Detection Score for Pulmonary Hypertension in Systemic Sclerosis Patients: Observations from the Pharos Registry

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Introduction/Rationale: Despite efforts at early detection, systemic sclerosis pulmonary hypertension (SSc-PH) patients present with advanced disease. It is unclear whether endothelial biomarkers (e.g. soluble endoglin [sEng], pentraxin-3 [PTX-3]) can detect SSc-PH or add to a clinical prediction score. Methods: Five-hundred fifty-eight patients from the PHAROS Registry, a prospective multicenter observational study of SSc patients at high risk for or with incident PH, were included in this analysis. Patients had PH (n=302) if their mean pulmonary artery pressure was ≥25mmHg on right heart catheterization. All other patients comprised the “no PH” group (n=256). PTX-3 and sEng were measured by ELISA in patients with available blood samples (n=118). Biomarker levels were compared between patients with and without PH using t-tests. Stepwise backwards regression was conducted using univariate parameters associated with PH that had a p value of <0.1. A composite score was derived for patients without blood samples using β coefficients of the retained variables. Receiver operating characteristics (ROC) analysis was performed for detection of PH. The composite detection score was internally validated in the group of patients with available blood samples. Results: Patients with blood samples had a lower six-minute walk distance (6WMD) and were more likely to be male than those without blood samples. One hundred forty-seven patients had Group 1 pulmonary arterial hypertension, 61 Group 2 PH (left-heart associated), and 94 Group 3 PH (from lung disease). PTX-3 was lowest in the Group 2 PH patients (59.9±29.9 pg/mL) compared to the Group 1 (87.2±41.0) or Group 3 PH patients (89.6±27.0, p=0.02). No significant differences were observed for sEng (394±134 vs 392±124 pg/mL, respectively, p=0.96) or PTX-3 (80.1±53.6 vs 80.7±36.4 pg/mL, p=0.93) when comparing the no PH and PH groups. Functional class (FC), autoantibodies, echocardiogram-estimated pulmonary artery systolic pressure (echo-PASP), B-type natriuretic peptide, 6MWD, and DLCO were univariate predictors of PH. Independent predictors of PH were FC, echo-PASP, and DLCO (all p<0.0001). The composite score calculated using these 3 parameters had an area under the ROC curve (AUC) of 0.85 (95%CI 0.81-0.89). Applying the composite score to the group of patients with blood samples yielded a similar AUC (0.85, 95%CI 0.78-0.92, Figure). Conclusions: A composite score of routine variables (functional class, echo-PASP, DLCO) accurately distinguished patients with SSc-PH from SSc patients at high-risk for but without PH. PTX-3 and sEng levels were not different between these groups and did not improve the ability to detect PH in this high-risk cohort.
Figure: ROC curves for presence of PH by the composite detection score in the derivation group (AUC=0.85) and the internal validation group (AUC=0.85).

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