Conclusion: The results from this exploratory study in a large cohort of SSC patients provide valuable insights into factors associated with digital pitting. Patients with digital pitting often have an earlier onset of Raynauds and of SSC and significantly more debridements/amputations, suggesting that digital pitting is associated with vascular disease severity. Our findings indicate the need for further research investigating pathophysiology of digital pitting, to inform development of preventative treatment strategies.

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AB0623

RATE AND PREDICTIVE FACTORS ASSOCIATED WITH SUSTAINED REMISSION IN IDIOPATHIC INFLAMMATORY MYOSITIS

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Background: Idiopathic inflammatory myositis (IIM) is a group of heterogeneous connective tissue diseases, characterised by chronic muscle inflammation, myositis-specific or myositis-associated autoantibodies and different extra-muscular features. Achieving low disease activity or remission in patients with IIM has proven to be difficult due to the wide clinical spectrum of the different IIM types.

Objectives: To retrospectively assess any predictive factors for sustained remission in IIM patients

Methods: We retrospectively analyzed data taken from medical charts, which included age at disease onset, gender, laboratory data as well as clinical features present at onset, organ involvement and treatment history. A total of 151 adult patients with IIM followed-up for > 1 year were retrospectively enrolled. Remission was defined as no clinical and laboratory evidence of disease activity persistent for more than 6 months during follow-up, while undergoing myositis therapy or under no medication. The remission of cutaneous involvement was defined as no current activity of skin rash, absence of Gottron's papules as well as heliotrope rash and erythema, whereas the remission of pulmonary involvement was considered as no requirement for intensification of immunosuppressive therapy during follow-up. Likewise, absence of muscle weakness or hyposthenia was taken into account for evaluating muscle involvement. Moreover, the clinical features were accompanied by normalization of myogenic enzymes such as creatine kinase (CK) and lactate dehydrogenase levels.

Results: Among all 151 patients, 89 (58.9%) patients achieved sustained remission. By univariate analysis, overlap myositis (7.9% vs 27.4%; $p=0.003$; OR:0.22), cancer-associated myositis (CAM) (7.8% vs 19.35%; $p=0.046$; OR:0.3), as well as the presence of anti Ku (3.37% vs 12.9%; $p=0.05$; OR:0.23) and anti TIF-1 gamma (1.1% vs 8%; $p=0.043$; OR:0.13) antibodies and polyarthritis (11.2% vs 24.19%; $p=0.045$; OR: 0.397) at onset were significantly associated with active IIM, not achieving remission.

Out of 89 patients in remission, 79 (88.8%) achieved long-term sustained remission, lasting at least 2 years, whereas 10 patients (11.2%) showed relapse. Most of relapsed patients showed a relapse/remitting disease, with no evident trigger for the relapse. We divided our cohort of 89 patients in remission in 2 subsets: 83 patients in remission undergoing therapy (93.3%) and 6 patients in drug-free remission (6.7%). No significant differences were found between two groups, except for younger age at onset ($p=0.0002$) found in patients achieving drug-free remission.

Conclusion: Sustained remission occurs in about one half of patients with IIM. The presence of anti Ku and anti TIF-1 gamma antibodies as well as polyarthritis at onset lowers the chance of achieving sustained remission. Younger age at diagnosis has proved to predict drug-free long-lasting remission.

Disclosure of Interests: None declared

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AB0624

PREDICTIVE PARAMETERS FOR DEVELOPMENT OF INTERSTITIAL LUNG DISEASE IN IDIOPATHIC INFLAMMATORY MYOSITIS

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Background: Idiopathic Inflammatory Myositis (IIM) is a group of heterogeneous connective tissue diseases, primarily characterized by chronic muscle inflammation as well as myositis-specific or myositis-associated autoantibodies and a spectrum of different extra-muscular features.

The most frequent organ involvement in IIM is Interstitial Lung Disease (ILD), occurring in 5-80% of different IIMs cases and considered the hallmark of morbidity and mortality in patients with IIMs.

