PULMONARY INVOLVEMENT AND FUNCTIONAL LIMITATION IN SYSTEMIC SCLEROSIS

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Background: Pulmonary involvement is the main cause of death in Systemic Sclerosis patients (SS). However, there is little information whether its presence is capable of affecting the functional capacity of patients and if this influences the quality of life perceived by them.

Objectives: To determine whether the presence of pulmonary involvement in patients with SS (Intestinal Lung Disease and/or Pulmonary Arterial Hypertension) is related to greater functional disability.

Methods: Observational and cross-sectional study, with a prospectively performed protocol, of patients diagnosed of SS according to ACR/EULAR 2013 criteria. Demographic, clinical, analytical, activity (EUSTAR index), severity (Medsgar scale and modified Rodnan index), health perception (SF36) and disability (HAQ and Cochin test) variables were collected. Moreover, VideoCapillaroscopy (VCL) and Respiratory Function Test were made. All the patients had pulmonary TCDM and echocardiography in order to describe pulmonary features.

Results: 42 patients were included (95.4% women), with an average age of 59.2 years (SD 12.9 years). The median of years from diagnosis was 4, and 6 from the first not Raynaud symptom. Out of them, 20 were SS limited, 20 patients SS diffuse; and 2 patients SS sine sclerodermia. Pulmonary hypertension (HTP) was found in 11.9% of patients, as well as EPID in 33.3% (UIP 16.7%, NSIP 14.3%, bronchiolitis 2.3%). Analyzing the subgroup of patients with EPID, we can highlight higher score of HAQ, Cochin and activity index EUSTAR, with less influence in Rodnan index and without differences in SF36.

Regarding the subset of patients with PAH, we found higher score on HAQ, EUSTAR activity index, index Rodnan, Medsgar gravity scale and the SF36, than in non-PAH patients, with no differences in Cochin test.

Conclusion: We can highlight the presence of greater functional limitation that is especially reflected in rates of disability (HAQ and Cochin), as well as an increased activity of disease and a lower self-perception of health in patients with Systemic Sclerosis pulmonary involvement.

Disclosure of Interests: None declared


SELF-ADMINISTERED VERSUS INTERVIEW BASED INTERNATIONAL CONSENSUS CRITERIA FOR THE DIAGNOSIS OF RAYNAUD’S PHENOMENON QUESTIONNAIRE – A SINGLE CENTRE EXPERIENCE

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Background: International Consensus Criteria for diagnosing Raynaud’s Phenomenon (RP) have been proposed recently [1]. These criteria are based on the three-step conditional approach (i.e. a screening question, assessment of colour changes and an item of disease score calculation). It is unclear whether differences exist between the self-administered and interview-based way of questionnaire fulfilment.

Objectives: We aimed in our study to evaluate the two approaches in patients with suspected RP.

Methods: This cross-sectional study was conducted at our secondary/tertiary rheumatology centre between 1 October 2018 and 31 December 2018. Each patient referred for the video capillaroscopy was asked to complete the RP questionnaire on the day of the referral. The same questionnaire was applied as an interview by the rheumatologist before the video capillaroscopy. Differences in answers and level of individual agreement on individual three items, as well as the result of the questionnaire, between the two ways of questionnaire completion were assessed with Fisher exact test and Kappa coefficient, respectively.

Results: During the 3-month period we included 84 consecutive patients (88.1% female, median age (IQR) 49.5 (41.9–56.5) years). Results of answers to each item, final outcome of the questionnaire and the level of individual agreement for each item are presented in Table 1.

Table 1. Answers to Raynaud’s phenomenon International Consensus Criteria questionnaire according to mode of administration

<table>
<thead>
<tr>
<th>Item</th>
<th>Self-administered</th>
<th>Interview based</th>
<th>p-value*</th>
<th>Measure of individual agreement– Kappa coefficient (% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Item 1. Are your fingers unusually sensitive to cold?</td>
<td>79 (94.0%)</td>
<td>78 (92.9%)</td>
<td>0.514 (0.101–0.927)</td>
<td></td>
</tr>
<tr>
<td>Item 2. Occurrence of biphasic colour change during the vasospastic episodes (white and blue)</td>
<td>40 (47.6%)</td>
<td>42 (50.0%)</td>
<td>0.877 (0.623–0.900)</td>
<td></td>
</tr>
<tr>
<td>Item 3. RP disease score</td>
<td>53 (63.1%)</td>
<td>67 (79.8%)</td>
<td>0.026 (0.039–0.747)</td>
<td></td>
</tr>
<tr>
<td>Presence of RP according to RPICq</td>
<td>31 (36.9%)</td>
<td>36 (42.9%)</td>
<td>0.529 (0.640–0.915)</td>
<td></td>
</tr>
</tbody>
</table>

*RP – Raynaud’s phenomenon; RPICq – International Consensus Criteria for the Diagnosis of Raynaud’s Phenomenon questionnaire, ** Fisher-test

Conclusion: Overall, there was a substantial level of agreement between self-administered and interview-based application of International Consensus Criteria for the Diagnosis of Raynaud’s Phenomenon questionnaire. However, we observed that the patients were inclined to score the severity of their RP lower than the physicians.

REFERENCE

Disclosure of Interests: None declared


SPECIFIC ALTERATIONS OF NAILFOLD CAPILLARIES MIGHT ANTICIPATE THE APPEARANCE OF THE EARLY CAPILLAROSCOPIC SCLERODERMA-PATTERN IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Non-specific abnormalities (i.e. capillary enlargements) are usu- ally found at the nailfold videocapillaroscopy (NVC) analysis in patients affected by primary Raynaud’s phenomenon (PRP). We have previously demonstrated that capillary diameter is an independent predictor for development of systemic sclerosis (SSc) associated secondary Raynaud’s Phenomenon (SRP), so that progression to SRP is unlikely for subjects affected by RP when average nailfold capillary diameter is under 30 μm [1]. However, until now these findings have not been clearly classified in a defined pattern able to really predict the evolution to a scleroderma-pattern.

Objectives: This pilot study aimed to identify in a cohort of SSc patients a “very early” NVC pattern at high risk of evolution in the already defined NVC scleroderma-patterns.