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 (%)			
Heliotrope 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

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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.

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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).