Objectives: To retrospectively assess the predictive factors for development of ILD in IIM patients

Methods: We retrospectively analyzed the prevalence of ILD in a single-center cohort of 165 IIM patients. Patient data was collected from clinical charts. ILD was diagnosed by chest X-ray scan and chest CT scan. All chest CT and chest X-ray scans available and performed at our hospital were consequently re-evaluated by our expert pneumologist for uniform evaluation.

Results: Myositis-related ILD (M-ILD) was found in 52 IIM patients (31.5%): 46.15% was affected by anti-synthetase syndrome (ARS), 21.15% by polymyositis (PM), 19.23% by dermatomyositis (DM) and 13.46% by overlap myositis. The pulmonary involvement was characterized by Non-specific interstitial pneumonia (NSIP) (30.6%), Unusual Interstitial Pneumonia (UIP) (38.77%), Bronchiolitis Obliterans with Organizing Pneumonia (BOOP) (20.4%), overlap NSIP/BOOP (4.1%) and Undetermined/Unspecific pattern (6.12%). Eighty four percent of M-ILD consisted of non-smokers and 69.23% presented with dyspnea at onset.

ILD was diagnosed in 90.38% of patients within the first year of IIM diagnosis (early onset ILD) and was associated with dyspnea and/or cough in 70.2% and 17% respectively. On the other hand, late onset ILD presented mostly with dyspnea and/or cough in 60% of cases and was significantly associated with anti-Ku antibodies.

At onset ILD was significantly associated with: ARS ($p<0.0001$; OR:12.98), anti-Jo-1 ($p<0.0001$; OR:6.1), anti-Ro ($p=0.038$; OR:2.2), mechanic's hands ($p<0.0001$; OR:10.41), arthritis ($p=0.01$; OR:2.58), polyarthritis ($p=0.001$; OR:4.578), dyspnea ($p<0.0001$; OR:9.66), and high levels of CPK ($p=0.0001$) and GOT ($p=0.0146$). By contrast, the following features: DM ($p=0.012$; OR:0.36), facial rash ($p=0.003$; OR:0.31), anti-NXP-2 ($p=0.019$; OR<0.0001), anti-PL-12 ($p=0.03$; OR<0.0001) and myositis ($p<0.0001$; OR:0.173) present at onset were less frequently associated with M-ILD.

At multivariate analysis M-ILD was predicted by anti-Ro ($p=0.0448$), polyarthritis ($p=0.0093$) and dyspnea ($p=0.0001$) at onset. On the other hand, patients presenting myositis ($p=0.0383$) and facial rash ($p=0.0398$) at onset were less likely to develop M-ILD.

Conclusion: ILD occurs in about one third of patients with IIM, mostly affected by ARS. The presence of anti-Ro antibodies as well as polyarthritis and dyspnea at onset predict the development of ILD.

Disclosure of Interests: None declared

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AB0625

IS PULMONARY ARTERIAL HYPERTENSION, ASSOCIATED WITH SYSTEMIC SCLEROSIS REVERSIBLE?

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Background: Systemic sclerosis (SSc) is one of the frequent causes of the pulmonary arterial hypertension (PAH) which found in 6-15% patient with SSc. Patients with PAH, associated with SSc have a poorer prognosis than other forms of PAH. 6 World Symposium of PH lowered diagnostic cut-off to 21 mm Hg in hope of improved survival. PAH reversibility is described in congenital heart defects, HIV and some tumors

Objectives: The aim of the study to detect reversibility of PAH associated with SSc.

Methods: Hemodynamics (mean pulmonary artery pressure (MPAP), pulmonary artery wedge pressure (PAWP), cardiac output (CO) and pulmonary vascular resistance (PVR)), functional class (NYHA), 6-minute walk distance (6MWD), biomarkers and DLCO were assessed. Patients with pulmonary fibrosis and left heart diseases were excluded.

