

Supplementary Table S1. Demographical, clinical, laboratory features (baseline) and therapies (ever) of SSc patients: comparison between 253 patients included in this study and 395 patients excluded from this study.

Continuous variables are presented as median (IQR) and compared with Mann-Whitney test; categorical variables are presented as number/number available data (%) and compared with Chi-square test/Fisher's exact test. *P*-value <0.05 was considered statistically significant.

Characteristics	SSc patients n=648	SSc patients included in this study (≥10 years follow-up) n=253	SSc patients excluded from this study (<10 years follow-up/missing data) n=395	<i>p</i> -value
Female sex	584/648 (90%)	237/253 (94%)	347/395 (88%)	0.015
Caucasian ethnicity	638/648 (98%)	251/253 (99%)	387/395 (98%)	0.312
Age* (years)	55.5 (45.1-66.6)	52.4 (43.3-60.3)	59.1 (46.8-68.6)	<0.001
Ever smokers	191/622 (31%)	70/247 (28%)	121/375 (32%)	0.299
Active smokers*	109/622 (18%)	42/247 (17%)	67/375 (18%)	0.782
Age-adjusted-CCI*	1 (0-3)	1 (0-2)	2 (0-3)	<0.001
Age-adjusted-CCI (T10)	2 (1-3)	2 (1-3)	-	-
Anti-topoisomerase I	141/644 (22%)	51/253 (20%)	90/391 (23%)	0.391
Anti-centromere	333/644 (52%)	139/253 (55%)	194/391 (50%)	0.187
Anti-RNA polymerase III	31/644 (5%)	11/253 (4%)	20/391 (5%)	0.657
Other auto-antibodies	132/644 (20%)	50/253 (20%)	82/391 (21%)	0.710
Diffuse cutaneous subset*	122/648 (19%)	38/253 (15%)	84/395 (21%)	0.047
Interstitial lung disease*	75/648 (12%)	24/253 (9%)	51/395 (13%)	0.184
Pulmonary arterial hypertension*	19/648 (3%)	1/253 (0.4%)	18/395 (4.5%)	0.002
Scleroderma renal crisis*	8/648 (1.2%)	1/253 (0.4%)	7/395 (1.8%)	0.158
Digital ulcers*	88/648 (14%)	28/253 (11%)	60/395 (15%)	0.135
Corticosteroids use (ever)	276/530 (52%)	102/222 (46%)	174/308 (56%)	0.016
Prostacyclin analogues use (ever)	162/540 (30%)	59/218 (27%)	103/322 (32%)	0.221
Immunosuppressive therapy use (ever)	70/540 (13%)	28/218 (13%)	42/322 (13%)	0.946

*at diagnosis.

CCI: Charlson Comorbidity Index; SSc: systemic sclerosis.

Supplementary Table S2. Demographical, clinical, laboratory features (baseline) and therapies (ever) of SSc patients: males vs. females. Continuous variables are presented as median (IQR) and compared with Mann-Whitney test; categorical variables are presented as number/number available data (%) and compared with Chi-square test/Fisher's exact test. *P*-value <0.05 was considered statistically significant.

Characteristics	SSc PATIENTS n=648	SSc MALE PATIENTS n=64	SSc FEMALE PATIENTS n=584	<i>p</i> -value
Caucasian ethnicity	638/648 (98%)	61/64 (95%)	577/584 (99%)	0.067
Age* (years)	55.5 (45.1-66.6)	56.1 (45.6-67.1)	55.5 (44.8-65.3)	0.632
Follow-up duration (years)	10.1 (4.9-16.8)	5.8 (2.7-11.0)	10.9 (5.2-17.0)	0.001
Lost to follow-up	111/648 (17%)	12/64 (19%)	99/584 (17%)	0.727
Deaths	240/537 (45%)	34/52 (65%)	206/485 (42%)	0.002
Age at death (years)	76.1 (67.2-81.4)	70.1 (61.1-78.1)	76.5 (67.8-82.4)	0.016
Disease duration at death (years)	12.9 (7.3-20.4)	7.2 (3.8-11.6)	14.6 (8.8-20.8)	<0.001
Ever smokers	191/622 (31%)	41/61 (67%)	150/561 (27%)	<0.001
Active smokers*	109/622 (18%)	19/61 (31%)	90/561 (16%)	0.004
Age-adjusted-CCI*	1 (0-3)	2 (1-3)	1 (0-3)	0.008
Age-adjusted-CCI (T10)	2 (1-3)	1 (1-3)	2 (1-3)	0.242
Anti-Topoisomerase I	141/644 (22%)	20/63 (32%)	121/581 (21%)	0.047
Anti-centromere	333/644 (52%)	9/63 (14%)	324/581 (56%)	<0.001
Anti-RNA Polymerase III	31/644 (5%)	6/63 (10%)	25/581 (4%)	0.110
Other auto-antibodies	132/644 (20%)	25/63 (40%)	107/581 (18%)	<0.001
Diffuse cutaneous subset*	122/648 (19%)	25/64 (39%)	97/584 (17%)	<0.001
Interstitial lung disease*	75/648 (12%)	17/64 (27%)	58/584 (10%)	<0.001
Pulmonary arterial hypertension*	19/648 (3%)	2/64 (3%)	17/584 (3%)	1.000
Scleroderma renal crisis*	8/648 (1.2%)	3/64 (5%)	5/584 (1%)	0.036
Digital ulcers*	88/648 (14%)	11/64 (17%)	77/584 (13%)	0.375
Corticosteroids use ever	276/530 (52%)	40/53 (75%)	236/477 (49%)	<0.001
Prostacyclin analogues use ever	162/540 (30%)	21/55 (38%)	141/485 (29%)	0.162
Immunosuppressive therapy use ever	70/540 (13%)	8/55 (15%)	62/485 (13%)	0.712

