decisions of the treating physician. Different therapeutic strategies are adopted regarding the treatment, and in many cases highly expensive drugs as Ivlg are prescribed. Further studies and recommendations are certainly needed for homogenising the patient management for this severe aspect of the disease.

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AB0807 PLATELET INDICES AS MARKERS OF INFLAMMATION IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a connective tissue disease involving multiple organs with an unknown etiology. Platelet function may be associated with endothelial dysfunction and immune regulatory mechanisms. Recently, an increased tendency to platelet aggregation and enhanced platelet activation have been described in SSc patients, suggesting a role for platelets in the disease itself.^{1,2}

Objectives: To evaluate platelet indices in systemic sclerosis (SSc) patients and identify their clinical significance as novel inflammatory biomarkers in correlation to markers of endothelial dysfunction: vascular endothelial growth factor (VEGF) and flow mediated dilatation (FMD).

Methods: Thirty-five SSc patients were enrolled in addition to thirty-five age and sex matched healthy volunteers as controls. All patients and controls underwent full medical history taking, thorough clinical examination, assessment of severity extent of skin sclerosis using the modified Rodnan skin score, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), complete blood count with special consideration to mean platelet volume (MPV), platelet distribution width and platelets count, assay for serum VEGF concentration, and brachial FMD assessment by colour duplex sonography.

Results: There was a highly significant decrease in the mean MPV in SSc patients compared to the controls (8.65±0.6 fl vs. 9.55±0.52 fl). There was a significant increase in the mean platelet count in SSc patients compared to controls (331.63±64.66 × 10³/ml vs. 297.80±44.48 × 10³/ml). In SSc patients, a significant negative correlation was found between the mean MPV and each of ESR, CRP and VEGF (r=-0.42, -0.368 and -0.55 respectively, p<0.05); and a significant positive correlation was found between the mean MPV and mean FMD (r=0.378, p<0.05). Linear regression test, showed an association between mean MPV and each of ESR and CRP (t=-3.312, -2.92 respectively, p<0.05).

Conclusions: MPV levels could be an easily measurable parameter to reflect the inflammatory condition and disease activity in systemic sclerosis patients.

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AB0808 MULTICENTRIC STUDY OF SYSTEMIC SCLERODERMA IN TUNISIA

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Background: Scleroderma is an autoimmune connective tissue disorder which is characterised by fibrosis of visceral organs, skin and blood vessels. This condition can be localised or systemic. Its estimated prevalence is 250 cases in a million and it is more common in women than in men.

Objectives: The aim of this retrospective multicenter study was to analyse demographic, clinical, laboratory features and outcome of SSc in Tunisia throughout 8 Departments of Internal Medicine and to compare them with those of other geographic groups.

Methods: One hundred and eight cases of SSc were recorded (American College of Rheumatology criteria) during a 15 years period. They were 93 women and 15 men with an average age at SSc onset of approximately 46.9 years.¹⁷⁻⁷⁵

Results: Only 18 patients had limited cutaneous SSc, 11 patients had a CREST syndrome. Our Tunisian patients were characterised by a high frequency of cutaneous signs: sclerodactyly (80.6%), proximal sclerosis (81%), telangiectasia (34.3%), Raynaud's syndrome (91%), pigmentation disorders (36%), ulceration (24%) and subcutaneous calcification (4.6%). The other clinical manifestations were dysphagia in 54% (n=59), pulmonary involvement in 55.6% (n=60), cardiac

manifestation in 27% (n=29), arthritis in 19.4% (n=21), renal involvement in 13.8% (n=15), and neurological involvement in 12 cas. 78.7% were antinuclear antibody (ANA) positive, 25% were Antitopoisomerase-I antibodies (anti-Scl-70 antibody) positive and 10% with anti-centromere antibodies.