Results: The study includes 56 patients receiving start-up monotherapy with 1st generation PAH-specific drugs: bosentan (25 patients) and sildenafil (31 patients). The median age of the patients was 51.5 (37; 58) years. At the time of diagnosis, the MPAP in the sildenafil group was 49 (30; 50) mm Hg, bosentan - 50 (42; 56) mm Hg, differences not significant ($p = 0.11$). During observation against the background of sildenafil intake,

the MPAP decreased by -3 (-7; 0) mm Hg, bosentan -6 (-13; -2) mm Hg. Decreasing of MPAP $\leq$ 21 mm Hg was at seven patients receiving sildenafil. Initial hemodynamic values in this group of patients was MPAP 25 (21; 27) mm Hg., RAP 2 (1; 5) mm Hg, PAWP 7 (6; 10) mm Hg., CV 5.9 (4.7; 6.7) l/min, PVR 3.0 (1.5; 4.2) Wood Unit. Among patients taking sildenafil, there were seven who had MPAP less than 35 mmHg, and among patients taking bosentan, there were 4 patients with less than 35 mmHg. However, the decreasing was observed only group who received sildenafil. ROC – analysis shown that cut-off for reversibility is 29 mm Hg (AUC 0,977, sensitivity 97%, and specificity 100%)

Conclusion: In patients with PAH, associated with SSc, on sildenafil treatment, at MPAP < 29 mm Hg reversibility is possible. Further study is required.

Disclosure of Interests: None declared

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AB0626 IMATINIB FOR THE TREATMENT OF SYSTEMIC SCLEROSIS: RATIONALE, CLINICAL EVIDENCE AND FUTURE DEVELOPMENT

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Background: Systemic sclerosis (SSc) is a chronic disorder of connective tissue affecting the skin and internal organs. The molecular mechanisms behind SSc are not entirely understood, but recent advances highlight key signalling pathways¹ (see Figure 1). Fibrosis disrupts tissue architecture resulting in organ dysfunction. This causes significant morbidity and mortality¹, therefore there is a clear need for identifying efficacious antifibrotic treatment.

Imatinib is a tyrosine kinase inhibitor with established use in some malignancies, and existing evidence that it can treat SSc².

Objectives: The aim of this literature review is to summarise the current evidence and future developments of imatinib as antifibrotic treatment in SSc.

Methods: PubMed headings “systemic sclerosis,” “scleroderma,” “imatinib” and synonyms were used. See Figure 2.

Results:

Lead author and year	Study design	Population	Intervention (imatinib dose given daily)	Results
Hinchcliff ME. 2016	Case study	1 female (F) with an 8-month history of diffuse cutaneous systemic sclerosis (dcSSc)	400mg	Severe adverse events (AEs)
Pope J. 2014	Early phase proof of concept trial	10 with dcSSc.	9 given 400mg, 1 placebo	Poor drug tolerability
Prey S. 2012	Double blind RCT	28 with SSc	15 given 400mg, 13 placebo	No improvement (mRSS)
Khanna D. 2011	Phase I/IIa pilot trial	20 with SSc	Up to 600mg	AEs present
Pope J. 2011	Double blind RCT	10 with SSc.	9 given 400mg, 1 placebo	No improvement, AEs present
Gordon J. 2014	Open label, single arm, extension phase clinical trial	17 with average disease duration of 3.5 years	100-400mg	↓ modified Rodnan skin score (mRSS)
Fratlicelli P. 2014	Phase II pilot trial	30 with SSc	200mg	↑ lung function
Guo L. 2012	Case series	6 F, Chinese pts with SSc	200mg	↓ mRSS and ↑ lung function
Divekar AA. 2011	Single centre, open-label study	15 with SSc	100mg up to 600mg/day	↑ lung function
Spiera RF. 2011	1-year, phase IIa, single-arm, open-label clinical trial	30 with dcSSc	400mg	↓ mRSS and ↑ lung function
Freyhaas H. 2009	Case report	58yo F	400mg	↑ lung function
Chung L. 2009	Case report	2 with early dcSSc	200mg	↓ mRSS and ↑ lung function
Van Daele PL. 2008	Case report	69yo F	400mg	↓ mRSS and ↑ lung function
P. P. Sfikakis. 2008	Case report	24yo F	400mg	↓ mRSS and ↑ lung function

9 studies showed imatinib had positive efficacy in the treatment of SSc. 5 showed no improvement or adverse effects.

Conclusion: Overall, current evidence suggests that imatinib can be a useful drug to improve manifestations of SSc, for some. Despite inconclusive evidence, a dose-dependent relationship seems to exist for imatinib toxicities, with more research needed to ascertain a safe dose.