12/15 remained stable during the entire follow-up.These positive clinical changes were mirrored by the subjective improvement of patients' well being in all cases (HAQ from 1.45±0.49 to 0.98±0.38, VAS from 64.5±18.5 to 37.5±8.6). Finally, no significant side effects were observed.
Conclusions: The present study reinforces the previous trials showing the usefulness of RTX in the management of SSc patients, along with its good safety profile. The specific therapeutical activity of RTX, able to down-regulate the B-cell over expression, might explain its beneficial effects on some SSc clinical manifestations; in particular the improvement of both skin sclerosis and articular involvement, along with the possible stabilization of lung fibrosis.
Disclosure of Interest: None declared
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FRI0389 FOOT INVOLVEMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS: A SINGLE-CENTRE REPORT

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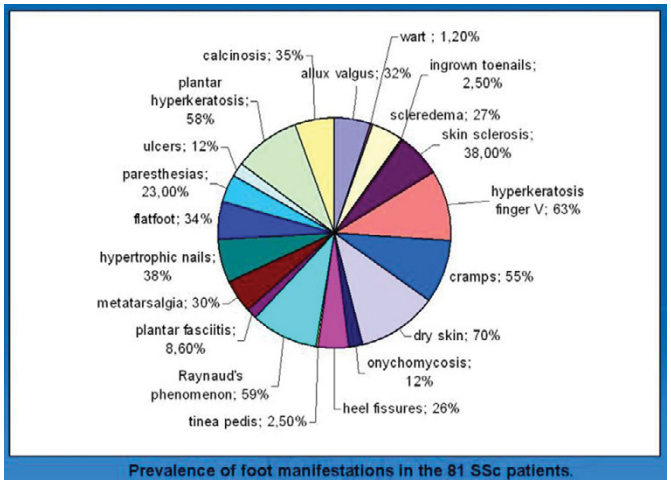
Background: Foot involvement can be a source of morbidity and disability in patients with systemic sclerosis (SSc). Some studies have previously reported severity, echographic and radiographic manifestations of foot involvement in SSc (1,2).
Objectives: The aim of our study was to assess the nature and prevalence of foot problems in patients with SSc reporting a single centre experience.
Methods: A podiatrist and a rheumatologist assessed 81 (76 female) consecutive patients attending our SSc outpatient clinic. The mean age was 50 years (range 21–70). Thirteen (16%) had diffuse cutaneous SSc with a median disease duration from Raynaud's Phenomenon of 5 years (range 3 –19); 68 (84%) had limited cutaneous SSc with a median disease duration of 11.5 years (range 1–41). The overall median disease duration was 11 years (range 1–41). Thirty (37%) were anticentromere antibodies positive, thirty five (43%) anti-scl70 antibodies positive. The two investigators evaluated the presence of the following features: colour changes, pain, previous ulceration, current ulceration, pre-ulceration (discoloration and thinning of the skin), toenail changes, hyperkeratosis, calcinosis, onychomycosis, dry skin, skin sclerosis, warts, scleredema, flatfoot. The presence of paresthesias, cramps, metatarsalgia were also investigated.
Results: The diagram reports the prevalence of foot manifestations in the 81 SSc patients.
Most SSc patients suffer from symptoms related to their feet, particularly dry skin (70%), hyperkeratosis (plantar 58%, finger V 63%), Raynaud's phenomenon (59%), cramps (55%). No statistically significant differences were found between diffuse and limited SSc groups.



Prevalence of foot manifestations in the 81 SSc patients.

Conclusions: Our study suggests that, in patients with SSc, foot problems are common and potentially disabling. A careful assessment of the feet should always be performed in these patients, in order to identify problems at an early stage.
References:
[1] La Montagna G et al. Semin Arthritis Rheum 2002.
[2] Sari-Kouzel H et al. Rheumatology (Oxford) 2001.
Disclosure of Interest: None declared
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FRI0390 EUROPEAN MULTICENTRE STUDY VALIDATES ELF TEST AS BIOMARKER OF FIBROSIS IN SYSTEMIC SCLEROSIS

