HYDROXYCHLOROQUINE MIGHT REDUCE MORTALITY IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a devastating disease that has a profound impact on life expectancy, reflected by a standardised mortality ratio of 3.5. There is still limited data regarding the predictive factors for mortality in patients with SSc. Determining those factors could guide in disease management and follow-up.

Objectives: We aimed to identify the predictive factors for death in SSc.

Methods: Patients followed in a tertiary rheumatology clinic in the last 5 years were included in this retrospective study. All of the patients met the ACR / EULAR SSc 2013 criteria. Medical records of the patients were reviewed. Follow up time was defined as the time period from the first admission of the patient to our rheumatology clinic until the date of death or the date on which the study was performed. Candidate predictive factors for mortality were tested by Kaplan-Meier (with Log rank) and Cox-regression analyses.

Results: In total 146 patients (mean age 55.6±12.3 years, female 89.7%, diffuse cutaneous type SSc 45.2%) were included in the study (Table 1). The mean age at diagnosis of study group was 48±13.7 years. The median duration of follow-up was 71 (6-228) months. Fourteen (10%) patients died during follow-up. The causes of death were: pulmonary (7), renal (2) and cardiac diseases (1), infection (3) and cancer (1).

Univariate analysis revealed that age at diagnosis (p=0.028), SSc subtype (p=0.035), the presence of interstitial lung disease (p=0.002), oesophageal involvement (on computed tomography) (p=0.030), pulmonary artery systolic pressure of ≥35 mmHg (measured by transthoracic echocardiography) (p=0.004), glucocorticoid (p=0.029), hydroxychloroquine (p=0.002) and cyclophosphamide (p=0.006) usage at any time were associated with mortality (Figure 1). Multivariate analyses model formed with age at diagnosis (B: 0.055, 95% CI, 0.005-0.105; p=0.033), SSc subtype (B: 0.963, 95% CI 0.541-12.684; p=0.231), glucocorticoid (B: 1.396, 95% CI, 0.487-3.357; p=0.198) and hydroxychloroquine usage (B: -1.50, 95% CI, 0.061-0.816; p=0.023) showed that age at diagnosis and hydroxychloroquine usage were independent predictive factors for mortality in patients with SSc.

Conclusion: The results of the study revealed for the first time that apart from the age at diagnosis hydroxychloroquine might reduce mortality in patients with SSc. Further studies are needed to prove of this information.

REFERENCES:

Table 1. The demographic and clinical features in patients with systemic sclerosis.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Baseline</th>
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</thead>
<tbody>
<tr>
<td>Age at diagnosis</td>
<td>48±13.7</td>
</tr>
<tr>
<td>Female sex, n (%)</td>
<td>131 (89.7)</td>
</tr>
<tr>
<td>Duration of follow-up, months*</td>
<td>71 (6-228)</td>
</tr>
<tr>
<td>Disease subtype, n (%)</td>
<td></td>
</tr>
<tr>
<td>Diffuse / Limited</td>
<td>66 (45.2) / 80 (54.8)</td>
</tr>
<tr>
<td>Autoantibodies, n (%)</td>
<td></td>
</tr>
<tr>
<td>Anti-Scl70 antibody</td>
<td>50/143 (35.0)</td>
</tr>
<tr>
<td>Anti-Centromere antibody</td>
<td>62/143 (43.4)</td>
</tr>
<tr>
<td>Immunosuppressive medication, ever, n (%)</td>
<td></td>
</tr>
<tr>
<td>Hydroxychloroquine</td>
<td>91/143 (63.6)</td>
</tr>
<tr>
<td>Mycophenolate mofetil</td>
<td>18/145 (12.4)</td>
</tr>
<tr>
<td>Azathioprine</td>
<td>47/145 (32.4)</td>
</tr>
<tr>
<td>Cyclophosphamide</td>
<td>24/145 (16.6)</td>
</tr>
<tr>
<td>Glucocorticoid</td>
<td>80/140 (57.1)</td>
</tr>
<tr>
<td>Others, n (%)</td>
<td></td>
</tr>
<tr>
<td>ILD</td>
<td>68/130 (52.3)</td>
</tr>
<tr>
<td>Percutaneous effusion, ever</td>
<td>26/133 (19.5)</td>
</tr>
<tr>
<td>Eosophageal dilation (detected by CT)</td>
<td>51/128 (39.8)</td>
</tr>
<tr>
<td>sPAP ≥35mmHg, ever (measured by ECHO)</td>
<td>46/142 (32.4)</td>
</tr>
</tbody>
</table>

Parameter presented as median (min-max); ILD: interstitial lung disease; sPAP: systolic pulmonary artery pressure
Raynaud’s phenomenon, current use of losartan and correlated with BAI scores. Also, a longitudinal study showed a reduction in volume of the hippocampus subfields volumes when compared to patient’s baseline associated with calcinosis and current use of prednisone.

