Supplementary Table S1. Demographic and clinical characteristics of 251 diffuse cutaneous SSc patients, according to the SSc pattern by nailfold videocapillaroscopy.

Demographic and clinical characteristics n (%) [a]	Late pattern 141 (56%)	Non-Late pattern 110 (44%)	Univariate analysis OR (95% CI)	*p*
Age at the first symptom, mean ± SD, years	44.8 ± 15.9	43.6 ± 14.0	-	0.545
Age at SSc diagnosis, mean ± SD, years	48.3 ± 15.7	47.9 ± 14.1	-	0.864
Time from first symptom to SSc diagnosis, mean ± SD, years	3.5 ± 7.8	4.5 ± 6.0	-	0.265
Female gender, n (%)	116 (82%)	89 (81%)	1.09 (0.58-2.08)	0.870
Peripheral vascular manifestations (n = 251)				
Raynaud's phenomenon as first symptom, n (%)	92 (68%)	80 (74%)	0.73 (0.42-1.28)	0.323
Raynaud's phenomenon, n (%)	132 (94%)	108 (98%)	0.27 (0.06-1.28)	0.119
Telangiectasia, n (%)	91 (65%)	72 (65%)	0.96 (0.57-1.62)	0.895
Acro-osteolysis, n (%)	41 (29%)	18 (17%)	2.12 (1.13-3.94)	0.024
Digital ulcers, n (%)	91 (65%)	72 (65%)	0.96 (0.57-1.62)	0.895
Digestive involvement, n (%) (n = 249)				
Oesophageal, n (%)	92 (65%)	77 (71%)	0.76 (0.44-1.30)	0.340
Gastric and/or intestinal, n (%)	31 (22%)	31 (29%)	0.70 (0.39-1.25)	0.240
Liver, n (%)	4 (2.8%)	4 (3.7%)	0.76 (0.19-3.11)	0.730
Pulmonary involvement:				
Interstitial lung disease, n (%) (n = 251)	98 (70%)	76 (69%)	1.02 (0.59-1.75)	1.000
FVC <70%, n (%) (n = 239)	59 (43%)	32 (31%)	1.70 (0.99-2.91)	0.060
DLCO/VA (% of expected), mean ± SD, (n = 221)	80.6 ± 57.5	73.4 ± 32.3		0.269
Pulmonary hypertension, n (%) (n = 218)	38 (31%)	27 (29%)	1.10 (0.61-1.97)	0.768
Pulmonary arterial hypertension, n (%) (n = 221)	3 (2.4%)	3 (3.1%)	0.76 (0.15-3.86)	1.000
sPAP >40 mm Hg, n (%) (n = 231)	68 (52%)	35 (35%)	2.07 (1.21-3.53)	0.008
Heart involvement				
Pericarditis, n (%) (n = 151)	14 (16%)	4 (6.5%)	2.71 (0.85-8.66)	0.124
Ischaemic cardiopathy, n (%) (n = 151)	12 (13%)	9 (15%)	0.92 (0.36-2.33)	1.000
Conduction alteration, n (%) (n = 150)	28 (31%)	16 (26%)	1.29 (0.63-2.67)	0.585
Left ventricle diastolic dysfunction, n (%) (n = 206)	21 (18%)	21 (23%)	0.74 (0.38-1.47)	0.486
Scleroderma renal crisis, n (%) (n = 250)	16 (11%)	3 (2.8%)	4.52 (1.28-15.94)	0.014
Osteomuscular involvement				
Arthritis, n (%) (N = 206)	26 (22%)	27 (31%)	0.62 (0.33-1.17)	0.149
Myositis, n (%) (N = 207)	29 (24%)	21 (24%)	1.03 (0.54-1.96)	1.000
Tendon friction rubs, n (%) (n = 205)	23 (19%)	16 (19%)	1.05 (0.52-2.13)	1.000
Calcinosis, n (%) (n = 204)	16 (14%)	6 (7.0%)	2.09 (0.78-5.59)	0.172
Flexion contractures, n (%) (n = 133)	41 (57%)	22 (36%)	2.34 (1.16-4.72)	0.023
Peripheral nervous system involvement, n (%) (n = 204)	15 (14%)	10 (10%)	1.39 (0.59-3.25)	0.524
Malignancy, n (%) (n = 249)	14 (10%)	20 (18%)	0.49 (0.24-1.03)	0.064