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Background: The Enhanced Liver Fibrosis (ELF) test is a serum test including the serum concentrations of amino-terminal pro-peptide of procollagen type III (PIIINP), tissue inhibitor of matrix metalloproteinase-1 (TIMP-1) and hyaluronic acid (HA). A recent single centre study showed that ELF score and its components are markers of overall fibrosis in systemic sclerosis (SSc) mainly reflecting skin and lung involvement (1).
Objectives: To determine the value of ELF score and its single analytes in an independent multicentre cohort of SSc patients.
Methods: 254 SSc patients from 6 European Rheumatology Centres were included in this study. Clinical data were collected at time of sampling. Serum samples were collected and stored according to EUSTAR biobanking recommendations (2). Sera were analysed employing a high-throughput in vitro diagnostic (Siemens Alpha-Centaur). Statistical analysis was performed with SPSS software V.24.
Results: The 254 SSc patients had a mean age 55.8±13.8 years, and included 209 females and 80 patients with diffuse cutaneous SSc (dcSSc). ELF score was overall higher in males than in females (p=0.0236) as well as in dcSSc compared to limited cutaneous SSc patients (p=0.0015). ELF score and the single markers significantly correlated with the degree of skin involvement (mRSS) and inversely correlated with FVC%, TLC% and DLCO%. Concordantly all markers significantly correlated with skin and lung severity as assessed by the Medsger's scale (Table 1). TIMP-1 and PIIINP levels were higher in patients with lung fibrosis assessed by chest HRCT scan (p=0.0126 and p=0.0308 respectively). Significant correlation (p<0.0001) was found between ELF score, TIMP-1, PIIINP, HA and total disease severity and activity. Multivariate analysis indicated that age (p<0.0001), mRSS (p<0.0001) and DLCO% (p=0.005) were independently associated with ELF score.

Table 1. Coefficient correlation (r) between ELF score, PIIINP, TIMP-1, HA serum levels and clinical variables

	ELF score	PIIINP (ng/mL)	TIMP-1 (ng/mL)	HA (ng/mL)
Serum values (median, range)	8.86, 6.22–12.2	6.44, 0.91–34.63	221, 19.09–595.4	36.98, 4.52–355.5
	r	r	r	r
Age	0.41****	0.06	0.26****	0.52****
mRSS	0.36****	0.29****	0.18**	0.16*
FVC%	-0.12	-0.25***	-0.2**	0.02
TLC%	-0.21**	-0.29****	-0.31****	-0.09
DLCO%	-0.25****	-0.39****	-0.30****	-0.18**
Sev_skin	0.28****	0.27****	0.22***	0.17**
Sev_lung	0.25****	0.29****	0.3****	0.18**
Sev_total	0.32****	0.27****	0.33****	0.23***
EScSG-AI	0.30****	0.26****	0.27****	0.22***

*p<0.05; **p<0.01; ***p<0.001; ****p<0.0001.

Conclusions: The value of ELF score as independent marker of skin and lung involvement is confirmed in a second independent multicentre cohort of SSc patients. A longitudinal study paired with analysis of large cohort of healthy controls is currently on going to identify a SSc specific test with the highest predictive value for skin and lung progression independently of age and gender.
References:
[1] Abignano G et al. Ann Rheum Dis. 2014.
[2] Beyer C et al. Ann Rheum Dis. 2011.
Disclosure of Interest: None declared
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FRI0391 A PERFUSION-METABOLIC MISMATCH IN 99MTL AND 123I-BMIPP SCINTIGRAPHY PREDICTS WORSE PROGNOSIS IN SYSTEMIC SCLEROSIS PATIENTS WITH ASYMPTOMATIC CARDIAC INVOLVEMENT

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Background: Cardiac involvement is a manifestation of systemic sclerosis (SSc) that contributes to significant mortality and morbidity. Since many SSc patients with cardiac involvement are asymptomatic and it may progress silently, early diagnosis is still challenging. The perfusion-metabolic mismatch in cardiac scintigraphy indicates functional abnormality due to myocardial injury and has been expected to detect early cardiac involvement in various diseases. However, its clinical utility has been poorly evaluated in patients with SSc.

