**Abstract P29 Table 1** Exercise intolerance in chronic thromboembolic disease

| | |
|---|---|
| **Gender** M/F number(%) | 11 (69)/5 (31) |
| **Age** [years] Median, IQR | 53, 46.6–61.5 |
| **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 $VO_2$ and oxygen pulse were decreased and VE/$VCO_2$ 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/$VCO_2$ at anaerobic threshold (p = 0.012, R 0.608). Resting mPAP and PVR did not correlate with peak $VO_2$ 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.

## REFERENCE

1 Galie N, *et al*. ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *EHJ* 2015;**58**(1):e129–e152.

## 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 PAH-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.

**Results** 551 responses were received. Participant mean age was 58.3 ± 16.6 years and age at diagnosis was 52.3 ± 18.8 years. 49% of patients had symptoms for >6 months before going to see a doctor and 22% were seen by 4 or more doctors before diagnosis. Time from first symptoms to diagnosis was >1 year in 49% with 31% of patients admitted as an emergency because of their symptoms. After diagnosis, 48% see a specialist at least every 6 months, 87% at least every 12 months. 62% think the support they receive is excellent and 26% good. 90% thought it was better to travel to a Specialist PH Centre rather than to be under the care of a non-PH specialist at a more local hospital.

**Discussion** This survey found that care of patients with PH is generally good or excellent and patients were keen to travel to Specialist PH Centres for their care. The early symptoms of pulmonary hypertension can be mild and are common to many diseases so it is often a lengthy process to arrive at the diagnosis. Compared with the previous survey the percentage of patients seeing >4 doctors before diagnosis was reduced (22% *vs* 47%) suggesting an increasing awareness of pulmonary hypertension amongst physicians. However, 49% of patients had symptoms for >6 months before presenting, which has not improved since previous surveys. As earlier diagnosis of patients results in better long-term survival, further work should be undertaken to continue to raise awareness in the UK of pulmonary hypertension.

## P32 A NEW NURSE LED P.E CLINIC 2015

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In 2015 Dr R Limbrey (Respiratory Physician) and Sr S.Goodman (CNS Pulmonary Vascular Service) set up a Nurse Led (new patient) Pulmonary Embolism (P.E) clinic to support increasing demand on the traditional model of consultant care.

We aim for a CNS review of all new patients with P.E at 3 weeks following diagnosis, and provide follow up for 2 years. Previously a CNS led follow up clinic was in place.

We aim to provide our patients with early high quality education and information to reduce anxiety, improve physical functioning and quality of life. We suggest that this will reduce perceived ongoing symptoms and lessen follow up requirement.

Referral is made electronically. The referral system has been developed as a learning opportunity to enable medical referrers to identify provoking factors and quantify risk associated with the event. The data obtained from referrals provides support for service provision.

In 2015, 260 patients were reviewed in the Nurse led clinic, 95 of these were new referrals.

Of the 95 new referrals to the service 54 (57%) were provoked by an identifiable transient provoking factor. ESC (2014). Of these 42 (78%) also had persistent risk factors by ESC

Unprovoked group:

The remaining 41 (43%) had no identifiable transient provoking factor. Of these 32 (78%) did have persistent risk factors as per ESC 2014.

We note that 44% of all identified PE patients in this cohort, provoked and unprovoked, had a BMI $\geq$ 30kg/m$^2$

In the provoked group 16 new patients were reviewed and discharged after an average of 2.62 appointments per person with an additional telephone review planned at the 2 year point.

In the unprovoked group 12 patients have been discharged to date with an average of 3.3 appointments per person with an additional telephone review planned for the 2 year mark.

The remainder of these patients can reasonably be expected to be discharged from follow up with similar levels of review.

Feedback from our patients has been overwhelmingly positive, most commonly expressed as a significant reduction in anxiety and improvement in quality of life.

**Abstract P32 Table 1**

| No of patients seen in Nurse led P.E clinic 2015 by identified transient provoking factors (new patients) | No of patients 54 in total | Persistent provoking factors identified | No of patients |
|---|---|---|---|
| Post operative | 17/54 | Obesity | 26/54 |
| Travel | 11/54 | Hypertension | 12/54 |
| Immobility | 8/54 | Previous VTE | 6/54 |
| Pneumonia | 7/54 | A.F | 2/54 |
| Oral Contraceptive Pill | 4/54 | Age > 80yrs | 2/54 |
| Pregnancy | 3/54 | Known thrombophilia | 2/54 |
| Cancer ( identified prior to diagnosis of VTE) | 1/ | Tamoxifen | 0/54 |
| Other | 3/54 | Diabetes | 1/54 |

Unprovoked group:

| Persistent risk factors identified in Nurse led P.E clinic 2015 in Unprovoked group | No of patients |
|---|---|
| **Obesity** | 16/41 |
| **Hypertension** | 13/41 |
| **Previous VTE** | 7/41 |
| **Age > 80 yrs at time of event** | 8/41 |
| **Diabetes** | 5/41 |
| **Known Thrombophilia** | 2/41 |
| **Atrial Fibrillation** | 1/41 |
| **Tamoxifen** | 1/41 |

## P33 PATIENTS WITH PULMONARY ARTERIOVENOUS MALFORMATIONS AND HEREDITARY HAEMORRHAGIC TELANGIECTASIA REPORT FORCED EXPIRATORY MANOEUVRES DURING PULMONARY FUNCTION TESTS PROVOKE NOSEBLEEDS AND MIGRAINES

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**Introduction and objectives** Forced expiratory manoeuvres during lung function testing produce major pressure swings that are often overlooked by referring clinicians. Standard tests use a noseclip to prevent air leakage through the nose. Our goal was to examine how often the tests caused clinical sequelae such as nosebleeds in people with abnormal nasal and pulmonary vasculature due to hereditary haemorrhagic telangiectasia (HHT).

**Methods** With ethical approval, self-reported migraine features and exacerbations were examined in HHT subjects with and