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 and oxymetry 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

| Parameter               | Baseline | At the end of inhalation of 5 µg iloprost |
|-------------------------|----------|-------------------------------------------|
| PAPm (mm Hg)            | 32       | 20                                        |
| PVR (dyn s cm⁻¹)        | 339      | 178                                       |
| CI (l/min/m²)           | 3.3      | 3.5                                       |
| RRm (mm Hg)             | 81       | 82                                        |
| SVR (dyn s cm⁻¹)        | 1058     | 1015                                      |
| RAP (mm Hg)             | 3        | 2                                         |
| PCWP (mm Hg)            | 7        | 6                                         |
| SaO₂ (volume %)         | 97       | 98                                        |
| SvO₂ (volume %)         | 73       | 74                                        |

CI, cardiac index; PAPm, mean pulmonary arterial pressure; PCWP, pulmonary capillary wedge pressure; PVR, pulmonary vascular resistance; RAP, right atrial pressure; RRm, 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.

ACKNOWLEDGEMENTS
Dr Halank has received speaker’s honorariums from Actelion and Schering.

Authors’ affiliations
M Halank, C Marx, G Hoeffken, University Hospital Carl Gustav Carus, Internal Medicine I, Dresden, Germany

Correspondence to: Dr M Halank, University Hospital Carl Gustav Carus, Internal Medicine I, Fetscherstrasse 74, 01307 Dresden, Germany; Michael.Halank@uniklinikum-dresden.de

Accepted 17 March 2004

REFERENCES
1 D’Alonzo GE, Barst RJ, Ayres SM, et al. Survival in patients with primary pulmonary hypertension: results from a national prospective registry. Ann Intern Med 1991;115:343–9.
2 Olschewski H, Rohde B, Behr J, et al. Pharmacodynamics and pharmacokinetics of inhaled iloprost aerosolized by three different devices, in severe pulmonary hypertension. Chest 2003;124:1294–304.
3 Rich S, Kaufmann E, Levy PS. The effect of high doses of calcium-channel blockers on survival in primary pulmonary hypertension. N Engl J Med 1992;327:76–81.
4 Galle N, Humber M, Vachiery JL, et al. Effects of beraprost sodium, an oral prostacyclin analogue, in patients with pulmonary arterial hypertension: a randomised, double-blind, placebo-controlled trial. J Am Coll Cardiol 2002;39:1496–502.
5 Channick RN, Simonneau G, Sitbon O, et al. Effects of the dual endothelin receptor antagonist bosentan in patients with pulmonary hypertension: a randomised, double-blind, placebo-controlled study. Lancet 2001;358:1119–23.
6 Rubin LJ, Badesch DB, Barst R, et al. Bosentan therapy for pulmonary arterial hypertension. N Engl J Med 2002;346:696–703.
7 Olschewski H, Simonneau G, Galie N, et al. Inhaled iloprost for severe pulmonary hypertension. N Engl J Med 2002;347:322–9.
8 Simonneau G, Barst RJ, Galie N, et al. Continuous subcutaneous infusion of treprostinil, a prostacyclin analogue, in patients with pulmonary arterial hypertension. Am J Resp Crit Care Med 2002;165:800–4.
9 Barst RJ, Rubin LJ, Long WA, et al. A comparison of continuous intravenous epoprostenol (prostacyclin) with conventional therapy for primary pulmonary hypertension. N Engl J Med 1996;334:296–302.
10 Hoeper MM, Spiekerkoetter E, Westerkamp V, et al. Intravenous iloprost for treatment failure of aerosolised iloprost in pulmonary arterial hypertension. Eur Respir J 2002;20:339–43.