Objectives: To evaluate clinical utility of perfusion and myocardial fatty acid metabolism mismatch in SSc patients without cardiac failure.
Methods: All patients who visited St. Marianna University Hospital from 2009 to 2015 and performed cardiac scintigraphy using ^{99m}Tl and ^{123}I-BMIPP were retrospectively evaluated. Patients who fulfilled American Collage of Rheumatology classification criteria of SSc and systemic lupus erythematosus (SLE) were selected. We subtracted %uptake of metabolism (^{123}I-BMIPP) from that of perfusion (^{99m}Tl) on each 17 myocardial segments standardized by American Heart Association. We compared sum of all the scores and each score in 3 coronary artery regions between patients with SSc and SLE. Furthermore, we evaluated incidence of cardiac death or cardiac failure depending on the scores of each coronary artery regions.
Results: Among 177 patients, we analyzed the data in 22 cases with SSc and 23 with SLE. The sum of all mismatch scores was not significantly different between the 2 groups (p=0.37). The mismatch score of left anterior descending coronary artery (LAD) region was significantly higher in SSc than SLE (p=0.05) (Figure 1A). We next divided SSc patients into 2 groups depending on degree of the LAD mismatch score and found the group with low score had higher incidence of cardiac death or cardiac failure (p=0.04) than that with high score (Figure 1B).

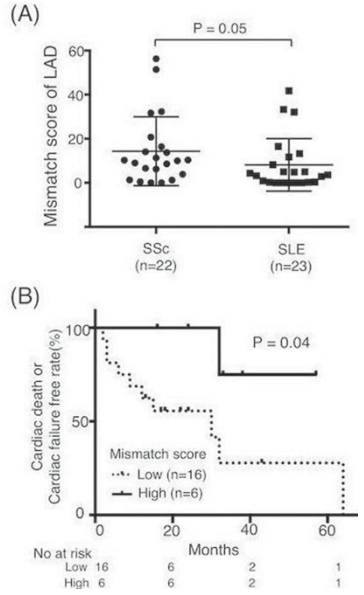

Figure 1

Conclusions: The low mismatch score in LAD region of cardiac scintigraphy could be associated with cardiac death or cardiac failure in SSc patients. Since myocardial fibrosis might replace viable cardiomyocytes leading to loss of ventricular volume, metabolic defect by scintigraphy might be less detectable.
Disclosure of Interest: None declared
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FRI0392 OPPORTUNISTIC INFECTIONS IN PATIENTS WITH MYOSITIS. A RETROSPECTIVE COHORT STUDY

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Background: Idiopathic inflammatory myopathies, also known as myositis, are a heterogeneous group of acquired diseases of probable autoimmune origin, characterized by the presence of inflammatory muscle infiltrates.Infectious complications are not uncommon in myositis, and some predisposing factors, such as upper esophageal involvement, calcinosis cutis, ventilatory insufficiency due to diaphragm involvement, and immunosuppressive drug therapies, seem to be implicated.
Objectives: To describe the prevalence, clinical characteristics, and risk factors for opportunistic infection (OI) in a cohort of patients with inflammatory myopathies, and compare mortality rates between those with and without OIs.
Methods: In total, 204 patients from our myositis cohort were reviewed to identify patients who had experienced an OI during the period 1986–2014. The patients' clinical characteristics, treatment received, and outcome were systematically recorded. Disease activity at the OI diagnosis and cumulative doses of immunosuppressive drugs were analyzed, as well as the specific pathogens involved and affected organs.
Results: The prevalence of OI in the total cohort, was 6.4%: viruses, 44.4% (varicella-zoster virus, cytomegalovirus); bacteria, 22.2% (*Salmonella sp.*, *Mycobacterium tuberculosis*, *M. chelonae);* fungi, 16.7% (*Candida albicans*, *Pneumocystis jirovecii);* and parasites, 16.7% (*Toxoplamosis gondii, Leishmania spp.*) were the pathogens detected. Lung and skin/soft tissues were the organs most commonly affected (27.8%). Overall, 55.6% of OIs developed during the first year after the myositis diagnosis and were significantly associated with administration of high-dose glucocorticoids (p=0.0148). Fever at onset of myositis (p=0.0317), biological therapy (p<0.001) and sequential administration of 4 or more immunosuppressive agents during myositis evolution (p=0.0032) were significantly associated with OI. All-cause mortality in the OI group was 3.69 deaths per 100 patients/year versus 3.40 in the remainder of the cohort (p=0.996).
Conclusions: The prevalence of OI was 6.4% in our myositis cohort. High-dose glucocorticoids at disease onset and severe immunosuppression are implicated in the development of these complications. Mortality did not differ from the remainder of the cohort.
Disclosure of Interest: None declared
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FRI0393 PREVALENCE OF MYOSITIS-SPECIFIC ANTIBODIES IN IDIOPATHIC INFLAMMATORY MYOPATHY COMPARED TO DISEASE AND HEALTHY CONTROLS

