Abstract Group: 4.3. Pulmonary Circulation and Pulmonary Vascular Disease
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Title: Prognostic risk factors of disease worsening in patients with functional class II pulmonary arterial hypertension

Body: Introduction: The open-label extension (OLE) phase of the EARLY trial provides a unique opportunity to analyse long-term data from WHO functional class II pulmonary arterial hypertension (PAH) patients. Here we investigated prognostic factors associated with PAH worsening. Methods: PAH worsening was defined as initiation of parenteral prostanoids, atrial septostomy, lung transplantation or death. Data on these 4 parameters, collected annually until study end regardless of OLE participation or treatment discontinuation, for all 185 patients originally randomised to placebo or bosentan were included in this analysis. OLE continued until at least 50% of patients had the opportunity to be treated with bosentan for at least 5 years. Cox regression analyses, univariate (significance set at P<0.1 cut-off) and multivariate (backward selection set at P<0.1 cut-off, including all variables having a P-value <0.1 in the univariate analysis), were employed to determine prognostic factors of PAH worsening. Results: In the univariate analysis, significant prognostic factors for a low risk of PAH worsening were time since PAH diagnosis >16 months, 6-minute walk distance >437.0 m and mixed venous oxygen saturation >68%. High values in NT-proBNP and PAH associated with connective tissue disease versus idiopathic, heritable or HIV-associated PAH were significant risk factors for PAH worsening. Apart from time since diagnosis, these factors were confirmed as significant in the multivariate analysis. Conclusions: This analysis provides firm evidence of risk factors significantly associated with PAH worsening in patients at a functionally early stage of the disease.