DLCO/VA: diffusing capacity for carbon monoxide corrected by alveolar volume; FVC: forced vital capacity; OR (95% CI), odds ratio, and 2-sided 95% confidence interval of the mean; sPAP: estimated systolic pulmonary artery pressure; SSc: systemic sclerosis.
[a]All data derived from 251 patients except when indicated (n =).

Supplementary Table S2. Immunological features, causes of death and survival of 251 diffuse cutaneous SSc patients according to the SSc pattern by nailfold videocapillaroscopy.

Autoantibodies, causes of death, and survival, n (%) [a]	Late pattern 141 (56%)	Non-Late pattern 110 (44%)	Univariate analysis OR (95% CI)	*p*
Autoantibodies, n (%)				
Anti-nuclear antibodies (n = 251)	133 (94%)	105 (95%)	0.79 (0.25-2.49)	0.780
Anti-centromere antibodies (n = 229)	14 (11%)	8 (8.1%)	1.37 (0.55-3.41)	0.652
Anti-Topoisomerase I (n = 238)	65 (50%)	56 (52%)	0.93 (0.56-1.55)	0.796
Anti-RNA polymerase III (n = 63)	8 (24%)	10 (33%)	0.64 (0.21-1.92)	0.578
Anti-PM-Scl (n = 151)	8 (9.3%)	7 (11%)	0.85 (0.29-2.48)	0.789
Death from all causes, n (%) (n = 71)	46 (33%)	25 (23%)	1.65 (0.93-2.91)	0.092
Causes of death, n (%)				
Interstitial lung disease	11 (24%)	1 (4.0%)	7.54 (0.91-62.34)	0.046
Pulmonary arterial hypertension	6 (13%)	1 (4.0%)	3.60 (0.41-31.74)	0.409
ILD related pulmonary hypertension	5 (11%)	4 (16%)	0.64 (0.16-2.64)	0.711
Scleroderma renal crisis	7 (15%)	1 (4.0%)	4.31 (0.50-37.21)	0.246
Malignancy	2 (4.3%)	10 (40%)	0.07 (0.01-0.35)	<0.001
Ischaemic cardiopathy	1 (2.2%)	0 (0.0%)	-	1.000
Stroke	0 (0.0%)	0 (0.0%)	-	-
Chronic renal failure	0 (0.0%)	0 (0.0%)	-	-
Sepsis	2 (4.3%)	1 (4.0%)	1.09 (0.09-12.66)	1.000
Other causes	8 (17%)	5 (20%)	0.84 (0.24-2.91)	0.760
Not specified	1 (2.2%)	1 (4.0%)	0.53 (0.03-8.91)	1.000
Scleroderma-related causes of death, n (%) (n = 69)	33 (73%)	9 (38%)	4.58 (1.59-13.20)	0.005
Median survival time since first SSc symptom, y (IQR)	24.1 (16.1-32.2)	28.6 (24.5-32.7)	-	0.049
Cumulative survival rates since disease onset				
At 5 years	0.879	0.990	-	0.001
At 10 years	0.791	0.944	-	0.001
At 20 years	0.564	0.739	-	0.006
At 30 years	0.410	0.322	-	0.026

ILD: interstitial lung disease; IQR: interquartile range; SSc: systemic sclerosis.
[a]All data derived from 251 patients except when indicated (n =).

Supplementary Table S3. Demographic and clinical characteristics of 987 limited cutaneous SSc patients, according to the SSc pattern by nailfold videocapillaroscopy.

