Capillary dimension has been evaluated in 2 studies, with no unequivocal results [3,4]. Corrado et al. found abnormal morphology to be more commonly associated with SSc-PAH [4]. No association was found between presence of haemorrhages and SSc-PAH [4]. Ricciari et al. found that NVC score and avascular area grade were more commonly associated with SSc-PAH [5]. Concerning qualitative assessment, severe NVC patterns have been unequivocally found to be more commonly associated with SSc-PAH [4,5].

Conclusion: This is the first systematic literature review to investigate the role of NVC in SSc-PAH using standardized definitions as proposed by the EULAR SG MC/RD. Unequivocal associations with incident SSc-PAH were found in longitudinal studies between capillary density, abnormal morphology and NVC pattern. Unequivocal associations with SSc-PAH were found in cross-sectional studies between capillary density and severe NVC pattern.

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FRI0346

PERFORMANCE OF THE 2017 EUROPEAN LEAGUE AGAINST RHEUMATISM/AMERICAN COLLEGE OF RHEUMATOLOGY (EULAR/ACR) CLASSIFICATION CRITERIA FOR ADULT IDIOPATHIC INFLAMMATORY MYOPATHIES IN A HONG KONG COHORT

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Background: The new classification criteria for idiopathic inflammatory myopathy (IIM) endorsed by the European League Against Rheumatism and American College of Rheumatology (EULAR/ACR) were published in 2017 [1]. However, the majority of the patients in the development cohort were Caucasians. External validation in other populations were advised. With the increasing availability and experience of myositis specific autoantibodies (MSAs) testing, the inclusion only anti-Jo-1 antibody is apparently insufficient.

Objectives: The objective of the study was to evaluate the performance of the 2017 EULAR/ACR classification criteria in a cohort of Hong Kong adult IIM patients. The secondary objectives included examining the level of agreement between the new criteria and the traditional criteria and assessing the effect of including other MSAs into the criteria.

Methods: This was a multi-centre retrospective cross-sectional study. Consecutive patients with a clinical diagnosis of IIMs and MSA tested seen in the rheumatology clinic or admitted to the rheumatology wards of the participating hospitals in Hong Kong up till August 2018 were recruited. Patients with juvenile onset myositis were excluded. Clinical parameters required by the two criteria will be collected by reviewing the medical records. A commercial line blot immunoassay kit (EUROIMMUN) was used to detect the MSAs.

Results: Two hundred and four patients with IIM were recruited. The mean age was 59.3 years. There was a female predominance of 76.5%. The subgroups of the patients were: polymyositis 40.7%, dermatomyositis 38.2%, clinically amyopathic dermatomyositis patients 21.1%. MSAs were detected in 59.3% of the patients with anti-Jo-1 antibody being the commonest (13.2%). The new 2017 EULAR/ACR Criteria could classify 96.1% of the patients as having definite or probable IIM. The Bohan and Peter criteria could only classify 76.0% of the patients. When combining with the Sontheimer's criteria for CADM, 93.1% of the patients could be classified. The percentage agreement of the new and the Bohan and Peter criteria also increased from 77.0% to 93.1% when the latter was supplemented by the Sontheimer's criteria. If the presence of any MSAs is considered one of the criteria, the performance of the 2017 criteria improved to 97.5% while the combined Bohan and Peter/Sontheimer's criteria to 96.1%. However, the new criteria still failed to highlight the important subtype of IIM associated with anti-MDA5 autoantibody.

Conclusion: In a population with a significant proportion of CADM patients, the new 2017 EULAR/ACR classification criteria outperformed the old criteria. Finally, a clinico-serological criteria for "anti-MDA5 syndrome" is proposed in which a patient must have positive serologic testing for an anti-MDA5 autoantibody, plus one of the following conditions:

myositis by the new EULAR/ACR criteria, interstitial lung disease or typical rash (skin ulceration, palmer papules).

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FRI0347

TUBERCULIN SKIN TEST FOR DETECTION OF TUBERCULOSIS IN SYSTEMIC SCLEROSIS

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Background: The tuberculin skin test (TST) is a screening tool for detection of occult or remote tuberculosis infection. Although skin tightness is a classic manifestation in systemic sclerosis (SSc), there are no reports vis-à-vis any limitation of the TST in SSc patients nor a definition on the cut-off for a positive TST test size for a diagnosis of tuberculosis.

Objectives: Our aims were to determine (a) the indurated reaction size of the TST; (b) the cut-off size for the indurated TST; and, (c) the sensitivity and specificity of the test for the diagnosis of tuberculosis in SSc patients.

Methods: A cross-sectional study was conducted among Thai adult SSc patients, followed up at the Scleroderma Clinic, Khon Kaen University, Thailand between November 1, 2016 and November 30, 2017. The TST was performed using 0.1 ml purified protein derivatives (PPD) injected intradermally (Figure 1), and interpreted 72 hours after testing.