SSc was associated to Sjögren's syndrome n=27, systemic lupus erythematosus (n=8), Polymyositis (n=5) and Rheumatoid arthritis (n=1). Treatment regimen included, calcium channel blockers (65.7%), steroids (47%), Colchicine (47.2%), D penicillamine (21%), immunosuppressive therapy was added in patient with partial control (n=13). Median follow-up period was 3.7 years. With the above treatment protocol, (29.6%) patients achieved disease control on treatment, (43.3%) had partial control while (26.8%) showed no response or progressive disease. Six patients died, three of them with scleroderma renal crisis.

Conclusions: The findings of this study documents The high frequency of extensive cutaneous sclerosis, Potential limitations and biases in our study need discussion. Specific recruitment of patients in tertiary referral centres may be the source of selection bias. Patients were evaluated by different doctors. The therapeutic management and outcome monitoring were heterogeneous. This study remains the most representative of Tunisian Scleroderma patients recruited from all parts of Tunisia

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AB0809 A NEW SCORE TO PREDICT DIGITAL ULCERS COMBINING CLINICAL DATA, IMAGING AND PATIENT HISTORY IN SYSTEMIC SCLEROSIS

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Background: Ischaemic complications such as digital ulcers (DU) are a common complication in systemic sclerosis (SSc) patients.

Objectives: The aim of this study was to combine clinical characteristics and imaging methods to a composite predictive score.

Methods: Seventy-nine SSc patients received clinical examination and their patient history was taken. Furthermore, we performed nailfold capillaroscopy (NC), colour Doppler ultrasonography (CDUS) and fluorescence optical imaging (FOI) of the hands at baseline. Newly developed digital ulcers over a period of approximately 12 months were registered. We used criteria with significant (p<0.5) OR values above 3.5 in regard to the development of these new DU to create the score (CIP-DUS, clinical features, imaging, patient history – digital ulcer score)

Results: Twenty-nine percent of the patients developed new DU during follow-up (48.1% diffuse SSc, 18.4% limited SSc). The following criteria were used: SSc diffuse subtype (OR 4.127, p=0.0087), modified Rodnan skin score >8 (OR 9.429 [95%CI: 3.0–29.2], p<0.0001), pulmonary arterial hypertension (OR 6.854 [95%CI: 1.6–9.7], p=0.0088), present digital ulcers or pitting scars at baseline (OR 15.71 [95%CI: 3.3–74.3], p<0.0001), history of digital ulcer or pitting scars (OR 36.15 [95%CI: 2.1–626.9], p<0.0001), NC pattern (OR 18.6 [95%CI: 1.1–326.4], p=0.0035), reduced capillary density (n<7/mm) in digit III of the right hand in NC (OR 9.0 [95%CI: 1.1–73.6], p=0.0266), missing initial enhancement in FOI in digit III of the right hand (OR 3.857 [95%CI: 1.2–12.8], p=0.0323), percentage of pathologic (i. e. narrowed or occluded) vessels >35% in CDUS (OR 4.286 [95%CI: 1.5–12.4], p=0.0099). Criteria with greater OR should impact the score to a higher degree so we appointed three points to dichotomous criteria with OR >10, two points for criteria with OR between 5–10, and one point for criteria with OR <5. Regarding the NC pattern, 3 points were given to patients with late pattern, 2 points for active and 1 point for early pattern.

Best results were found for a cut-off of >10 points with obtained sensitivity levels of 95% and specificity levels of 74% in regard to new DU (AUC=0.8687, p<0.0001). In the absence of CDUS and FOI data, specificity levels dropped slightly to 72% with unchanged sensitivity values of 95%.

Conclusions: A new score was introduced with the aim to predict digital ulcers. If applied correctly and with the new imaging techniques proposed, 95% of patients at risk of digital ulcers throughout 12 months could be identified.

Disclosure of Interest: None declared

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AB0810

A COMPARISON OF CLINICAL PRESENTATION AND INCIDENCE RATE OF CARDIOPULMONARY INVOLVEMENT BETWEEN MALE AND FEMALE PATIENTS WITH EARLY SYSTEMIC SCLEROSIS

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Background: Data regarding the incidence rate (IR) of cardiopulmonary involvement in comparison between male and female patients with early systemic sclerosis (SSc) are limited.

