disease 20 (35%) pts, esophageal involvement 29 (51%) pts of them 5 (9%) pts needed enteral feeding. 2 (4%) pts had oncopathology. CK was increased in 49 (86%) pts: < 10 N - 15 (26%) pts, > 10 N - 28 (49%) pts, > 50 N - 6 (11%). ANA positive - in 39 (70%) pts. Antibodies identified in 25(44%) pts: Pm-Scl 7 (12%) pts, PL-7 1 (2%), Mi-2 10 (17.5%), Ku 2 (3.5%) pts, AMA-M2 2 (2%)pts, a-SRP 4 (7%) and SS-A/Ro-52 13 (23%), SS-A/Ro-60 2 (3%) pts.

Conclusion: The group of inflammatory myopathies is characterized by clinical and immunological heterogeneity. Finding antibodies specific for myositis can help diagnose the disease.

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.3101

AB0445

FIRST CLINICAL ANALYSIS OF MYOSITIS PATIENTS REGISTERED AT REUMA.PT/MYOSITIS PROTOCOL: DATA FROM A SINGLE-CENTER

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Background: Idiopathic inflammatory myopathies are a group of rare heterogeneous diseases that require a multidisciplinary and standardized approach.

Table 1. Clinical characteristics and immunological profile of the patients.

Diagnosis (n)	Clinical features					Myositis antibodies (n)
	Skin disease, median [IQR]/ (n)	MMT-8, median disease [IQR] (n)	Lung (n)	Others (n)	Malignancy (n)	
Definite DM (28)	DAS Skin 2 [0-2] (3) Calcinosis (3) Mechanic hands (2)	80 [75-80] (2)	NSIP (2); COP (1)	Arthritis (6)	Breast cancer (1)	anti-Ro52 (6); anti-Mi2b (5); anti-PmScl100 (4); anti-Mi2a (3); anti-NXP2 (3); anti-SAE (2); anti-MDA5 (2); anti-Ku (2); anti-Tif1g (1)
Antisynthetase syndrome (21)	DAS Skin 0 [0-0] (1) Calcinosis (1) Mechanic hands (4)	80 [80-80] (1)	NSIP (1); UIP (3); LIP (1)	Arthritis (15); RP (10)	—	anti-Jo1 (14); anti-Ro52 (12); anti-Mi2b (1); anti-PL7 (3); anti-PL12 (1)
Probable DM (19)	DAS Skin 0 [0-0] (1)	80 [78-80] (1)	COP (1)	Arthritis (5); RP (6)	—	anti-Mi2a (4); anti-Mi2b (2); anti-Tif1g (2); anti-Ku (2); anti-PmScl75 (2); anti-Mi2b (5); anti-Ro52 (2); anti-Mi2a (1); anti-MDA5 (1); anti-EJ (1); anti-SAE (1); anti-SRP (1); anti-Ku (1); anti-Tif1g (1)
CADM (16)	DAS Skin 1 [0-2] (1) Calcinosis (1) Mechanic hands (1)	80 [77-80] (1)	NSIP (1); COP (1)	Arthritis (5); RP (4)	—	anti-Ro52 (2); anti-Mi2a (1); anti-MDA5 (1); anti-EJ (1); anti-SAE (1); anti-SRP (1); anti-Ku (1); anti-Tif1g (1)
MCTD (12)	DAS Skin 0 [0-1] (1)	80 [79-80] (1)	NSIP (1)	Arthritis (8); RP (10)	—	anti-U1 RNP (12); anti-Ro52 (7)
PM (7)	DAS Skin 0 [0-0] (1) Calcinosis (1)	80 [72-80] (1)	NSIP (1)	Arthritis (1)	Ovarian cancer (1); non-Hodg-kin lymphoma (1)	anti-Ro52 (2); anti-SRP (1)
UCTD (5)	DAS Skin 0 [0-2] (1) Calcinosis (1) Mechanic hands (1)	80 [79-80] (2)	NSIP (2)	Arthritis (1); RP (5)	—	anti-ThTo (2); anti-SRP (1); anti-PL12 (1)
Overlap syndromes (12)	DAS Skin 0 [0-2] (2) Mechanic hands (2)	80 [77-80] (2)	NSIP (3); UIP (1)	Arthritis (4); RP (8)	—	anti-PmScl75 (4); anti-PmScl100 (1); anti-Ro52 (1); anti-Ro60 (1); anti-RNAPIII (1); anti-NOR90 (1); anti-MDA5 (1); anti-Ku (1)
Necrotizing myopathy (1)	DAS Skin 0	80	—	—	—	—