Demographic and clinical characteristics, n (%) [a]	Late pattern 367 (37.2%)	Non- Late pattern 620 (62.8%)	Univariate analysis OR (95% CI)	*p*
Age at the first SSc symptom, mean ± SD, y	47.1 ± 16.8	46.6 ± 16.2	-	0.672
Age at SSc diagnosis, mean ± SD, y	54.9 ± 16.1	54.6 ± 14.5	-	0.767
Time from first symptom to SSc diagnosis, mean ± SD, y	8.2 ± 10.6	8.2 ± 10.9	-	0.975
Female gender, n (%)	325 (89%)	556 (90%)	0.89 (0.59-1.35	0.596
Peripheral vascular manifestations				
Raynaud's phenomenon as first symptom, n (%) (n = 974)	308 (86%)	524 (85%)	1.22 (0.77-1.62)	0.638
Raynaud's phenomenon, n (%) (n = 986)	357 (97%)	597 (96%)	1.32 (0.62-2.81)	0.579
Telangiectasia, n (%) (n = 982)	242 (66%)	384 (62%)	1.19 (0.91-1.57)	0.216
Acro-osteolysis, n (%) (n = 623)	26 (12%)	14 (3.4%)	3.84 (1.96-7.53)	<0.001
Digital ulcers, n (%) (n = 985)	146 (40%)	213 (34%)	1.26 (0.96-1.64)	0.100
Digestive involvement				
Oesophageal, n (%) (n = 979)	203 (56%)	383 (62%)	0.76 (0.59-0.99)	0.050
Gastric and/or intestinal, n (%) (n = 979)	69 (19%)	133 (22%)	0.85 (0.61-1.17)	0.328
Liver, n (%) (n = 984)	38 (10%)	63 (10%)	1.03 (0.67-1.57)	0.914
Pulmonary involvement				
Interstitial lung disease, n (%) (n = 983)	148 (41%)	215 (35%)	1.29 (0.99-1.68)	0.065
FVC <70%, n (%) (n = 874)	41 (13%)	78 (14%)	0.88 (0.59-1.32)	0.610
DLCO/VA <70%, n (%) (n = 798)	116 (40%)	197 (39%)	1.02 (0.76-1.38)	0.881
Pulmonary hypertension, n (%) (n = 795)	75 (26%)	127 (25%)	1.01 (0.72-1.40)	1.000
Pulmonary arterial hypertension, n (%) (n = 800)	23 (7.8%)	56 (11%)	0.68 (0.41-1.13)	0.142
sPAP >40 mm Hg, n (%) (n = 888)	140 (42%)	236 (43%)	0.98 (0.74-1.29)	0.944
Heart involvement				
Pericarditis, n (%) (n = 431)	21 (13%)	18 (6.7%)	2.03 (1.05-3.94)	0.038
Ischaemic cardiopathy, n (%) (n = 435)	21 (13%)	51 (19%)	0.62 (0.36-1.08)	0.110
Conduction alteration, n (%) (n = 435)	37 (22%)	86 (32%)	0.62 (0.40-0.97)	0.037
Left ventricle diastolic dysfunction, n (%) (n = 837)	89 (29%)	137 (26%)	1.17 (0.86-1.60)	0.333
Scleroderma renal crisis, n (%) (n = 985)	4 (1.1%)	3 (0.48%)	2.27 (0.50-10.19)	0.434
Osteomuscular involvement				
Arthritis, n (%) (n = 625)	45 (21%)	82 (20%)	1.04 (0.69-1.56)	0.917
Myositis, n (%) (n = 625)	15 (6.9%)	46 (11%)	0.58 (0.32-1.07)	0.090
Tendon friction rubs, n (%) (n = 625)	7 (3.2%)	7 (1.7%)	1.92 (0.66-5.54)	0.259
Calcinosis, n (%) (n = 984)	92 (25%)	125 (20%)	1.32 (0.97-1.80)	0.080
Flexion contractures, n (%) (n = 388)	22 (19%)	22 (8.1%)	2.58 (1.37-4.88)	0.005
Peripheral nervous system involvement, n (%) (n = 893)	16 (5.1%)	54 (9.4%)	0.52 (0.29-0.92)	0.026
Malignancy, n (%) (n = 985)	41 (11%)	70 (11%)	0.99 (0.66-1.49)	1.000

