Conclusion: The period prevalence of AEs recorded during MMF treatment appeared as high as 71.7% in our real life setting. Even though we did not record any serious AE, our analysis suggests a close monitoring of this therapy in SSc.
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OP0064 EVIDENCE-BASED CONSENSUS RECOMMENDATIONS FOR THE IDENTIFICATION AND MANAGEMENT OF INTERSTITIAL LUNG DISEASE IN SYSTEMIC SCLEROSIS

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Background: Interstitial lung disease in systemic sclerosis (SSc-ILD) occurs frequently and carries a high burden of morbidity and mortality. To date, there are no existing guidelines for screening, diagnosis and management of SSc-ILD that would aid early recognition and treatment and improve the care of these patients.
Objectives: To develop expert consensus recommendations for the identification and management of SSc-ILD.
Methods: Based on the results of a comprehensive systematic literature analysis conducted in line with NICE/CRD and IQWiG guidelines and PRISMA methodology, evidence-based statements on SSc-ILD risk, screening, diagnosis, treatment and follow-up were developed. A modified Delphi process was then used to establish consensus statements for the identification and management of SSc-ILD. Briefly, an expert panel of 27 European-based pulmonologists, rheumatologists and internists with experience in treating SSc-ILD was established. Between July and November 2018, the panel took part in 3 rounds of online surveys, a face-to-face discussion and a WebEx meeting to establish consensus-based recommendations for the management of SSc-ILD. Statements were categorised by topic: risk factors (including biomarkers); screening; diagnosis; assessment of severity; treatment initiation; treatment options; disease progression; treatment escalation; other management options. Panellists indicated their level of agreement with proposed statements on a scale of 1 (strong disagreement) to 7 (strong agreement), and consensus was considered achieved when $\geq$80% either disagreed (score of 1–3) or agreed (score of 5–7) with a statement. Based on panel feedback, statements that did not reach consensus were modified and re-voted in later rounds.
Results: At the close of the Delphi process, the panel agreed on the following:

1. Risk factors: The presence of anti–topoisomerase I antibodies, male gender and diffuse cutaneous SSc all increase risk for ILD
2. Screening: All SSc patients should undergo screening for ILD, using HRCT and lung function testing. Frequency of screening using HRCT should be guided by risk of developing ILD, in combination with clinical symptoms and lung function
3. Diagnosis and assessment of severity: Use of HRCT to diagnose SSc–ILD and assess severity, with supporting findings from lung function testing and clinical assessment, is also recommended
4. Treatment initiation and options: All patients with severe or progressive SSc–ILD should be considered for pharmacotherapy, with mycophenolate mofetil and cyclophosphamide recommended as treatments. Patients not receiving treatment should be followed closely for signs of disease progression
5. Disease progression: Indicators of progression include sustained decline in lung function, worsening of clinical symptoms, and change in extent and/or pattern of fibrosis on HRCT
6. Treatment escalation: Patients with inadequate treatment responses should be considered for treatment escalation. Suitability for lung transplant should be evaluated early, especially for patients diagnosed with advanced disease. Autologous haematopoietic stem cell transplant may be considered in carefully selected patients

Conclusion: These evidence-based expert consensus recommendations, developed using a modified Delphi process, provide important guidance for the identification and management of SSc-ILD.
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OP0065 THE VERY EARLY DIAGNOSIS OF SYSTEMIC SCLEROSIS (VEDOSS) PROJECT: PREDICTORS TO DEVELOP DEFINITE DISEASE FROM AN INTERNATIONAL MULTICENTRE STUDY

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Background: Early identification of patients is of key importance for the management and treatment of inflammatory rheumatic diseases.
Objectives: The aim of the VEDOSS project (1) is to determine through an at-risk population the predictive factors for the progression toward a definite systemic sclerosis (SSc).
Methods: VEDOSS investigators prospectively recruited patients with Raynaud phenomenon (RP), with or without anti-nuclear antibodies (ANA)for this longitudinal, observational study. Fulfilling the 2013 classification criteria at baseline was an exclusion criterion. Patients with primary Raynaud syndrome were recruited as controls. Patients had an annual assessment according to EUSTAR standards to determine organ involvement and severity. The endpoint was defined as fulfilment of the 2013 classification criteria. The time to fulfilling 2013 classification criteria was evaluated with Kaplan-Meier analysis, and predictors of evolution were determined by univariate and multivariate Cox regression.

Figure 1: A matrix was built to show the PPV of various variables alone or in combination to predict subsequent progression of VEDOSS patients toward definite SSc after 3 years

	alone	Disease specic atb	scleroderma patten on NVC	puffy fingers
	PPV%	PPV%	PPV%	PPV%
Disease specific atb	70.2		82.2	94.1
scleroderma patten on NVC	75.0	82.2		77.8
puffy fingers	78.9	94.1	77.8	

Results: 735 patients with RP were recruited into the study. The sample is distributed as follows: i) *237* patients (143 with follow up) RP/ANA negative (ANA$^-$/pRP) as the control group, ii) *498 patients* (401 with follow up) RP/ANA positive (ANA$^+$/pRP)): 87 had puffy fingers (PF), 199 had anti-centromere antibodies (atb) positive, 45 had anti-topoisomerase I atb positive and 182 had nailfold videocapillaroscopy (NVC) abnormalities at baseline. Out of 401 ANA$^+$/pRP patients, 7.4% within 1 year, 29.3% within3 and 44.1% within 5 years satisfied the 2013 classification criteria. Out of the 143 ANA$^-$/pRP patients, none (0%) within 1 year, 4.6% within 3 years, and 4,6% within 5 years satisfied SSc criteria. After adjustment for age, the following baseline parameters were identified as independent predictors for progression into definite SSc by multivariate analysis: puffy fingers (OR=3.4 [2.0;5.6]), anti-centromere atb (OR=2.6 [1.6;4.1]) and anti-topoisomerase 1 atb (OR=3.1 [1.6;5.8]), and NVC abnormalities (OR=1.9 [1.3;2.9]) The presence of PF had a positive predictive value (PPV) of 79% and combination of PF + specific auto-antibodies showed 94% PPV to satisfy ACR/EULAR 2013 criteria within 5 years (figure 1).
Conclusion: the data show that patients with very early SSc develop definite, classification criteria fulfilling SSc within 5 years of follow up. The VEDOSS study