DM – dermatomyositis; CADM – clinically amyopathic dermatomyositis; MCTD – mixed connective tissue disease; PM – polymyositis; UCTD – undifferentiated connective tissue disease; ILD – interstitial lung disease; NSIP – nonspecific interstitial pneumonia; LIP – lymphocytic interstitial pneumonitis; UIP – usual interstitial pneumonia; COP – cryptogenic organizing pneumonia; RP – Raynaud phenomenon

Objectives: To clinically characterize patients with inflammatory myopathies followed at an hospital's Rheumatology Department, using the Rheumatic Diseases Portuguese Register (Reuma.pt).

Methods: An observational transversal study was performed. Data on demographic variables, clinical features, antibodies and treatment were collected.

Results: Of the 121 included patients, 78% were female, with a median age of 56 [44-68] years and a median disease duration of 2 [0-4] years. The most frequent diagnosis was dermatomyositis (DM; n=28, 23%) followed by antisynthetase syndrome (ASyS; n=21, 17.4%). At the time of the analysis, the median Manual Muscle Test (MMT-8) was 80 [78-80], the median modified skin Disease Activity Score was 0 [0-2] and global disease activity was 0.5 [0-0.75]. Calcinosis was found in 6% of the patients (n=7), mostly DM cases. Interstitial lung disease (ILD) was present in 29 patients (24%), of those, 52% (n=15) had anti-synthetase syndrome (ASyS). Three patients presented as a paraneoplastic syndrome. Ninety-nine patients (82%) had a myositis autoantibody identified: antisynthetase autoantibodies were the most commonly identified (n=20, 16%). 62% of the patients were treated with steroids and 35% with ≥2 disease-modifying anti-rheumatic drugs.

Table 1 depicts the main clinical characteristics and the immunologic profile for each diagnosis

Conclusion: In our cohort the most frequent myositis subtype was DM. Almost a quarter of patients had associated ILD, which is an important cause of morbidity and mortality. ILD was more frequent in ASyS patients and was most commonly related to anti-Jo1 antibodies, which is consistent with the literature.

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.3150

AB0446

DEMOGRAPHIC AND CLINICAL FEATURES OF IDIOPATHIC INFLAMMATORY MYOSITIS AND PREVALENCE OF MYOSITIS-RELATED ANTIBODIES

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Background: Idiopathic inflammatory myopathies (IIM) are heterogeneous groups of connective tissue diseases. Diagnosis of IIM is sometimes challenging, but newly defined myositis specific autoantibodies (MSAs) are helpful in diagnosing of IIM and revealing the clinical signs [1].

Objectives: To describe demographic and clinical features of IIMs and to evaluate the frequency and clinical associations of myositis-specific and myositis associated antibodies (MAAs) in IIM.

Methods: Samples obtained from patients tested for MSA/MAA as of 2017 to 2020 were retrospectively evaluated. Patients were diagnosed with IIM according to clinician diagnosis. The prevalence and clinical associations of myositis-related antibodies were assessed with univariate and multivariate analysis.

Results: 152 patients (70.4% female) were tested for myositis-related antibodies during the study period. 81 patients were diagnosed with IIM (38 DM, 43PM). DM patients were diagnosed at an earlier age and its association with malignancy was more relevant, although the relationship with malignancy was not statistically significant. Differences of clinical characteristics between DM and PM patients were skin features which were more frequent in DM, and interstitial lung disease which was more common in PM patients.

MSAs/MAAs were found in 65.4% and 43.2% in IIM patients, more frequently than patients without IIM (p=0.000 and 0.0225, respectively). MSAs were equally frequent in both IIM groups, whereas MAAs were more common in the PM group (although no significant difference in overlap with other connective tissue diseases within these two diseases). The most common MSAs in DM were anti-TIF1-γ (23%), anti-MDA5 (16%), anti-NXP2 (13%) and in PM were anti-Jo-1 (28%), anti-MDA5 (12%), anti-ku (12%), anti-PL7 (12%) respectively. Predictors of ILD in IIM were Anti-Jo-1 positivity and age (RR:10 [CI:2-41] p=0.003 and RR:1.07 [CI:1.02-1.12] p=0.04 respectively)

Conclusion: The general characteristics of DM and PM patients and the frequencies of myositis-specific autoantibodies were similar to the literature, except that anti-TIF1-γ antibodies (14%) were more common in our Turkish cohort [2].