Objectives: To compare the prevalence of clinical manifestations and the IR of cardiopulmonary involvement comparing between male and female patients with early SSc.

Methods: An inception cohort of early SSc patients (disease duration ≤ 3 years from the first non-Raynaud's phenomenon) seen at the Rheumatology clinic, Maharaj Nakorn Chiang Mai Hospital, Thailand, between January 2010 and June 2016, was used. All patients were assessed for clinical manifestations and underwent ECG, echocardiography, and HRCT at the study entry and every 12 months thereafter. In the last visit, the cumulative clinical manifestations were recorded. Patients who continue to follow up at least 12 months were included for analysis.

Results: One hundred and fifteen patients (89 dcSSc) with a mean (SD) disease duration of 11.6 (8.8) months at cohort entry were enrolled during a mean (SD) observation period of 3.8 (1.6) years. There were 46 male patients (40%). Tests for anti-topoisomerase I and anti-centromere antibodies were positive in 91 (79.1%) and 7 (6.1%) patients, respectively. The male group had higher prevalence of dcSSc subtype (91.3% vs. 69.5%, $p=0.006$) and positive test for anti-topoisomerase I antibody (89.1% vs. 72.5%, $p=0.031$) compared with the female group. At enrollment, the male group had higher prevalence of hypo-hyperpigmentation (84.8% vs. 65.2%, $p=0.021$), suspected myositis (26.1% vs. 10.1%, $p=0.024$), and right ventricular dysfunction (8.7% vs. 0%, $p=0.024$) compared with the female group. In the last visit, the male group had higher cumulative prevalence of digital ulcer (47.8% vs. 27.5%, $p=0.026$), telangiectasia (93.5% vs. 69.6%, $p=0.002$), joint contracture (69.6% vs. 43.5%, $p=0.006$), tendon friction rub (39.1% vs. 20.3%, $p=0.027$), LVEF $<50\%$ (21.7% vs. 8.7%, $p=0.048$) and right ventricular dysfunction (34.8% vs. 7.2%, $p<0.001$) compared with the female group. The male group had significant higher IR of right ventricular dysfunction (8.21 vs. 1.99 per 100 person-years, $p=0.006$) and interstitial lung disease (ILD) determined by HRCT (63.66 vs. 39.19 per 100 person-years, $p=0.022$) compared to the female group. However, no significant IR of LVEF $<50\%$ (5.63 vs. 2.10 per 100 person-years, $p=0.078$), and elevated systolic pulmonary artery pressure ≥ 50 mmHg determined by echocardiography (2.81 vs. 1.55 per 100 person-years, $p=0.339$) were observed among the two groups.

Conclusions: Our study cohort found that male gender had higher proportion of dcSSc subtype and had more severe clinical manifestations compared with female. In addition, during the follow-up period, the IR of right ventricular dysfunction and ILD were higher in male than that in female patients in the early stage of SSc.

Disclosure of Interest: None declared

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AB0811

INCREASED LEVELS OF SERUM WISTERIA FLORIBUNDA AGGLUTININ-POSITIVE MAC-2 BINDING PROTEIN IN SYSTEMIC SCLEROSIS

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Background: Mac-2 binding protein is a cell-adhesive glycoprotein of the extracellular matrix secreted as a ligand of galectin-3 (Mac-2). Recently, a Wisteria floribunda agglutinin positive-M2BP (M2BP) assay developed using a lectin-antibody sandwich immunoassay has shown promise as a new fibrotic marker in liver fibrosis and interstitial lung disease (ILD) to detect unique fibrosis-related glycoalteration. Systemic sclerosis (SSc) is an intractable connective tissue disease that causes skin and organ fibrosis. Progressive fibrosis is now recognised to be one of the major causes of morbidity and mortality in SSc.

Objectives: The aim of this study is to evaluate the utility of serum M2BP levels in patients with SSc.

Methods: We retrospectively measured serum M2BP levels in 76 patients with SSc and 16 healthy controls (HC). We excluded patients of viral hepatitis and cirrhosis. Serum levels of M2BP were measured using HISCL[®] M2BP glycosylation isomer Assay Kit. IHC was performed on 5 samples of normal skin, 7 stored samples of SSc skin and 2 samples of SSc-ILD lung tissue. We examined the relationship between serum M2BP levels and clinical parameters in patients with SSc.

