

Applying REVEAL Registry Risk Score Calculator in Connective Tissue Disease Associated Pulmonary Arterial Hypertension: Latvian National Retrospective Study

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Introduction. Connective tissue disease associated pulmonary arterial hypertension (CTD-APAH) classified under the World Health Organization group I pulmonary hypertension, is a severe complication and contributor to morbidity and mortality in all forms of CTD. The REVEAL Registry predictive algorithm published in 2012 showed reliable risk assessment and one year survival prediction of newly or previously diagnosed PAH, thus derivatively possess major prognostic and therapeutic implications. The score divides individuals into low (1-7), average (8), moderate (9), high (10-11), very high risk (≥ 12).

Aim, Material and Methods. The aim of the study was to explore the application of the REVEAL risk score calculator on the population of CTD-APAH in Latvia. We retrospectively analysed data obtained from the Latvian National Registry. 25 CTD-APAH patients, diagnosed between August 2008-2015. After the database was completed, we calculated their individual risk scores at the time of diagnosis. Patients with incomplete data were excluded ($n = 5$). We used Microsoft excel and SSPS 20.0 programmes.

Results. Population: 90% women ($n = 18$) and 10% men ($n = 2$). Mean age and standard deviation $\bar{x} = 58.5 \pm 14$ years. The CTD distribution: 55% Systemic Scleroderma, 25% Systemic Lupus Erythematosus, 10% Mixed connective tissue disease and 10% others. At the time of diagnosis, 85% were in New York Heart Association functional class III or IV. REVEAL risk score: low $n = 4$, average $n = 4$, moderate $n = 4$, high risk $n = 5$ and very high $n = 3$. Mean score was $\bar{x} = 9.2 \pm 2.1$; range 6-13 (95% CI from 8.2-10.2). Significant correlation of the risk score with pulmonary vascular resistance and pulmonary vascular resistance index: Pearson correlation $r = + 0.78$ and $+ 0.73$, respectively ($p < 0.01$). 1 year survival 85% ($n = 17$), SE 8%, all 1st year death cases ($n = 3$) were attributable to the very high risk group. Death incidence rate 0.11 deaths/1 person years (95% CI = 0.015 to 0.21). Incidence proportion estimates 1.66 cases/Million adult inhabitants and estimates prevalence - 12.04 cases/Million adult inhabitants.

Conclusions. In our small scale population of CTD-APAH in Latvia, the REVEAL risk score accurately predicted the highest risk, manifested as the one year survival. We conclude that the relatively simple and easy to use REVEAL risk calculator should be applied in our clinical practice for all consecutively diagnosed PAH patients, while establishing their initial management strategy.