**Background:**

Pulmonary arterial hypertension (PAH) and PH secondary to ILD are the leading cause of death and late disease morbidity in SSC. Patients with SSC-PAH have a blunted response to modern therapies by measures of exercise capacity, functional class and survival. The natural history of SSC-PAH/PH and its response to treatment is thus still unknown. PHAROS is a multicenter effort by members of the Scleroderma Clinical Trials Consortium. Its objective is to determine the timeline of progression from pre-PAH/PH to diagnosable PAH/PH; to determine the natural history of the disease and clinical worsening event rate. Earlier diagnosis of SSc-PAH/PH would offer information relevant to design of prevention trials and lead to strategies that alter the natural history of the disease and clinical worsening of PAH/PH.

**Methods:**

PHAROS is the prospective observational study of patients with SSC who have PAH/PH < 6 months or who meet criteria for pre-PAH: SPAP by echocardiogram > 35mmHg, DLco<55% of predicted or ratio predicted %FVC/%DLco>1.6. Data are collected bi-annually and include demographics, review of organ involvement, Rx history, medical event history and functional assessment (SHAQ-DI, Borg index, UCSD Dyspnea questionnaire, SF36). Objective data gathering includes: PFT, echocardiogram, 6MWT and right heart catherization (RHC) parameters. Data are electronically entered by both investigators and subjects.

**Patients:**

- Ongoing enrollment
- Inclusion criteria:
  * patients with SSC who have PAH/PH < 6 months or
  * patients who meet criteria for pre-PAH/PH:SPAP
  1. echocardiogram > 35mmHg
  2. DLco<55% of predicted
  3. %FVC/%DLco>1.6

**Methods:**

PHAROS is a prospective observational study. Data are collected bi-annually and entered electronically both by investigators and patients in a secure web-based system.

**General data include:**

- Demographics
- Review of organ involvement
- Rx history
- Medical event history
- Functional assessment (SHAQ-DI, Borg index, UCSD Dyspnea questionnaire, SF36)

**Objective data include:**

- PFT
- Echocardiogram
- 6MWT and right heart catherization (RHC) parameters

**Results:**

- 12 SSC centers are actively enrolling and data are preliminary
- Of 139 enrollees, 29 had established PAH/PH, 102 have suspected pre-PAH- PH and 8 withdrew consent

<table>
<thead>
<tr>
<th>PAH/PH</th>
<th>Mean (n=139)</th>
<th>Mean (n=102)</th>
<th>Mean (n=8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAH/PH</td>
<td>51.9 (100%)</td>
<td>45.2 (89%)</td>
<td>33.4 (20)</td>
</tr>
<tr>
<td>PAH/PH</td>
<td>1.78 (65%)</td>
<td>1.39 (22%)</td>
<td>0.95 (12)</td>
</tr>
</tbody>
</table>

Early observations include information about treatment effect on PAH/PH. Of 20 subjects initially on bosentan, 1 progressed to epoprostenol, 2 stopped it secondary to AEs and 10 required additional Rx (sildenafil 8 and iloprost 2).

**Conclusions:**

- PHAROS prospectively gathers data to develop tools for early PAH/PH diagnosis, to characterize time to diagnosis of PAH/PH and clinical worsening event rate.

- Earlier diagnosis of SSC-PAH/PH would offer information relevant to design of prevention trials and lead to strategies that alter the natural history of disease.