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Background: Myositis-specific autoantibodies (MSA) are increasingly recognized as important diagnostic and prognostic markers in idiopathic inflammatory myopathies (IIM) (polymyositis, dermatomyositis, sporadic inclusion body myositis and necrotizing autoimmune myositis). The prevalence of these MSAs in other systemic autoimmune rheumatic diseases and neuromuscular diseases is unclear.
Objectives: To compare positivity of MSA in a cohort of IIM patients to positivity in healthy controls and different systemic autoimmune rheumatic diseases (SARD) or chronic inflammatory demyelinating polyneuropathy (CIDP).
Methods: A line immunoassay (Myositis 12 IgG DOT for BlueDiver) for IgG autoantibodies against Jo-1, PL-7, PL-12, EJ, SRP, Mi-2, MDA-5, TIF1-γ, HMGCR, SSA/Ro52kD, SAE1/2 and NXP-2 antigens was performed in patients with IIM (n=146), healthy controls (blood donors, n=40) and disease controls (n=200). The disease control group consisted of patients with other SARD (rheumatoid arthritis, RA; systemic sclerosis, Ssc; Sjögren's syndrome, SjS; and systemic lupus erythematosus, SLE) (n=40 for each disease group) and CIDP (n=40). A result >10 arbitrary units was considered significantly positive.
Results: 50% of 146 patients with IIM tested positive for an MSA (table 1), compared to 3,5% of 200 disease controls (table 1). 1 SSc patient was positive for Jo-1, 1 CIDP patient was positive for PL12, 2 SSc patients were positive for TIF1-gamma, 2 patients (1 SSc and 1 SjS patient) were positive for SAE1/2, and 1 SjS patient was positive for NXP2. The prevalence of SAE1/2 and TIF1-gamma positivity was similar in the IIM and disease control group. No healthy control had a significantly positive MSA.

Antibody detected	IIM (n=146)	Disease controls (n=200)	Healthy controls (n=40)
1 positive (>10 AU) MSA detected	68 (47%)	7 (3,5%)	0 (0%)
Jo-1	28 (19%)	1 (0,4%)	0 (0%)
PL12	3 (2%)	1 (0,4%)	0 (0%)
PL7	1 (1%)	0 (0%)	0 (0%)
EJ	2 (1%)	0 (0%)	0 (0%)
Mi-2	7 (5%)	0 (0%)	0 (0%)
MDA-5	7 (5%)	0 (0%)	0 (0%)
SAE1/2	3 (2%)	2 (0,8%)	0 (0%)
TIF1-gamma	2 (1%)	2 (0,8%)	0 (0%)
NXP2	5 (3%)	1 (0,4%)	0 (0%)
SRP	3 (2%)	0 (0%)	0 (0%)
HMGCR	7 (5%)	0 (0%)	0 (0%)
>1 positive (>10 AU) MSA detected	4 (3%)	0 (0%)	0 (0%)
Jo-1 + EJ	1		
Jo-1 + NXP2	1		
SAE1/2 + NXP2	1		
Jo-1 + SAE1/2 + NXP2	1		

Conclusions: MSA positivity in patients with a clinical non-IIM diagnosis (other SARD or CIDP) is infrequent compared to positivity in the IIM group. For TIF1-gamma and SAE1/2 assay performance may need to be optimized. The distribution of subtypes of MSA in this IIM cohort is consistent with data of previous studies.[1]
References:
[1] Allenbach Y, Benveniste O. Diagnostic Utility of Autoantibodies in Inflammatory Muscle Diseases. J Neuromuscul Dis. 2015; 2:13–25.