



External validation of a refined four-stratum risk assessment score from the French pulmonary hypertension registry

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A four-stratum risk assessment method with low, intermediate-low, intermediate-high and high risk categories was better at discriminating survival in pulmonary arterial hypertension than a three-stratum method with low, intermediate and high risk groups <https://bit.ly/3mA6kj7>

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Abstract

Introduction Contemporary risk assessment tools categorise patients with pulmonary arterial hypertension (PAH) as low, intermediate or high risk. A minority of patients achieve low risk status with most remaining intermediate risk. Our aim was to validate a four-stratum risk assessment approach categorising patients as low, intermediate-low, intermediate-high or high risk, as proposed by the Comparative, Prospective Registry of Newly Initiated Therapies for Pulmonary Hypertension (COMPERA) investigators.

Methods We evaluated incident patients from the French PAH Registry and applied a four-stratum risk method at baseline and at first reassessment. We applied refined cut-points for three variables: World Health Organization functional class, 6-min walk distance and N-terminal pro-brain natriuretic peptide. We used Kaplan–Meier survival analyses and Cox proportional hazards regression to assess survival according to three-stratum and four-stratum risk approaches.

Results At baseline (n=2879), the four-stratum approach identified four distinct risk groups and performed slightly better than a three-stratum method for predicting mortality. Four-stratum model discrimination was significantly higher than the three-stratum method when applied during follow-up and refined risk categories among subgroups with idiopathic PAH, connective tissue disease-associated PAH, congenital heart disease and portopulmonary hypertension. Using the four-stratum approach, 53% of patients changed risk category from baseline compared to 39% of patients when applying the three-stratum approach. Those who achieved or maintained a low risk status had the best survival, whereas there were more nuanced differences in survival for patients who were intermediate-low and intermediate-high risk.



Conclusions The four-stratum risk assessment method refined risk prediction, especially within the intermediate risk category of patients, performed better at predicting survival and was more sensitive to change than the three-stratum approach.