

Supplementary Table S1. Characterisation of subdomains according to SSDDI and SSDI scores.

Damage	Number of patients affected (n; %)	Score	
		Mean $\pm$ SD score	Time to event min; max
Ocular domain			
Corneal scarring	2; 1.0	0.1 $\pm$ 1.3	0; 18
Corneal ulcers	43; 20.7	1.7 $\pm$ 4.5	0; 30
Schirmer result 0mm/5min in both eyes	21; 10.1	0.4 $\pm$ 2.1	0; 22
Schirmer result 5mm/5min in both eyes	47; 22.6	0.7 $\pm$ 2.4	0; 22
Tear duct surgery		0	0
Cataract	49; 23.6	2.8 $\pm$ 6.5	0; 34
Retinal change	25; 12.0	1.3 $\pm$ 4.4	0; 25
Chronic blepharitis	35; 16.8	1.4 $\pm$ 4.5	0; 28
Oral domain			
Caries	32; 15.4	1.3 $\pm$ 4.4	0; 40
Teeth loss	62; 29.8	2.8 $\pm$ 5.9	0; 39
Salivary gland swelling	84; 40.4	1.7 $\pm$ 4.0	0; 31
Unstimulated salivary flow <1.5mL/15min	14; 6.7	0.4 $\pm$ 2.2	0; 22
Unstimulated salivary flow 0mL/15min	4; 1.9	0.2 $\pm$ 1.9	0; 22
Oral infection	56; 26.9	2.1 $\pm$ 5.1	0; 36
Parotid surgery	5; 2.4	0.1 $\pm$ 0.6	0; 8
Gum disease	39; 18.8	1.6 $\pm$ 4.5	0; 31
Oral ulceration	50; 24.0	1.1 $\pm$ 2.9	0; 18
Dysphonia	14; 6.7	0.3 $\pm$ 1.6	0; 14
Neurological domain			
CNS involvement	25; 12.0	1.3 $\pm$ 4.9	0; 32
Cranial neuropathy	18; 8.7	1.0 $\pm$ 4.3	0; 32
Peripheral neuropathy (SSDDI)	66; 31.7	2.5 $\pm$ 5.8	0; 39
Peripheral neuropathy (SSDI)	64; 30.8	2.5 $\pm$ 5.8	0; 39
Other CNS involvement	13; 6.3	1.3 $\pm$ 3.9	0; 26
Mononeuritis multiplex	3; 1.4	0.1 $\pm$ 0.5	0; 6
Cognitive impairment	13; 6.3	0.7 $\pm$ 3.5	0; 26
Renal			
Nephrocalcinosis	11; 5.3	0.5 $\pm$ 2.6	0; 21
Renal tubular acidosis	4; 1.9	0.1 $\pm$ 0.7	0; 8
Increased serum creatinine level or reduced GFR	22; 10.6	1.1 $\pm$ 3.7	0; 24
Glomerular filtration rate <50% predicted	20; 9.6	1.0 $\pm$ 3.6	0; 24
Proteinuria >3.5g/24h	0	0	0
End stage renal disease	0	0	0
Chronic cystitis	1; 0.5	0.0 $\pm$ 0.6	0; 9
Pulmonary domain			
Significant irreversible functional damage (confirmed by spirometry)	5; 2.4	0.1 $\pm$ 0.9	0; 8
Pleural fibrosis	1; 0.5	0.0 $\pm$ 0.5	0; 7
Pulmonary fibrosis	12; 5.8	0.5 $\pm$ 2.8	0; 21
Pulmonary hypertension	5; 2.4	0.2 $\pm$ 1.3	0; 13
Pulmonary infarction	0	0	0
Cardiovascular domain			
Cardiomyopathy	14; 6.7	1.0 $\pm$ 4.1	0; 28
Hypertension	38; 18.3	2.2 $\pm$ 6.0	0; 37
Ischaemic heart disease	11; 5.3	0.9 $\pm$ 4.3	0; 39
Heart valve disease	13; 6.3	0.7 $\pm$ 3.1	0; 21
Pericarditis	1; 0.5	0.0 $\pm$ 0.1	0; 1
Myocardial infarction	1; 0.5	0.1 $\pm$ 1.5	0; 22
Gastrointestinal domain			
Chronic pancreatitis	0	0	0
Celiac disease	3; 1.4	0.1 $\pm$ 0.9	0; 10
Primary biliary cirrhosis	2; 1.0	0.0 $\pm$ 0.3	0; 3
Primary sclerosing cholangitis	0	0	0
Chronic autoimmune hepatitis	6; 2.9	0.2 $\pm$ 2.0	0; 26
Upper gastrointestinal surgery	1; 0.5	0.2 $\pm$ 0.3	0; 4