Results: A total of 168 SSc patients were enrolled (female to male ratio = 1.8:1). The median age and duration of disease was 57.2 and 6.4 years, respectively. The majority (71.8%) was the diffuse cutaneous SSc subset. Seventeen cases (10.1%) were defined as tuberculosis infection. All of the patients had a history of BCG vaccination at birth. An indurated skin reaction size TST of 20 U/L had a high specificity for tuberculosis (99.3%: 95%CI 96.4-100) (Kappa 0.86; p=0.03). The modified Rodnan skin score (mRSS) had a significant negative correlation with the indurated skin reaction size (Rho -0.23; p=0.003). While other clinical parameters—such as BMI, SSc subset, serum albumin level, steroid or immunosuppressant use—were not correlated with the TST result (p=0.06, 0.14, 0.09, 0.23 and 0.89, respectively).

Conclusion: Indurated skin ≥ 20 mm indicated a high specificity for tuberculosis infection in SSc patients with history of BCG vaccination. The high mRSS resulted in a smaller skin reaction size when using the TST. The TST is thus less useful as a diagnostic tool for tuberculosis among SSc patients, especially among those with severe skin tightness.

Figure 1. Tuberculin skin test in a systemic sclerosis patient

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FRI0348

USE OF INTRAVENOUS IMMUNOGLOBULIN THERAPY IN PATIENTS WITH SYSTEMIC SCLEROSIS: A SPANISH MULTICENTER EXPERIENCE

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Objectives: To describe the efficacy of intravenous immunoglobulin (IVIG) therapy in different organic conditions of Systemic Sclerosis (SSc).

Methods: Retrospective multicenter observational study that enrolled patients with SSc treated with IVIG. We collected epidemiological data, SSc complications, treatments and functional tests. Regarding IVIG treatment we recorded the reason to use, number of cycles, and the clinical efficacy at the end of the last IVIG cycle were collected at investigator's discretion. For the comparison of variables, the Chi-square, Fisher's F test and the T test was used.

Results: 41 patients (83% women) were recruited, with a mean age of 58 ± 18.2 years old. The age of diagnosis was 48 ± 8 years old. The diffuse cutaneous SSc was the most frequent in the sample (61%) and 24/41 had Overlap syndrome (49% myositis). Regarding the different SSc complications in treated patients the most frequent were digestive involvement, arthritis and interstitial lung disease (78%, 73%, and 63% respectively). 12% of patients developed cancer and 25% died during the follow-up. The most frequent indication IVIG was myositis (51%) followed by cutaneous (17%). The mean of cycles was 10. 37% of patients had previously undergone treatment with biological therapy, being RTX the most used therapy. 80% of patients had a history of use with corticosteroid therapy. When evaluating the degree of skin involvement (mRSS) patients showed a significant improvement of -2.89 ± 5.2 (p < 0.03) at the end of the follow-up. Indeed we observed better results in the groups of patients with myositis. No differences were observed in the % FVC or % DLCO outcomes during the follow-up. However, when we compared patients with or without overlap syndrome we found differences in %FVC values at the beginning of the study that were not present at the end of the follow-up. Patients with antiScl70 seemed to have less response to IVIG therapy.

Conclusion: Our results suggest that IVIG can be useful for the management of some conditions in specific profiles of patients with SSc.

Prospective studies and randomized clinical trials are necessary to establish the efficacy and safety of IVIG to treat the different conditions of SSc.

	IVIG N=41(%)	Diffuse n=25 (%)	Limited n=16(%)
Women	34(83)	21(84)	13(81)
Age	58 ± 18		
Overlap	24(59)	11(44)	13(81)
Clinical manifestations			
Joint	30(73)	22(88)	8(50)
Muscular	30(73)	16(64)	14(88)
Reflux GE	32(78)	20(95)	12(75)
Interstitial lung disease	26(63)	16(64)	10(63)
Pulmonary hypertension	5(12)	2(8)	3(19)

	myositis	Overlap	AntiScl70	PM/ Scl
Initial mRss	16±12	13±11	29±9	15 ±10
Final mRss	12±12	7±7	32±7	13 ±15
Initial FVC	68±25	64±24	56±23	60 ±13
Final FVC	73±26	72±25	58±20	67 ±18
Initial DLCO	56±16	56±17	55±16	49 ±13
Final DLCO	68±25	71±24	45±12	69 ±18

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FRI0349

RELATIONSHIPS BETWEEN BODY COMPOSITION AND NAILFOLD VIDEOCAPILLAROSCOPY PATTERNS IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Systemic sclerosis (SSc) is a chronic autoimmune disease, characterized by microvascular damage and progressive fibrosis. Among clinical complications, abnormal body composition and sarcopenia have been reported in SSc patients(1,2). Nailfold videocapillaroscopy (NVC) is a safe diagnostic tool to assess microvascular progressive damage and may predict severe organ involvement in SSc(3).

Objectives: The aim of this study was to evaluate body composition in SSc patients and assess possible differences between patients with specific NVC patterns of microangiopathy.

Methods: 38 patients (6 men and 32 women) fulfilling ACR 2013 criteria for SSc underwent NVC to assess their NVC patterns ("Early", "Active" and "Late" pattern)(4) and DXA to evaluate body composition(5). In particular we analyzed: total mass, total lean mass, total fat mass, bone