Results: The mean age was 62.7 years in SSc patients and 54.6 years in HC. There were 57 limited cutaneous SSc patients and 19 diffuse cutaneous SSc patients. The mean duration of SSc patients was 9.8 years. Serum M2BP levels were significantly higher in patients with SSc than in HC (0.853 ± 0.701 cutoff index [COI], 0.325 ± 0.111 COI, respectively; $p<0.00001$). Receiver operating characteristic (ROC) curve analysis indicated that serum M2BP correctly differentiated between SSc patients and HC with a sensitivity of 81.6% and specificity of 87.5%. In patients with SSc, a significant correlation was not found between serum M2BP levels and age, sex, mRSS, ILD, pulmonary artery hypertension (PAH), SSc-types, dermatoscopy findings. In multivariate analysis, disease duration was a only independent factor for higher serum M2BP levels more than 0.64 COI, serum M2BP median (odds ratio 0.935, 95% CI 0.878–0.996, $p=0.0385$). We detected the protein level of M2BP in SSc patient's fibroblast of skin and SSc Patient's proliferating type II alveolar cells adjacent to fibrotic lesions of lung tissue.

Conclusions: In this study, for the first we demonstrated that M2BP is expressed in skin and lung tissue in SSc patients in IHC and more over the serum M2BP level tends to be higher in patients with short disease duration. It is reveal that M2BP play an important role of fibrosis in early stage of SSc. The serum level of M2BP may be a novel biomarker of SSc.

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AB0812

ULTRASONOGRAPHY FOR THE ASSESSMENT OF SKIN INVOLVEMENT IN SYSTEMIC SCLEROSIS: A SYSTEMATIC REVIEW

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Background: Skin involvement is the clinical hallmark of systemic sclerosis (SSc). A palpation-based semi-quantitative tool, the modified Rodnan skin score, is the current gold standard for the assessment of skin involvement. However, this method has significant limitations, with emphasis on low sensitivity to change and a high inter-observer variability. Ultrasound has been explored, in the past decades, as a basis for more objective, sensitivity and reproducible measure of skin involvement.

Objectives: To identify and synthesise the best available evidence on the use of ultrasound as a source of skin outcome measures in patients with SSc.

Methods: Two independent reviewers systematically searched Pubmed MEDLINE and Embase (up to December 2017). A study was eligible if it included at least one defined group of patients with SSc, and reported a structured evaluation of the skin with ultrasound and/or ultrasound elastography. This search was augmented by review of bibliographic references from the included studies. The same two reviewers independently extracted the data and classified the quality of evidence of the included studies by the Effective Public Health Practice Project system.

Results: A total of 30 studies (21 observational cross-sectional and 9 longitudinal) were included. They enrolled a total of 1171 SSc patients, mostly middle-aged, female and with a limited form of SSc (59.0%). The most common ultrasound outcome measure used was skin thickness (in 28 studies), although the definition of this parameter was a highly heterogeneous. Other outcome measures were echogenicity (in 7), and/or stiffness (in 6) and/or vascularity (in 1). There was a substantial discordance in the number and exact location of skin sites examined. The main comparator was global and site specific mRSS.

There was a lack of construct and criterion validity (only 1 study assessed the correspondence between ultrasound measures and histological findings). Few studies reported information about inter- and intra-rater reproducibility, but when reported, it showed excellent results. Data regarding evidence for responsiveness to change and feasibility were also scarce.

Conclusions: This systematic review highlights a remarkable literature heterogeneity and limited quality of most reported studies. This hinders the evidence currently supporting the use of skin ultrasound evaluation in clinical practice, but the very promising data (e.g good reliability and early detection of skin involvement) support its use in clinical research. Further well-designed and dimensioned studies are needed to support the role of skin ultrasound assessment in the early diagnosis and monitoring SSc patients. These may be crucial to improve our