DLCO/VA: diffusing capacity for carbon monoxide corrected by alveolar volume; FVC: forced vital capacity; OR (95% CI), odds ratio, and 2-sided 95% confidence interval of the mean; sPAP: estimated systolic pulmonary artery pressure; SSc: systemic sclerosis.
[a]All data derived from 987 patients except when indicated (n =).

Supplementary Table S4. Immunological features, causes of death and survival of 987 limited cutaneous SSc patients according to the SSc pattern by nailfold videocapillaroscopy.

Autoantibodies, causes of death, and survival, n (%) [a]	Late pattern 367 (37.2%)	Non-Late pattern 620 (62.8%)	Univariate analysis OR (95% CI)	*p*
Autoantibodies, n (%)				
Anti-nuclear antibodies (n = 984)	348 (95%)	581 (94%)	1.13 (0.64-2.01)	0.774
Anti-centromere antibodies (n = 896)	199 (60%)	353 (62%)	0.92 (0.69-1.21)	0.569
Anti-Topoisomerase I (n = 876)	40 (13%)	48 (8.5%)	1.58 (1.01-2.47)	0.046
Anti-RNA polymerase III (n = 223)	7 (11%)	8 (5.0%)	2.43 (0.84-7.03)	0.132
Anti-PM-Scl (n = 534)	14 (7.7%)	21 (5.9%)	1.33 (0.66-2.67)	0.462
Death from all causes, n (%) (n = 152)	51 (14%)	101 (16%)	0.83 (0.58-1.19)	0.362
Causes of death				
Interstitial lung disease	2 3.9%)	9 (8.9%)	0.42 (0.09-2.01)	0.336
Pulmonary arterial hypertension	12 (24%)	17 (17%)	1.52 (0.66-3.49)	0.383
ILD related pulmonary hypertension	1 (2.0%)	8 (7.9%)	0.23 (0.03-1.91)	0.273
Scleroderma renal crisis	1 (2.0%)	2 (2.0%)	0.99 (0.09-11.18)	1.000
Ischaemic cardiopathy	2 (3.9%)	4 (4.0%)	0.99 (0.18-5.59)	1.000
Stroke	2 (3.9%)	2 (2.0%)	2.02 (0.28-14.78)	0.602
Chronic renal failure	0 (0.0%)	2 (2.0%)	-	0.551
Malignancy	6 (12%)	14 (14%)	0.83 (0.30-2.30)	0.804
Sepsis	6 (12%)	7 (6.9%)	1.79 (0.57-5.64)	0.362
Pulmonary embolism	1 (2.0%)	1 (0.99%)	2.00 (0.12-32.64)	1.000
Other causes	14 (27%)	34 (34%)	0.75 (0.36-1.56)	0.466
Not specified	3 (5.9%)	1 (0.99%)	6.25 (0.63-61.67)	0.110
Scleroderma-related causes, n (%) (n=83)	22 (46%)	46 (46%)	0.99 (0.50-1.98)	1.000
Median survival time since first SSc symptom, y (IQR)	48.4 (40.8-56.0)	45.9 (41.1-50.8)	-	0.415
Cumulative survival rates since disease onset				
At 5 years	0.970	0.973	-	0.785
At 10 years	0.940	0.948	-	0.673
At 20 years	0.893	0.837	-	0.231
At 30 years	0.761	0.747	-	0.552

ILD: interstitial lung disease; IQR: interquartile range; SSc: systemic sclerosis.
[a]All data derived from 987 patients except when indicated (n =).