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POS0319

PERFORMANCE OF PATIENT REPORTED OUTCOMES (PROS) IN SCLERODERMA PATIENTS WITH REDUCED LUNG FUNCTION IN AN OBSERVATIONAL COHORT

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Background: Patient Reported Outcomes (PROs) are used to capture disease impact on patients' Health Related Quality of Life (HRQoL) and they have been increasingly used as endpoints in clinical trials for Systemic Sclerosis (SSc). The DeSScIPHER project within the EUSTAR group highlighted dyspnea as one of the factors more strongly related with the highest SHAQ scores (1). Nevertheless, the SENSICIS trial in SSc-ILD (2) showed the efficacy of Nintedanib in reducing the annual rate of FVC loss, as compared to placebo, without significant changes in the PROs used as secondary endpoints (St. George's Respiratory Questionnaire and the FACIT-Dyspnoea questionnaire). Since patient's perspective is a crucial determinant to define the overall relevance of an intervention, the performance of PROs in reflecting different lung functional stages is a relevant issue in SSc.

Objectives: To analyse in a prospective SSc cohort the inference of reduced lung function as measured by FVC on median PRO scores and the correlation among distinct commonly used PROs.

Methods: A cross-sectional study was performed on data exported from the STRIKE database regarding SSc patients followed in Leeds Scleroderma Programme for SSc. Data included records of periodical visits with scores of different PROs commonly used in SSc (Scleroderma-Health Assessment Questionnaire (SHAQ), Cochin Hand Function Scale (CHFS) and Borg dyspnoea scale) and FVC%-predicted (%pFVC) and DLCO%-predicted (%pDLCO). SHAQ score was calculated as the mean value of HAQ (0-3) with the average of the 7-VAS scores divided by 3.33. The 7-VAS (score 1-10) were 1: pain, 2: general function, 3: arthritis, 4: gastro-intestinal, 5: dyspnoea, 6: Raynaud Phenomena; 7: digital ulcers. The correlation of FVC with distinct PROs, and the inter-PRO correlation, were analysed through the non-parametric Spearman test.

Results: Complete data were available from 182 visits of 87 SSc patients (41 with diffuse and 46 with limited cutaneous involvement). Mean %pFVC was 95.16 ±24.93 (median 95) and mean %pDLCO was 59.31±16.51 (median 59). Overall, FVC and DLCO showed a moderate correlation with SHAQ ($r=-0.36$, $p<0.001$ and $r=-0.24$, $p<0.001$ respectively), while Borg score showed a stronger negative correlation with FVC and DLCO ($r=-0.42$ and $r=-0.38$, $p<0.001$ for both). In a sub-analysis of patients grouped by FVC, patients with FVC 50-70% showed a significant correlation of FVC with SHAQ ($r=-0.47$, $p<0.012$), which was not present in patients with FVC 70-90% ($r=0.23$, $p<0.13$). VAS-5 dyspnoea and Borg were not associated with FVC in these two subgroups of patients. Inter PROs analysis showed that CHFS score had a stronger correlation with SHAQ than Borg dyspnoea score in the overall population ($r=0.86$ vs. $r=0.57$, both $p<0.001$).

Conclusion: The analysis of a single centre prospective cohort of SSc patients, suggests a small inference of lung function in the overall SHAQ. The stronger correlation of SHAQ with CHFS, than with Borg score, suggests a higher weight of hand function on SHAQ in this population with relatively conserved lung function. In patients with %pFVC <70%, the correlation with SHAQ was stronger than in patients with %pFVC >70%, suggesting that mild reductions in FVC might not be perceived by the patients, or at least they might not modify HrQoL as measured by SHAQ.

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POS0320

POOR PROGNOSIS PREDICTION IN ANTI-MDA5 POSITIVE DERMATOMYOSITIS ASSOCIATED WITH INTERSTITIAL LUNG DISEASE: THE CROSS-CAR DECISION TREE MODEL

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Background: The prognosis of anti-melanoma differentiation-associated gene 5 positive dermatomyositis (anti-MDA5+ DM) – associated interstitial lung disease (ILD) is poor and heterogeneity.

Objectives: The aim of this study was to evaluate prognostic factors and to develop a simple and generally applicable bedside decision tree model for predicting outcomes in patients with anti-MDA5+ DM and to guide treatment.

Methods: We analyzed data for 246 anti-MDA5+ DM patients from Myositis Study Group-Jiangsu, a multicenter cohort across eighteen tertiary hospitals in Jiangsu province, from March 2019 to October 2020. The primary end point was all-cause death, and the secondary end point was occurring of rapidly progressive-ILD (rp-ILD). We used a multivariable Cox proportional hazards model to identify the independent prognostic risk factors of death and rp-ILD respectively. A decision-tree prediction model was developed by using data from 10 hospital of southern region (n=163), with validation by using contemporaneous data from northern region (n=83).

Results: To assess the risk of rp-ILD, we developed a combined risk score, the CROSS score, that included the following values and scores: C-reactive protein ($\leq 8\text{mg/L}$, 0; $>8\text{mg/L}$, 3), anti-Ro52 antibody (negative, 0; positive, 4), Sex (Female, 0; Male, 2) and Short course of disease (More than 3 months, 0; Less than 3 months, 2). The mortality risk was identified by the CAR score, including C-reactive protein ($\leq 8\text{mg/L}$, 0; $>8\text{mg/L}$, 1), Alanine Transaminase ($\leq 50\text{units/L}$, 0; $>50\text{units/L}$, 1) and rp-ILD (non-rpILD, 0; rp-ILD, 3). We divided patients into three risk groups according to the CROSS score: low, 0 to 3; medium, 4 to 7; and high 8-11. And then Use of a simple decision tree prediction model permitted stratification into three different outcome prediction groups. High-risk patients had significantly higher mortality rates than low- and medium-risk patients in both discovery and validation cohorts ($p < 0.0001$).

Conclusion: The CROSS-CAR decision tree model is easy to evaluate the poor prognostic risk in MDA5+ DM patients during any follow-up period. Unnecessary lung examination, such as chest CT scan and arterial blood gas analysis was avoided in low- and medium- rpILD risk patients. The special ambulance, with red cross sign tagged on car in China, may help to screen the high risk patients and to guide further treatment.

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POS0321

USE OF HYDROXYCHLOROQUINE AND SYSTEMIC SCLEROSIS: RESULTS FROM A PROSPECTIVE OBSERVATIONAL STUDY ON THE EUSTAR COHORT

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