THE PROGNOSTIC IMPACT OF ETIOLOGIC SUBGROUP AND BASELINE CLINICAL, HEMODYNAMIC AND NEUROHORMONAL CHARACTERISTICS IN PATIENTS WITH PULMONARY HYPERTENSION AND THE RISK OF PARADOXICAL DETERIORATION IN THE INTERMEDIATE RISK GROUPS: INSIGHTS FROM A SINGLE-CENTER STUDY

Poster Contributions
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Background: Functional class (FC), 6 minute walk distance (6MWD), plasma brain natriuretic peptide (BNP) levels and hemodynamic measures (HM) are used to assess disease severity, stability and prognosis in patients with pulmonary hypertension (PH). In this single-center study we assessed prognostic impact of etiology and baseline FC, 6MWD, BNP, HM in PH patients.

Method: 184 patients (F110, M74, age 49±16 yrs) with PH included in our single center Evaluation of Pulmonary Hypertension Risk factors Associated with Survival - EUPHRATES registry. Groups were; Idiopathic pulmonary arterial hypertension (IPAH) (n=44), PAH associated with congenital heart defects (CHD-APAH) (n=70), other subgroups of PAH (n=9), group 3 PH (n=17) and group 4 PH (n=44).

Results: Pulmonary arterial systolic and mean pressures were 96.3±28 and 60.2±14 mmHg and mean pulmonary and systemic vascular resistance (PVR, SVR) and PVR/SVR were 10.6±3.5 and 21±2.5 WU and 0.49±0.2 respectively. All patients were under targeted therapy. Mean follow-up was 25±20 (2-82) months. The 1, 3 and 5 year survival were 90, 83 and 60%, respectively. Baseline 6MWD and BNP distribution were divided into tertiles and cut offs were 250 and 340 m for 6MWD and 89.2 and 464 pg/dl for BNP. CHD-APAH subgroup (p<0.001) and FC-I-II (p<0.05) but not age and sex, were associated with higher 5 year survival. However, after 3rd year FC-III patients showed a trend for an increased mortality compared to FC-IV patients (p=NS). Moreover, patients in 2nd tertile of 6MWD had higher risk of worsening and 5 year mortality compared to other tertiles (p<0.01) and difference became significant after 2nd year. Although baseline BNP tertile was not associated with survival, patients in 2nd tertile showed a time dependent trend for increased mortality (p=NS). Baseline PVR>13 WU (AUC=0.69; 95% CI:0.54-0.85, p<0.05) and PVR/SVR>0.51 (AUC=0.76;95% CI:0.57-0.94, P<0.05) predicted higher 5 year mortality.

Conclusion: CHD etiology and baseline FC, 6MWD, PVR and PVR/SVR, but not age, sex and BNP seem to predict 5 year survival in patients with PH. However, FC-II and 2nd tertile of 6MWD and BNP may be associated with a paradoxically increased risk of deterioration in a time dependent fashion.