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Title: Prognostic value of pulmonary function testing in idiopathic pulmonary hypertension – The most relevant differences between survivors and non-survivors in three-years observation study

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Body: Introduction: The risk stratification in idiopathic pulmonary hypertension (IPAH) is currently based on hemodynamic and functional parameters. Clinical significance of pulmonary function testing (PFT) is not well defined.The aim of the study was to analyze the prognostic value of PFT in IPAH patients (pts) during three-years observation period. Material: 65 IPAH pts, mean age 41.5 (+/-14.5) years, mean(m)PAP - 57.3 (+/-12.5) mmHg, mRAP - 8.9 (+/- 5.2) mmHg, mCI - 2.5 (+/-0.7) L/min/m2, msatO2mv - 56.8 (+/-9.1)%. PFT was performed in all of the patients according to ERS guidelines. Survival time was assessed from PFT till the death, lung transplantation or the end of observation time. The prognostic value of selected variables was tested by Cox proportional hazards regression analysis. Results: The following differences in median PFT values between survivors (50 pts) and non-survivors (15 pts) were noted: FEV1 (%pred.) - 100.2 vs 85.8 (p=0.01), FEV1/VC – 0.77 vs 0.74 (p=0.03), VC (%pred.) 109.2 vs 103.1 (ns), MMEF (%pred.) – 75.8 vs 46.4 (p=0.02), DLCO (%pred.) – 74.2 vs 52.0 (p=0.005), TLC-VA (ml) – 660 vs 1170 (p=0.005). DLCO (%pred) (HR: 0.97, p=0.005) and TLC-VA (HR: 2.15, p=0.03) were significant prognostic variables in univariate analysis. DLCO<60%pred, pO2<60 mmHg and satO2mv< 63% were significant prognostic indicators in multivariate analysis. Conclusions: Three- years survivors comparing to non-survivors had significantly higher DLCO% pred. and less disturbances of gas distribution expressed as TLC-VA. Nevertheless only DLCO %pred. was an independent prognostic variable.