Reviewer’s report

Title: Epidemiology and long-term survival in patients with pulmonary arterial hypertension in the Czech Republic: a retrospective analysis of a nationwide registry

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Reviewer: Jiri Widimsky

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The paper demonstrates the results of the register of pulmonary arterial hypertension (PAH) in the Czech republic, being the first register of PAH in Eastern countries.

When comparing the results with the French register I would like also to know the following information: how many patients had in the Czech register PH induced by drugs and toxins, how many patients had portopulmonary hypertension and how many patients had PH due to HIV infection - were some such patients in the analysis ?

How many patients with PH were detected in the years 1980-1999?

Is there any information available on survival when comparing PH detected in the years 1980-1999 with those after 2007?

If this is not available could a comparison of survival be performed between patients enrolled in the years 2000-2006 and patients enrolled between 2007 and later e.g. 2011?

The group of congenital PH should be analysed separately. The reviewer assumes that these patients are mostly patients with an Eisenmenger syndrome. The diagnosis of this syndrome is usually made in the early childhood. These patients survive usually without vasodilation treatment up till 40 years.

The extremely good survival of PH-CHD in the present register are in agreement with my opinion.

At what age were the patients with congenital PH included into the register?

The authors do not comment the low incidence of a positive vasodilation test – only 3,1%?

The reviewer hopes that the above questions can be answered by the authors.