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

<table>
<thead>
<tr>
<th>Gender</th>
<th>MF</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age [years]; Median, IQR</td>
<td>53, 46.6–61.5</td>
</tr>
<tr>
<td>WHO class I/II/III</td>
<td>2/9/5</td>
</tr>
<tr>
<td>NT-proBNP [pg/ml] Mean ± SD</td>
<td>76.87 ± 81</td>
</tr>
<tr>
<td>6MWT Median, IQR</td>
<td>444, 366–521</td>
</tr>
<tr>
<td>• Distance [m]</td>
<td>1.5, 1–3.75</td>
</tr>
<tr>
<td>• % SpO2</td>
<td>9.3</td>
</tr>
<tr>
<td>mPAP [mmHg]; Median, IQR</td>
<td>20.5, 18–23</td>
</tr>
<tr>
<td>PVR [dyn·s·cm−5]; Median, IQR</td>
<td>158, 112–195.7</td>
</tr>
<tr>
<td>PAWP [mmHg]; Median, IQR</td>
<td>10.5, 8–12</td>
</tr>
<tr>
<td>CO [L/min]; Median, IQR</td>
<td>5.35, 4.1–5.8</td>
</tr>
</tbody>
</table>

REFERENCE

P28 CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: LONG TERM OUTCOMES IN SURGICAL AND NON-SURGICAL PATIENTS

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 has 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 underwent 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 (± standard deviation), 65 ± 22 years, mean pulmonary arterial pressure 48 ± 13 mmHg and median pulmonary vascular resistance 480 ± 463 dynes/sec/cm−5 were identified and followed for 4.3 ± 3.2 years. 5 year survival was significantly (p < 0.001) better in CTEPH-surgical-operated (n = 279) at 82.9 ± 3.1% compared to CTEPH-surgical-not-operated (n = 206) at 44.4 ± 5% (66.7 ± 9.1% patient choice, 39.4 ± 6% comorbidities) and 53.4 ± 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.
**Abstract P29 Table 1**

<table>
<thead>
<tr>
<th>Gender</th>
<th>M/F number(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age [years] median, IQR</td>
<td>53, 46.6–61.5</td>
</tr>
<tr>
<td>Newborn (earliest age)</td>
<td>4, 4.2–12.8</td>
</tr>
<tr>
<td>Camphor score median, IQR</td>
<td>9.5, 5–12.3</td>
</tr>
<tr>
<td>• Symptoms</td>
<td>3, 1.8–6.8</td>
</tr>
<tr>
<td>• Activity</td>
<td>5, 0.8–12</td>
</tr>
<tr>
<td>QOL</td>
<td></td>
</tr>
</tbody>
</table>

| 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 |
| PVR on exercise [WU] | 2.6, 2.1–3.9 |
| Peak VO₂ [%pred.] mean ± SD | 90 ± 19.5 |
| VE/VCO₂ at AT, median, IQR | 36, 31–44.9 |
| Peak O₂ 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.

**REFERENCE**


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**Abstract P30**

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

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10.1136/thoraxjnl-2016-209333.173

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 3 week period (June–July 2016).

**Results**

- 56 patients
- **Median age:** 62 (31–88)
- **Sex:** Male 17 (30%); Female 39 (70%)
- **WHO FC:** II: 15 (median), 1
- **Aetiology:** Familial: 2 (3%) Portal hypertension: 2 (3%)
- **Emphasis 10 score (50):** FC II: 15 (median), 1

**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.

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**Abstract P31**

**PHA-UK LIVING WITH PULMONARY HYPERTENSION 2016 SURVEY**

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10.1136/thoraxjnl-2016-209333.174

**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.