REFERENCES:

Disclosure of Interests: None declared.
DOI: 10.1136/annrheumdis-2021-eular.3815

AB0458 SURVIVAL OF SCLERODERMA PATIENTS WITH INTERSTITIAL LUNG DISEASE: OBSERVATION DATA FROM A MALAYSIAN TERTIARY CENTRE
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Background: Intestinal lung disease (ILD) is the leading cause of death in scleroderma (SSc) with decline in FVC as a predictor of mortality in patients with SSc-ILD, especially in the early course of the disease.

Objectives: The aim of this study is to determine the survival rate of SSc-ILD in a Malaysian cohort of patients from University Malaya Medical Centre (UMMC).

Methods: 61 patients clinically diagnosed with SSc-ILD were identified and prospectively recruited. Baseline demographic data were collected. Kaplan-Meier analysis was used to estimate the survival.

Results: Females were predominant (56, 91.8%). 39 (64%) had limited cutaneous SSc. Majority were ethnicity Chinese 30 (49.2%), followed by Malays 20 (32.8%). Indians 7 (11.4%) and others 4 (6.6%). Mean age was 56.25 (SD ± 12.5) years while mean duration of disease (non-Raynaud’s disease onset) was 10.5 years (SD ± 9.2) (range of 1 year to 44 years). 29 (47.5%) patients were positive for anti-Scl-70, whereas 6 (9.8%) patients were anti-centromere positive. There were 16 (26.2%) deaths. Median survival was 24 years. Patients had a sharper drop in survival probability for the first 10 years compared to the next 20 years (Figure 1). Median survival in limited subset was 24 years whereas in diffuse subset was 11 years. Patients from the limited subset appeared to have higher chance of surviving for 10 years and above, compared to those in the diffuse subset (Figure 2).

Conclusion: The results demonstrate the poor survival in SSc-ILD patients. The survival rate tends to be worse in the first 10 years of SSc disease duration. Survival rate was poorer in patients with diffuse cutaneous subset.

REFERENCES:

Disclosure of Interests: JASMIN RAJA Speakers bureau: For Boehringer Ingelheim for topic on Scleroderma-ILD, Grant/research support from: From Boehringer Ingelheim for scleroderma research, Shantini Muthusamy: None declared., CHOUNG MIN NG: None declared.
DOI: 10.1136/annrheumdis-2021-eular.3815

AB0459 ARTICULAR INVOLVEMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS
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Background: SSc (systemic sclerosis) is a connective tissue disease characterized by small vessel vasculopathy, production of autoantibodies, and fibroblast dysfunction leading to increased deposition of extracellular matrix in the skin and internal organs mainly. Therewithal, many SSc patients develop musculoskeletal symptoms during the course of their illness. Different rheumatic complaints such as arthralgia, arthritis, contractures, tendon friction rubs, calcinosis, and acroosteolysis can be seen as musculoskeletal symptoms in SSc patients.

Objectives: To provide an overview of the spectrum of articular involvement in SSc and determine the relationship between these involvements and Rheumatoid factor (RF) and Anti-cyclic citrullinated peptide (Anti-CCP) positivity and organ involvements.

Methods: We performed a retrospective cohort study involving 232 SSc patients who were followed up in our department of rheumatology between 2000 and 2020 years. The patients were divided into two groups as limited and diffuse SSc. Age, gender, weight, height, smoking habits, duration of illness, follow-up duration, other systemic organ involvement, and radiographic findings were recorded. Diagnostic tests such as RF, Anti-CCP, ANA, ENA panel tests, direct radiographs were examined.

