Primary pulmonary hypertension (PPH) is a disease of unknown origin. It is characterised by a progressive increase in pulmonary arterial pressures. Individual mortality is associated with variables of right ventricular dysfunction. The mean survival of patients with severe PPH is <3 years without appropriate medical treatment. To our knowledge, there are no long term reports on the spontaneous course of mild PPH over a period of three decades in the literature.

We present a stable long term follow up of a young patient with PPH without specific medical treatment over a period of 30 years.

**CASE REPORT**

Thirty years ago, a now 39 year old woman was evaluated by right heart catheterisation for the first time. At that time, the 9 year old girl had exertional dyspnoea and fatigue. Height and weight were in the normal range. The girl had no signs of cyanosis and there were no other abnormal findings. Physical examination showed a mild right parasternal systolic murmur over the fifth intercostal space. Spirometry, lung auscultation, and neuropsychiatric status were normal. Appetite suppressant use was ruled out. At this time, right heart catheterisation was performed. The diagnosis of PPH was suspected. The young girl was advised to avoid physical efforts and was released from school sports but continued her normal life. Further examinations confirmed the diagnosis of PPH. It is suggested that PPH with modestly limited physical activity (New York Heart Association functional class II) does not always seem to coincide with progression of the disease and, therefore, it may be feasible to withhold treatment while closely monitoring these patients.

The mean survival of patients with severe primary pulmonary hypertension (PPH) is <3 years without appropriate treatment. There are no long term reports on the spontaneous course of mild PPH over a longer period. Stable long term follow up is described of a 39 year old patient with PPH without treatment over a 30 year period. PPH had been diagnosed 30 years previously after right heart catheterisation (mean pulmonary artery pressure 35 mm Hg) and 30 years later, repeated measurements showed nearly unchanged haemodynamic parameters. Further examinations confirmed the diagnosis of PPH. It is suggested that PPH with modestly limited physical activity (New York Heart Association functional class II) does not always seem to coincide with progression of the disease and, therefore, it may be feasible to withhold treatment while closely monitoring these patients.

**Table 1** Cardiopulmonary haemodynamic data at baseline after 30 years of known primary pulmonary hypertension and at the end of inhalation of 5 μg iloprost

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Baseline</th>
<th>At the end of inhalation of 5 μg iloprost</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAPm (mm Hg)</td>
<td>32</td>
<td>20</td>
</tr>
<tr>
<td>PVR (dyn s cm⁻⁵)</td>
<td>339</td>
<td>178</td>
</tr>
<tr>
<td>CI (l/min/m²)</td>
<td>3.3</td>
<td>3.5</td>
</tr>
<tr>
<td>RAP (mm Hg)</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>PCWP (mm Hg)</td>
<td>7</td>
<td>6</td>
</tr>
<tr>
<td>SaO₂ (volume%)</td>
<td>97</td>
<td>98</td>
</tr>
<tr>
<td>SvO₂ (volume%)</td>
<td>73</td>
<td>74</td>
</tr>
</tbody>
</table>

CI, cardiac index; PAPm, mean pulmonary arterial pressure; PCWP, pulmonary capillary wedge pressure; PVR, pulmonary vascular resistance; RAP, right atrial pressure; Rm, mean arterial pressure; SaO₂, arterial oxygen saturation; SvO₂, mixed venous oxygen saturation; SVR, systemic vascular resistance.
DISCUSSION
In this case report we describe an untreated long term 30 year follow up of a patient with mild PPH without progression of the disease. In a non-randomised cohort trial the efficacy of high dose calcium channel blocker among patients with severe PPH and acute vasodilator responses was shown. Because of side effects, however, our patient could not be given the planned long term treatment with amlodipine. Randomised placebo controlled clinical trials have shown the clinical efficacy of oral beraprost sodium, the oral dual endothelin receptor antagonist bosentan, inhaled iloprost, and subcutaneous treprostinil. Intravenous prostaglandins are an alternative for the treatment of severe pulmonary hypertension. Prostaglandins or endothelin receptor antagonists are now recommended for first line treatment of patients with severe PPH if no acute vasodilator response is present. Treatment recommendations for milder forms of PPH (NYHA class I and II) are lacking.

On the basis of the case presented here, we suggest that PPH with modestly limited physical activity (NYHA class II) does not always seem to coincide with progression of the disease. Therefore, it may be feasible, with close monitoring, to withhold treatment for these patients.

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