High altitude living and its relationship with Pulmonary Arterial Hypertension (PAH) symptom severity and clinical characteristics: The Pulmonary Hypertension Association Registry (PHAR).

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Background

• Pulmonary Hypertension is a progressive and potentially fatal disease that often presents with exertional dyspnea and functional limitation.
• Pulmonary arterial hypertension (PAH) is a subtype of pulmonary hypertension that primarily affects the pulmonary arterial circulation. PAH is defined as a mean resting pulmonary artery pressure of 25 mm Hg or above, pulmonary capillary wedge pressure (PCWP) below 15 mm Hg, in the absence of significant left heart disease, chronic lung disease, or venous thromboembolism ¹ ²
• Individuals living at high altitude are exposed to lower barometric pressure and hypoxic hypoxemia which can lead to pulmonary vascular remodeling with chronic exposure ³
• We sought to examine the relationship between high altitude living and PAH.

Methods

• Forty-two centers across the United States screened and enrolled adult patients (age ≥ 18) in the Pulmonary Hypertension Association Registry (PHAR) who met the definition of group I PAH.
• All PAH subgroups were included. Patients with chronic thromboembolic pulmonary hypertension (CTEPH) where excluded.
• Upon enrollment patients and staff completed a baseline questionnaire reporting clinical characteristics, hemodynamic data, patient demographics and patient reported quality of life metrics.
• Patients were divided into two groups; defined as high elevation living (>4000 ft) and low elevation living (<4000 ft) based on provided zip codes.

Results

• Among 1021 patients enrolled in the registry, 851 met the diagnostic definition for PAH and had elevation data available with 776 living at low altitude and 75 living at high altitude.
• Subjects at high elevation had a 6-minute walk distance (6MWD) 26 meters greater than those at low elevation (p=0.08), despite having a pulmonary vascular resistance (PVR) that was 2.1 Wood Units (WU) higher (p=0.002). Additionally, those at high elevation had 3.2 times greater odds of using supplemental oxygen (p=0.001).

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<thead>
<tr>
<th>Response</th>
<th>Unadjusted</th>
<th>Adjusted</th>
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<tbody>
<tr>
<td>Difference</td>
<td>CI</td>
<td>p-value</td>
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<tr>
<td>Six minute walk distance (m)</td>
<td>42</td>
<td>(8.4, 75.6)</td>
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<tr>
<td>Mean pulmonary artery pressure</td>
<td>4.5</td>
<td>(1.2, 7.8)</td>
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<td>Right arterial pressure</td>
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<td>(-1.3, 1.6)</td>
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<td>PVR (Wood units)</td>
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<td>(1.2, 3.9)</td>
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<td>Emph 10 score</td>
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<tr>
<td>Physical health score</td>
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<td>(-1.5, 1.8)</td>
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<tr>
<td>Odds Ratio</td>
<td>CI</td>
<td>p-value</td>
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<tr>
<td>Use of supplemental oxygen</td>
<td>2.2</td>
<td>(1.3, 3.5)</td>
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Future Directions

• More study is needed to further evaluate the relationship between chronic high-altitude exposure and PAH development and progression.

Conclusions

• Patients presenting with PAH who live at high altitude have a higher PVR, and are more likely to need supplemental oxygen.
• Despite their higher PVR, they appear to have a better 6MWD, which could be related in part to better overall conditioning.
• It is interesting to speculate that there might exist a “high-altitude phenotype” of PAH.

References