular function (inframedian LVSVI) was associated with impaired survival in CTD-PAH (p = 0.002), but not in IPAH(p = 0.21)(B)
RVSVI, right ventricular stroke volume index;LVSVI, left ventricular stroke volume index

S120 RIGHT VENTRICULAR DYSFUNCTION IN PULMONARY HYPERTENSION WITH COMBINED PULMONARY FIBROSIS AND EMPHYSEMA SYNDROME

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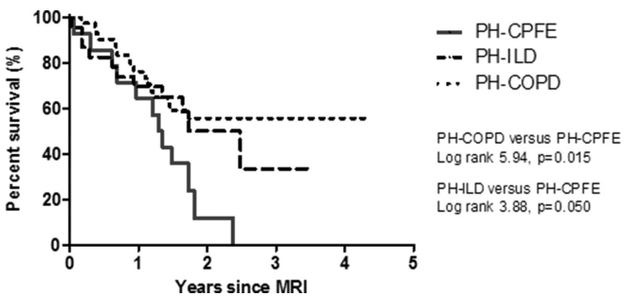
Introduction Recent studies have suggested that the coexistence of emphysema and fibrosis alters clinical outcome. The aim of this study was to investigate the comparative clinical characteristics, pulmonary function, haemodynamics and right ventricular (RV) function and outcome in patients with pulmonary hypertension associated with combined pulmonary fibrosis and emphysema (PH--CPFE), chronic obstructive pulmonary disease (PH-COPD) and interstitial lung disease (PH-ILD).

Methods In 79, incident patients with pulmonary hypertension associated with respiratory disease, cardiovascular magnetic resonance imaging was performed at 1.5T. Emphysema and fibrosis were scored on high resolution computed tomography scans. Demographic data, lung function tests and right heart catheterisation were also performed.

Results Patients with pulmonary hypertension associated with combined pulmonary fibrosis and emphysema syndrome had lower right ventricular ejection fraction when compared to both patients with PH-COPD and PH –ILD (p < 0.05). At Kaplan-Meier analysis, patients with PH-CPFE patients had significantly



Abstract S120 Figure 1 Kaplan Meier plot showing survival of patients with pulmonary hypertension associated with combined fibrosis and emphysema (PH-CPFE) in comparison to pulmonary hypertension interstitial lung disease (PH-ILD) and pulmonary hypertension associated with COPD (PH-COPD)

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.

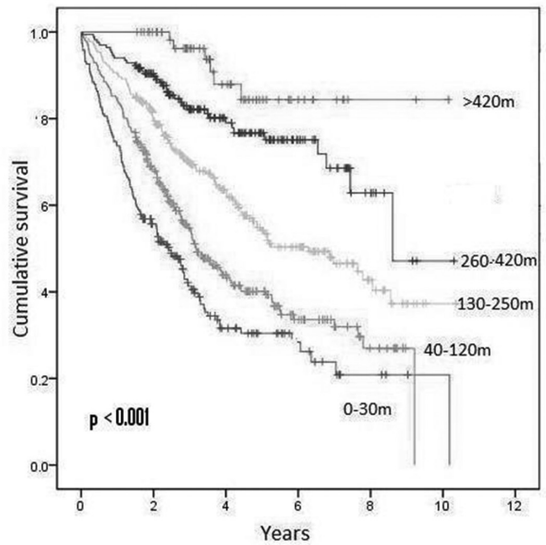
Kaplan-Meier analysis showed that increasing level of ISWT was associated with increased survival (Figure 1), including the PAH sub-group, with no ceiling effect.

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.

Figure 1. 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