Results: The mean age of the patients was 59.4 ± 12.8 and 88.4% of them were women. 69.3% of patients were limited SSc. At any stage of the disease, 39.1% of all patients had arthralgia and 34.1% had arthritis. The arthritis rates were similar between the SSc groups (p = 0.396). RF and anti-CCP positivity rates were similar between the SSc groups. Although RF and anti-CCP positivity rates were higher in the patient group with arthritis, it was not statistically significant (respectively p<0.063, p<0.070). Interestingly, the lung involvement rate was higher in patients with arthritis (63.3% vs 46.4%) (p<0.015). Other clinical, demographic characteristics, laboratory, and radiographic findings of the patients are shown in Table 1.

Conclusion: Articular involvement in SSc is a common clinical feature seen in one-third of patients regardless of the type of disease. Although RF and Anti-CCP positivity are more common in patients with arthritis, it was not statistically significant. Interestingly, arthritis is a more common manifestation in patients with lung involvement.

REFERENCES:

Table 1. Demographic and clinical characteristics of b/tsDMARDs patients

Table-1

<table>
<thead>
<tr>
<th>Limited SSc</th>
<th>Diffuse SSc</th>
<th>All patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>(n=161)</td>
<td>(n=71)</td>
<td>(n=232)</td>
</tr>
</tbody>
</table>

Age (years) (Means±SD) 60.5 ± 12.7 58.6 ± 12.9 59.9 ± 12.8 0.295
Disease duration (years) (Means±SD) 15.5 ± 6.6 12.4 ± 8.1 11.8 ± 7.1 0.384
Weight (kg) (Means±SD) 68.2 ± 13.3 66.2 ± 15.6 67.0 ± 14.1 0.331
BMI (kg/m²) (Means±SD) 27.6 ± 5.5 26.2 ± 6.1 27.1 ± 5.7 0.102
Female, n (%) 146 (90.7) 59 (83.1) 205 (88.4) 0.097
Current and ex smoker, n (%) 60 (37.2) 18 (25.4) 78 (33.6) 0.077
Digital ulcer, n (%) 53 (32.9) 44 (37.9) 87 (37.5) 0.03*
Contracture on hand 28 (17.4) 28 (39.4) 56 (24.1) <0.001*
Arthralgia, n (%) 63 (39.1) 29 (31.5) 92 (39.7) 0.806
Arthritis, n (%) 52 (32.3) 27 (38) 79 (34.1) 0.396
Joint space narrowing on X-ray, n (%) 44 (51.2) 60 (38.3) 74 (55.6) 0.160
Joint erosion on X-ray, n (%) 17 (19.8) 14 (29.8) 31 (23.3) 0.205
Acroosteolysis on X-ray, n (%) 9 (10.5) 16 (34) 25 (18.8) 0.001
ANA positivity, n (%) 155 (96.3) 69 (97.2) 224 (96.6) 0.738
Anti-Scl positivity, n (%) 41 (28.7) 48 (71.6) 89 (42.4) <0.001*
Anti-centromere positivity, n (%) 65 (45.5) 10 (14.9) 75 (33.7) <0.001*
RF positivity, n (%) 27 (17.5) 14 (20.6) 41 (18.5) 0.589
Anti-CCP positivity, n (%) 16 (12.7) 7 (11.3) 23 (12.2) 0.782

P*Independent Samples T Test, Pearson Chi-Square Test, BMI: Body mass index. ANA: Anti nuclear antibody. RF: Rheumatoid factor, Anti-CCP: Anti- Cyclic citrullinated peptide

Disclosure of Interests: None declared.
DOI: 10.1136/annrheumdis-2021-eular.4103

AB0460 THE UTILITY OF USB CAPILLAROSCOPE FOR ASSESSMENT OF RAYNAUD’S PHENOMENON PATIENTS
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Background: Videocapillaroscopy is the gold standard for evaluation of nailfold capillaries and the major tool used for differentiation of primary and secondary Raynaud’s phenomenon (RP) in rheumatology practice. However, nowadays, there are also accessible alternatives such as USB capillaroscopes, which offer the opportunity to apply capillaroscopic examination at a significantly lower price.

Objectives: The aim of the current study was to study the utility of USB capillaroscope (Dinolite) via assessment of capillaroscopic images obtained by patients with primary and secondary RP in rheumatic diseases.

Disclosure of Interests: None declared.
DOI: 10.1136/annrheumdis-2021-eular.17

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