*at diagnosis.

CCI: Charlson Comorbidity Index; SSc: systemic sclerosis.

Supplementary Table S3. Demographical and clinical features, causes of mortality and SCTC-DI values (T0, T1, T5, T10): patients with early vs. late mortality.

Continuous variables are presented as median (IQR) and compared with Mann-Whitney test; categorical variables are presented as number/number available data (%) and compared with Chi-square test/Fisher's exact test. *P*-value <0.05 was considered statistically significant.

	Patients with early mortality n=133	Patients with late mortality n=90	<i>p</i> -value
Characteristics			
Female sex	111/133 (84%)	83/90 (92%)	0.056
Caucasian ethnicity	130/133 (98%)	90/90 (100%)	0.275
Age* (years)	66.5 (58.9-73.2)	57.6 (50.2-63.2)	<0.001
Age-adjusted-CCI*	3 (2-4)	2 (1-2)	<0.001
Anti-topoisomerase I	31/131 (27%)	28/90 (31%)	0.219
Anti-centromere	63/131 (48%)	37/90 (41%)	0.306
Anti-RNA Polymerase III	5/131 (4%)	2/90 (2%)	0.703
Other auto-antibodies	31/131 (24%)	21/90 (23%)	0.955
Diffuse cutaneous subset*	28/133 (21%)	18/90 (20%)	0.849
Interstitial lung disease*	28/133 (21%)	15/90 (17%)	0.415
Pulmonary arterial hypertension*	15/133 (11%)	6/90 (7%)	0.247
Scleroderma renal crisis*	2/133 (1.5%)	1/90 (1%)	1.000
Digital ulcers*	38/133 (29%)	18/90 (20%)	0.148
Causes of mortality			
SSc-related deaths	61/133 (46%)	30/90 (33%)	0.062
Interstitial lung disease	12/61 (20%)	15/30 (50%)	0.003
Pulmonary arterial hypertension	26/61 (43%)	6/30 (20%)	0.034
Gastrointestinal involvement	7/61 (11%)	4/30 (13%)	1.000
Cardiovascular involvement	8/61 (13%)	4/30 (13%)	1.000
Scleroderma renal crisis	7/61 (11%)	1/30 (3%)	0.263
Other	1/61 (1.6%)	0/30 (0%)	1.000
No SSc-related causes of death	43/133 (32%)	44/90 (49%)	0.013
Cancer	17/43 (39.5)	18/44 (40.9)	0.896
Infection	2/43 (4.7)	4/44 (9.1)	0.676
Cardiovascular	11/43 (25.6)	6/44 (13.6)	0.160
Other	7/43 (16.3)	10/44 (22.7)	0.448
Undetermined causes of death	29/133 (22%)	16/90 (18%)	0.462
SCTC-DI values			
SCTC-DI (T0)	3 (2-6)	3 (1-4)	0.052
SCTC-DI (T1)	4 (2-8)	3 (1-4)	0.006
SCTC-DI (T5)	6 (4-11)	4 (2-6)	<0.001
SCTC-DI (T10)	-	6 (3-8)	-

*at diagnosis.

DI: damage index; SCTC: Scleroderma Clinical Trial Consortium; SSc: systemic sclerosis.