Anticoagulation and survival in pulmonary arterial hypertension: results from the Comparative, Prospective Registry of Newly Initiated Therapies for Pulmonary Hypertension (COMPERA).

For almost 30 years, anticoagulation has been recommended for patients with idiopathic pulmonary arterial hypertension (IPAH). Supporting evidence, however, is limited, and it is unclear whether this recommendation is still justified in the modern management era and whether it should be extended to patients with other forms of pulmonary arterial hypertension (PAH). We analyzed data from Comparative, Prospective Registry of Newly Initiated Therapies for Pulmonary Hypertension (COMPERA), an ongoing European pulmonary hypertension registry. Survival rates of patients with IPAH and other forms of PAH were compared by the use of anticoagulation. The sample consisted of 1283 consecutively enrolled patients with newly diagnosed PAH. Anticoagulation was used in 66% of 800 patients with IPAH and in 43% of 483 patients with other forms of PAH.
In patients with IPAH, there was a significantly better 3-year survival (P=0.006) in patients on anticoagulation compared with patients who never received anticoagulation, albeit the patients in the anticoagulation group had more severe disease at baseline. The survival difference at 3 years remained statistically significant (P=0.017) in a matched-pair analysis of n=336 IPAH patients. The beneficial effect of anticoagulation on survival of IPAH patients was confirmed by Cox multivariable regression analysis (hazard ratio, 0.79; 95% confidence interval, 0.66-0.94). In contrast, the use of anticoagulants was not associated with a survival benefit in patients with other forms of PAH. The present data suggest that the use of anticoagulation is associated with a survival benefit in patients with IPAH, supporting current treatment recommendations. The evidence remains inconclusive for other forms of PAH. http://www.clinicaltrials.gov. Unique identifier: NCT01347216.