REFERENCES:

- [1] Cruellas, M.G., et al., *Myositis-specific and myositis-associated autoantibody profiles and their clinical associations in a large series of patients with polymyositis and dermatomyositis*. Clinics (Sao Paulo), 2013. **68**(7): p. 909-14.
- [2] Gonzalez-Bello, Y., et al., *Myositis-Specific Antibodies and Myositis-Associated Antibodies in Patients With Idiopathic Inflammatory Myopathies From the PANLAR Myositis Study Group*. J Clin Rheumatol, 2020.

Table 1. Baseline characteristics of the patients with DM and PM

	DM (n=38)	PM (n=43)	P
Female, n (%)	27 (71.1)	29 (67.4)	0.457
Age at onset, years	33 (15-48)	47 (38-58)	0.002
Skeletal muscle features			
Muscle weakness, n (%)	34 (89.5)	37 (86.0)	0.451
Myalgia, n (%)	23 (60.5)	30 (69.8)	0.261
Arthritis, n (%)	8 (21.1)	9 (20.9)	0.601
Skin features, n (%)			
Heliotope rash, n (%)	18 (47.4)	0 (0)	<0.001
Gotttron papules, n (%)	25 (65.8)	0 (0)	<0.001
Gotttron sign, n (%)	18 (47.4)	0 (0)	<0.001
Generalize erythema, n (%)	10 (26.3)	0 (0)	<0.001
Calcinosis, n (%)	4 (10.5)	1 (2.3)	0.143
Dysphagia, n (%)	10 (26.3)	12 (27.9)	0.537
Interstitial lung disease (ILD), n (%)	8 (21.1)	20 (47.6)	0.012
Other connective tissue diseases overlap, n (%)	6 (15.8)	13 (30.2)	0.102
Malignancies, n (%)	5 (13.5)	3 (7)	0.275
Laboratory data			
Creatine kinase (IU/L)	505 (110-3616)	801 (402-1868)	0.655
C-reactive protein (mg/dl)	0.3 (0.1-0.8)	1.2 (0.5-2.8)	<0.001
ESR (mm/h)	19 (8-27)	25 (17-48)	0.005
MAAs (Ro-52, pm-scl 100, pm-scl75), n (%)	11 (28.9)	24 (55.8)	0.013
MSAs, n (%)	28 (73.7)	25 (58.1)	0.108
Anti-TIF1-γ, n (%)	9 (23.7)	2 (4.7)	0.014
Anti-Jo-1, n (%)	2 (5.3)	12 (27.9)	0.007
Anti-PL-7, n (%)	0	5 (11.6)	0.038

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.3349

AB0447

PREVALENCE OF PRIMARY BILIARY CIRRHOSIS IN SYSTEMIC SCLEROSIS AND SJÖGREN'S SYNDROME OVER TIME: A SYSTEMATIC REVIEW

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Background: Primary biliary cirrhosis (PBC) is a rare slowly progressive autoimmune disease characterized by inflammatory destruction and fibrosis of intra-hepatic bile ducts. It is known to coexist together with rheumatological conditions such as Sjögren's syndrome (SS) and systemic sclerosis (SSc). There is a wide range in reported prevalence of disease overlap with these entities; however, the exact prevalence rates remain unclear.

Objectives: The objectives were to determine the prevalence of: 1) PBC in patients with SS and SSc (and the subsets of limited cutaneous SSc (lcSSc) and diffuse cutaneous SSc (dcSSc)), 2) SSc and SS in patients with PBC, and 3) to analyze changes in frequency over time. SSc occurs in 3/10,000 and PBC in 4-40/10,000 but these rare autoimmune diseases are known to coexist together. We speculated that there could be more cases diagnosed due to increasing availability of standardized antibody tests such as ANA, centromere antibodies, ENA and mitochondrial antibodies.

Methods: A systematic review of the literature was performed using Medline, EMBASE, CINAHL, and the Cochrane Library databases up till June 16, 2020. Only full text articles in the English language with at least 40 patients were included. Cohorts, case series, cross-sectional studies, correspondences and registries with reported prevalence rates of both PBC in patients with SS and SSc as well as SSc and SS in patients with PBC were included. Data on frequency of co-existent diseases was studied by year of publication to determine if prevalence changed over time using linear regression. We used the Strengthening of Reporting of Observational Studies in Epidemiology (STROBE) checklist to assess the quality of the studies.