Damage	Number of patients affected (n; %)	Score	
		Mean $\pm$ SD score	Time to event min; max
Musculoskeletal			
Erosive arthropathy	14; 6.7	0.6 $\pm$ 2.7	0; 21
Osteoarthritis	111; 53.4	3.2 $\pm$ 5.6	0; 35
Osteoporosis	39; 18.8	2.4 $\pm$ 6.3	0; 33
Avascular necrosis	4; 1.9	0.2 $\pm$ 2.1	0; 21
Skin ulcers	15; 7.2	0.9 $\pm$ 3.7	0; 26
Endocrine			
Hypothyroidism	21; 10.1	0.9 $\pm$ 3.2	0; 26
Pernicious anaemia	3; 1.4	0.1 $\pm$ 1.3	0; 18
Hyperthyroidism	2; 1.0	0.1 $\pm$ 0.8	0; 10
Diabetes	4; 1.9	0.2 $\pm$ 1.8	0; 18
Malignancy			
Paraproteinaemia	21; 10.1	1.2 $\pm$ 4.5	0; 25
Other malignancy	23; 11.1	1.1 $\pm$ 4.3	0; 37
Macroglobulinaemia	1; 0.5	0 $\pm$ 0.1	0; 1
Cryoglobulinaemia	4; 1.9	0.3 $\pm$ 2.1	0; 18
Multiple myeloma	0	0	0
Lymphoma	17; 8.2	0.6 $\pm$ 2.8	0; 23

Continuous values shown in mean $\pm$ standard deviation or median and interquartile range if not normally distributed. Proportions reported in absolute number and percentage.

Supplementary Table S2. Glandular and extra-glandular domains using SSDDI and SSDI scores.

Scores	First year			Final		
	Glandular	Extra-glandular	<i>p</i> -value	Glandular	Extra-glandular	<i>p</i> -value
SSDDI score	45 (21.6)	31 (14.9)	0.08	138 (66.4)	108 (51.9)	0.003
SSDI score	56 (26.9)	33 (15.9)	0.006	131 (63.0)	126 (60.6)	0.61
P value	0.2	0.8	-	0.5	0.1	-

Proportions reported in absolute number and percentage.

Supplementary Table S3. Comparison of characteristics of deceased and alive patients.

	Dead (n=32, 18.0)	Alive (n=146, 82.0)	p-value
Female	29 (90.6)	141 (96.6)	0.14
Ethnicity			NA
White	27 (87.1)	81 (57.0)	
African-American	0	16 (11.3)	
Afro-Caribbean	0	5 (3.5)	
Asian	3 (9.7)	23 (16.2)	
Mixed	0	4 (2.8)	
Other	1 (3.2)	13 (9.2)	
Age at diagnosis	59.2 ± 11.1	45.8 ± 13.9	< 0.001
Age at symptoms onset	53.2 ± 10.2	40.9 ± 13.9	< 0.001
Antibodies profile – positivity			
ANA	19 (76.0)	121 (89.0)	0.10
Anti-SSA/Ro	12 (48.0)	118 (84.9)	<0.001
Anti-SSB/La	10 (40.0)	64 (46.0)	0.67
Anti-RNP	0	7 (5.1)	NA
Rheumatoid factor	12 (63.2)	12 (63.2)	0.59
Treatment (ever)			
Hydroxychloroquine	18 (58.1)	104 (71.2)	0.20
Methotrexate	1 (3.2)	10 (6.9)	0.69
Azathioprine	1 (3.2)	5 (3.4)	0.96
Sulfasalazine	0	7 (4.8)	NA
Mycophenolate of mofetil	1 (3.1)	8 (5.5)	0.59
Cyclophosphamide	3 (9.7)	5 (3.4)	0.13
Rituximab	1 (3.1)	14 (9.6)	0.31
Steroids (any dose)	12 (38.7)	59 (40.4)	0.86
High dose of steroids (>6mg)	13 (41.9)	50 (36.2)	0.55
Follow-up period (years)	21.5 (IQR 13-28)	11 (IQR 7-19)	0.003
Lymphoma	5 (15.6)	10 (6.9)	0.15

Continuous values shown in mean ± standard deviation or median and interquartile range if not normally distributed. Proportions reported in absolute number and percentage. Thirty patients were excluded due to missing data on mortality.

Supplementary Table S4. Glandular and extra-glandular domains using SSDDI and SSDI scores.

Damage	SSDDI score			SSDI score		
	Lymphoma (n = 17, 8.2)	No lymphoma (n = 191, 91.8)	p-value	Lymphoma (n = 17, 8.2)	No lymphoma (n = 191, 91.8)	p-value
Early glandular damage	2 (11.8)	43 (22.5)	0.54	4 (23.5)	52 (27.2)	0.74
Early extra-glandular damage	5 (29.4)	26 (13.6)	0.08	5 (29.4)	28 (14.7)	0.11
Final glandular damage	11 (64.7)	127 (66.5)	0.88	11 (64.7)	120 (62.8)	0.88
Final extra-glandular damage	17 (100)	91 (47.6)	NA	17 (100)	109 (57.1)	NA
Time to glandular damage	7.6 ± 7.2	5.6 ± 6.6	0.33	8.5 ± 12.4	5.0 ± 6.6	0.13
Time to extra-glandular damage	6.8 ± 7.0	6.9 ± 7.5	0.96	5.8 ± 5.5	6.4 ± 6.2	0.68

Continuous values shown in mean ± standard deviation or median and interquartile range if not normally distributed. Proportions reported in absolute number and percentage.