Conclusion: In an unselected cohort of SSc patients, a third of the patients has ILD at baseline which is in line with previous prevalence reports. Importantly, this is the first study reporting incidence rates of SSc-ILD.

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THU0368

RAPID SKIN THICKNESS PROGRESSION RATE IS ASSOCIATED WITH HIGH INCIDENCE RATE OF CARDIOPULMONARY INVOLVEMENT AND MORTALITY IN PATIENTS WITH EARLY DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS (DCSSC)

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Background: There has been no prior inception cohort study regarding the incidence rate of cardiopulmonary involvement in patients with early dcSSc comparing among different groups of skin thickness progression rate.

Objectives: The aims of this study were to compare differences in early dcSSc patients, which were classified into 3 subgroups; (a) rapid skin progression [RPsp], (b) intermediate skin progression [IMsp], and (c) slow skin progression [SLsp]; regarding: (1) clinical characteristics, (2) incidence rate of left ventricular ejection fraction (LVEF) < 50 %, interstitial lung disease (ILD), and pulmonary hypertension (PH). In addition, to compare mortality rate between skin improved and non-improved skin group.

Methods: We used an inception cohort of early dcSSc patients seen at the Rheumatology clinic, Maharaj Nakorn Chiang Mai Hospital, between January 2010 and December 2017. All patients were assessed for demographic data, clinical manifestations, modified Rodnan Skin Score (mRSS) and underwent echocardiography, and HRCT at the study entry and then annually.

Results: One hundred and two dcSSc patients (55 females and 89 anti-ScI 70 antibody positivity) with a mean±SD age of 53.2 ± 8.8 years and mean disease duration of 11.2 ± 8.7 months were enrolled, during a mean observation period of 54.0 ± 24.0 months. Mean±SD of baseline mRSS was 21.9 ± 9.1. Patients were classified into 3 groups: (a) 41 (40.2 %) patients with RPsp; (b) 37 (36.3 %) IMsp and; (c) 24 (23.5 %) SLsp. At enrollment, the RPsp group had significantly differences in clinical manifestations compared to IMsp and SLsp group as the followings: mean disease duration (5.4 ± 2.3 vs. 11.5 ± 6.1 and 20.7 ± 10.3 months, p < 0.001), anti-ScI 70 antibody- positive (95.1 % vs. 75.7 % and 91.7 %, p = 0.035), mRSS (26.4 ± 7.9 vs. 22.1 ± 8.4 and 13.8 ± 6.5, p < 0.001), dry mouth (48.8 % vs. 37.8 % and 8.3 %, p = 0.005), LVEF < 50 % (26.8 % vs. 21.6 % and 0, p = 0.010), pericardial effusion (0 vs. 10.8 % and 0, p = 0.018), arrhythmia (7.3 % vs. 29.7 % and 8.3 %, p = 0.018), muscle weakness (26.8 % vs. 27.0 % and 0, p = 0.009), creatine kinase ≥ 500 U/L (34.1 % vs. 18.9 % and 4.2 %, p = 0.016, and NT-proBNP (935.7 ± 2300.3 vs. 746.4 ± 1385.2 and 164.0 ± 225.3 ng/L, p = 0.001). During the observation period, the RPsp group had a significantly higher incidence rate of LVEF < 50 % compared with the SLsp group (6.06 vs. 0 per 100 person-years, incidence rate ratio= 21.27, p= 0.001). The RPsp group had a significantly higher incidence of ILD compared with the SLsp group (69.69 vs. 34.66 per 100 person-years, incidence rate ratio= 2.01, p= 0.012). Skin non-improved

group had higher mortality rate compared with the skin improved group (28.6 % vs. 5.9 %, p= 0.004).

Conclusion: Our study cohort found that early dcSSc patients with rapid skin progression had higher incidence rate of LVEF < 50 % and ILD compared to those with slow skin progression. In addition, skin non-improved patients had significantly higher mortality rates than those with skin improved.

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THU0369

EVALUATION OF DIFFERENT CLASSIFICATION CRITERIA IN SYSTEMIC SCLEROSIS IN A TURKISH COHORT: THE IMPORTANCE OF NON-SKIN MANIFESTATIONS, SEROLOGY AND CAPILLAROSCOPY

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Background: Proximal scleroderma is the major criterion in both 1980 and 2013 classification criteria for systemic sclerosis (SSc). ACR(1980) criteria included digital lesions and bibasilar fibrosis, nonetheless ACR/EULAR(2013) criteria based on a scoring system including digital lesions, telangiectasia, abnormal nailfold video-capillaroscopy(NVC), PAH, Raynaud's and specific autoantibodies.

Objectives: We aimed to implement both criteria in a Turkish SSc cohort to evaluate the contribution of non-skin manifestations, NVC and autoantibodies.

Methods: A consecutive hundred and thirty-nine (125 females) SSc patients diagnosed and evaluated by the same experts (YY, MI) with relevant NVC records and at least 6 months follow-up were included into the study. Classification criteria were used retrospectively using a preformed database.

Results: Characteristics of the SSc patients were summarized in table-1. The mean age, duration of Raynaud's and non-Raynaud symptoms were 47.1±11.9, 8.9±7.9 and 5.7±5.8 years, respectively. Diffuse and limited cutaneous disease were diagnosed in 62(44.6%) and 60(43.2%) patients respectively. Asclerodermic disease was present in 17(12.2%) patients. ANA, anti-centromere and anti-ScI70(+) positivity was 80.5%, 18.0% and 37.4%, respectively.

Twelve patients (8.6%) could not be classified as SSc by both criteria; 5 with Raynaud's+specific antibodies (2 anti-centromere+, 2 anti-ScI70+), 4 with Raynaud's+puffy hands+NC abnormalities, 2 with Raynaud's+telangiectasia and a patient with Raynaud's+sclerodactyly. Nineteen (13.7%) patients could not be classified as SSc according to ACR (1980) can be classified according to ACR/ EULAR (2013) (table-1 and -2).

The sensitivity for ACR/EULAR (2013) and ACR (1980) criteria were found to be 91.4% vs 75.5%; 98.4% vs 96.8% in diffuse cutaneous SSc, 98.3% vs 68.3% in limited cutaneous SSc and 47.1 vs 23.5% in asclerodermic SSc, respectively.

Table 1. The sensitivity for ACR (1980) and ACR/EULAR (2013) classification criteria in SSc

	ACR/EULAR (2013) +	ACR/EULAR (2013)-	total
ACR (1980)+	108	0	108
ACR (1980)-	19	12	31
total	127	12	139

Table 2. SSc patients fulfilling ACR (1980) and/or ACR/EULAR (2013) criteria

ACR/EULAR (2013) ACR (1980)	Classified n=127	Not classified n=12
+Prox scleroderma +	87	
+Puffy hands	29	4
+Sclerodactyly +	112	1
+Digital ulcers + +Pitting scars	62 72	
+Telangiectasia	86	2
+Nailfold capillaroscopy	97	4
Normal	9	
Early	17	3
Active	40	
Late	50	1
+PAH	12	
+Interstitial lung disease +	60	
+Raynaud's	120	12
+SSc- specific antibodies	79	5

Conclusion: The sensitivity of ACR/EULAR (2013) criteria was shown to be higher than ACR (1980) criteria in our Turkish SSc cohort with established cases.