Predictors of survival in patients with precapillary pulmonary hypertension

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Introduction: Although the perceived prognosis of patients with precapillary pulmonary hypertension (PH) is poor, the natural history of this condition is very heterogeneous. In this study we sought to identify predictors of poor outcomes which could help refine prognosis.

Methods: We studied consecutive patients referred to our centre from 12/2016 to 11/2018 with confirmed precapillary PH. A range of clinical, laboratory, echocardiographic and right heart catheterization (RHC) data variables were collected to assess predictors of survival. Outcome was defined as mortality from any cause.

Results: Of the 80 included patients, 51 (64%) were female and mean age was 60.5 ± 16.0 years. The majority of patients (45%) had pulmonary arterial hypertension (group 1) and 41% were chronic thromboembolic pulmonary hypertensive disease patients (group 4). During a median follow-up of 18.7 [IQR 12.3 – 26.7] months, 10 patients (12.5%) died. New York Heart Association (NYHA) functional class (HR 19.4 [95% CI 2.56 - 147.5], p = 0.004) was the strongest predictor of mortality, whereas higher haemoglobin (HR 0.70 [0.49 - 0.99], p= 0.047) and 6-minute walking distance (6MWD) expressed as percentage of predicted (HR 0.96 [0.93 - 0.99], p = 0.004) were associated with better survival overall. Echocardiographic parameters such as eccentricity index (HR 3.35 (95% CI 1.11 - 10.0), p = 0.031), short pulmonary acceleration time (HR 0.98 [95% CI 0.96 - 0.99], p = 0.008), the presence of moderate to severe tricuspid regurgitation (HR 6.46 [95% CI 1.67 - 25.0], p = 0.007) and pericardial effusion (HR 3.86 [95% CI 1.12 - 13.4], p = 0.033) were also associated with death. Traditional right ventricular function parameters such as fractional area change, tricuspid annular plane systolic excursion (TAPSE) and S velocity of the lateral annular tricuspid annulus did not predict mortality in these patients. Invasive pressures and pulmonary vascular resistance measured by RHC were also not associated with mortality. In multivariable analysis, NYHA functional class was the only independent predictor of mortality in patients with precapillary PH (HR 14.5 [95% CI 2.3 - 146.8], p = 0.006).

Conclusion: Eccentricity index, short pulmonary acceleration time, moderate to severe tricuspid regurgitation and pericardial effusion were associated with poor survival. Functional class was the strongest independent predictor of mortality in precapillary PH patients. These parameters may help stratify the risk of death in this heterogenous population.