Validation of ESC/ERS 2015 guidelines risk score in patients with scleroderma associated pulmonary arterial hypertension (SSc-PAH)

Authors:
H Fayed¹, M Ahmad¹, R Abdelkhalak¹, T Kotecha¹, J Brown¹, N Okonkwo¹, DS Knight¹, P Marino¹, B Schreiber¹, C Handler¹, CP Denton¹, JG Coghlan¹, ¹Royal Free Hospital - London - United Kingdom of Great Britain & Northern Ireland,

On behalf: Royal Free Pulmonary Hypertension Service

Topic(s):
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Background: The ESC/ERS 2015 guidelines presented a comprehensive risk assessment model with three risk categories based on different clinical and biomarkers data. Low, intermediate and high risk were defined with one-year mortality of <5%, 5-10% and >10%. Different groups suggested different methods of risk assessment based on this model.

Purpose: We applied three different methods to validate the ESC/ERS risk prediction model for one-year survival in SSc-PAH.

Methods: 309 patients with SSc-PAH have been diagnosed and managed in our institution from 2006 to 2017. We used three different risk assessment models that have been previously suggested;
1. Suggested by the Swedish group¹: Having a score of 1 (low risk), 2 (intermediate risk) or 3 (high risk) resulting from the average of the sum obtained after grading each of the variables (whichever available) from 1 to 3 according to ESC/ERS guidelines cut-offs for WHO-functional class (FC), 6-minute walking distance (6MWD), NT-Pro BNP, right atrial pressure (RAP) and cardiac index (CI).
2. Suggested by the French group²: Having none, 1, 2, 3 or 4 of the following low-risk criteria of; FC, 6MWD, RAP and CI.
3. Instead of the invasive data, The French group also suggested the use of a non-invasive model including NT-Pro BNP.

Patients were divided into different risk groups according to data obtained at baseline and at their 6-month follow-up. Survival analysis over a 5-year period was performed using Kaplan-Meier analysis.

Results: Overall median follow-up was 33.3 months. One year survival was significantly different between the risk groups (p <0.001) using either baseline or follow-up data. Applying the French group non-invasive model, almost two thirds of the population ended up in the higher risk group. Whilst applying the Swedish model, two thirds of the population ended up in the intermediate risk group. In all the models used, there were significantly less number of patients in the lower risk groups at onset with improvement of risk profile at follow up. An important advantage of the Swedish model, that it can be calculated even in the presence of missing data, a problem commonly encountered. The French models are easier to calculate but they cannot be applied when there is missing data.

Conclusion: All models used were valuable in risk prediction of SSc-PAH both at onset and at follow up. However, each model has some caveats which should be considered. In all the methods used, the prevalence of high risk criteria is higher amongst the SSc-PAH population which indicates the higher risk profile at the time of diagnosis in comparison to other PAH populations, which could explain the poorer outcome.
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