Results: Of 2876 citations identified, 67 were included in the analysis (n=33 for PBC, 15 for SSc, 18 for SS and 1 for SSc/SS). STROBE checklist scores ranged from 7-21. The prevalence of PBC was 5% in patients with SSc. Within the subsets, the prevalence of PBC in lcSSc was 8% and in dcSSc was 1%. In patients with SS, the prevalence of PBC was 4%. The prevalence of SSc overall in those with PBC was 5% and, within the subsets was 6% in lcSSc and 0% in dcSSc. The prevalence of SS in PBC was 18%. There was also no significant association between year of publication and prevalence. There was a lack of standardized definitions so misclassification may have occurred.

Conclusion: PBC is increased in SSc but mostly in the lcSSc subset. SS in PBC is common at nearly 1 in 5. Over the years, there was no change in the prevalence of PBC in SSc indicating stability over time.

Acknowledgements: Meagan Stanley, Western University Librarian.

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.3433

AB0448

SYSTEMIC SCLEROSIS ASSOCIATED PULMONARY ARTERIAL HYPERTENSION: PREDOMINANCE OF PULMONARY FIBROSIS AS A RISK FACTOR FOR MORTALITY IN A SINGLE CENTER COHORT

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Background: Pulmonary arterial hypertension (PAH) is a severe vasculopathic complication for systemic sclerosis (SSc) patients. The availability of oral-specific vasodilator therapies has provided better outcomes.

Objectives: We aimed to analyze the characteristics of SSc-PAH patients and factors associated with mortality.

Methods: Medical records of 291 SSc patients fulfilling ACR/EULAR criteria (2013) and followed-up during 2008-2020 years were screened and the patients who diagnosed PAH evaluated by right heart catheterization (mean PAB≥20 mmHg) (n=26, 8.9%) were included into this retrospective cross-sectional study.

Results: The characteristics of 26 SSc-PAH(24 females) patients were summarised in Table 1.

Table 1. Characteristics of SSc patients with PAH

	SSc-PAH (n=26)
Demographics	
Age(yrs)	53.6±8.9
Duration of Raynaud's(yrs)	16.1±11.8
Duration of Non-Raynaud's(yrs)	9.2±6.6
Raynaud's to Diagnosis of PAH(yrs)	10.8±6.7
Clinical Characteristics (%)	
LcSSc	9 (34.6)
DcSSc	18 (65.4)
Digital ulcer	14 (53.8)
Gastrointestinal	20 (69.0)
Synovitis	5 (19.2)
Flex contractures	7 (26.9)
Tendon friction rubs	3 (11.5)
Renal crisis	1 (3.8)
Pulmonary fibrosis	19 (73.1)
Auto-antibodies (%)	
ANA	23 (88.5)
Anti-centromere	6 (23.1)
Anti-Scl70	13 (50)
Treatment (%)	
Specific vasodilator	
ERA (bosentan/macitentan/ambicentan)	15 (51.7)
PDE5-i (sildenafil/tadalafil)	17 (58.6)
Prostacyclin-analog (Iloprost/treprostenil/selexipag)	15 (51.7)
Riociguat	5 (17.2)
Immunosuppressives	21 (80.8)
Steroids	16 (61.5)

Twenty-four (92.3%) of the SSc-PAH patients had PAH-related symptoms at the time of diagnosis, 2 (7.7%) were asymptomatic and diagnosed by screening. RHC and treatment details were stated in Table 2.

Table 2. Right Heart Catheterization (RHC) ve treatment details of SSc-PAH patients

	n=26
Initial RHC	
-mean PAB	30.4±7.9 (median 28, 20-53mmHg)
-PVR	5.1±2.4 (median 4, 3-9 woods)
-PCWP	10.8±5.5 (median 10, 0-15 mmHg)
Initial treatment	
-monotherapy	5 (19.2%)
-combination	10 (38.5%)
-add-on combination	12 (46.2%)

Eleven out of 26 patients (42.3 %) were deceased after a mean follow up of 43.7±24.6 (median 48.1-84) from PAH diagnosis and 15.1±9.9 (median 13, 0.6-34) years after SSc diagnosis. Deceased patients were younger and had younger age at disease onset (49.1±8.8 vs 56.9±7.7, p=0.032 and 30.8±13.0 vs 42.0±10.9, 0.027). All deceased patients had associated pulmonary fibrosis (100 vs 53.3%, p=0.01). No significant difference was observed for initial RHC parameters between deceased and survived SSc-PAH patients. Specific monotherapy was found to be more frequent in deceased patients (45.5 vs 0%, p=0.007).