

was even more frequent than in young female patients, in contrast to the predominance of adenocarcinoma in young females in our series. Regional differences could be assumed. A genetic component in the evolution of lung cancer in young patients has already been described [1], leading to an ongoing prospective study on this subject, conducted by H.E. Wichmann and with the participation of our hospital.

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Long-term treatment of pulmonary hypertension with aerosolized iloprost

To the Editor:

MACHHERNDL *et al.* [1] report that long-term treatment of pulmonary hypertension with aerosolized iloprost was ineffective in a group of 12 patients with pulmonary arterial hypertension. These authors as well as GALIE [2] in an accompanying editorial state that the patients in this study were comparable to the patients in whom our group has recently reported favourable long-term effects of aerosolized iloprost [3]. We believe that this is not the case. The patients in our study suffered exclusively from primary pulmonary hypertension [3]. The study by MACHHERNDL *et al.* [1], in contrast, included only one patient with primary pulmonary hypertension. Five of their patients suffered from chronic thromboembolic pulmonary hypertension and six patients from Eisenmenger's syndrome. Although our experience with the therapeutic use of inhaled iloprost in patients with these forms of pulmonary hypertension is only limited, the report by MACHHERNDL *et al.* [1] supports our own impression that aerosolized iloprost is far less effective in these settings than in primary pulmonary hypertension.

We also agree with GALIE [2] that the results of controlled clinical trials should guide treatment of pulmonary arterial hypertension. However, all controlled clinical trials in pulmonary hypertension have studied the effects of new treatment modalities for a few months only. We have not learned anything from these trials about the long-term effects of the treatments that we now routinely apply to patients. Since long-term placebo controlled trials are unethical in a deadly disease, observational follow-up studies are a useful and necessary means to obtain at least some information about the long-term effects of new treatments.

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From the authors:

We value GALIE's [1] and HIGENBOTTAM's [2] editorial comments surrounding our report on the long-term treatment of pulmonary hypertension with aerosolized iloprost [3], and we appreciate M.M. Hoeper's recent comments. M.M. Hoeper points out that the apparent disagreements between his data [4] and our report may be due to the differences in the patient populations. We agree with him entirely. We have pointed out the fundamental differences in the patient populations in the discussion section of our manuscript. However, M.M. Hoeper states that six of our patients suffered from Eisenmenger's syndrome. This is not correct. As outlined in table 1 of our manuscript, two of the four patients with pulmonary hypertension related to congenital systemic pulmonary shunts had had prior surgical corrections with no residual shunts, while the other two had left-to-right shunts and were not Eisenmenger patients. Furthermore, we believe that differences in clinical status did indeed have a bearing on the study outcome rather than differences in diagnoses. While M.M. Hoeper's patients were on average 10 yrs younger than our patients, with five >50% haemodynamic responders and twelve >20% haemodynamic responders, there were only two patients in our study who demonstrated a >20% decrease in both pulmonary vascular resistance and mean pulmonary arterial pressure. Taken together, we treated sicker patients. In the pulmonary hypertension clinical outpatient unit at our institution, old inoperable patients with chronic thromboembolic pulmonary hypertension are the most frequent referral. In these patients, one would