Supplemental Online Content

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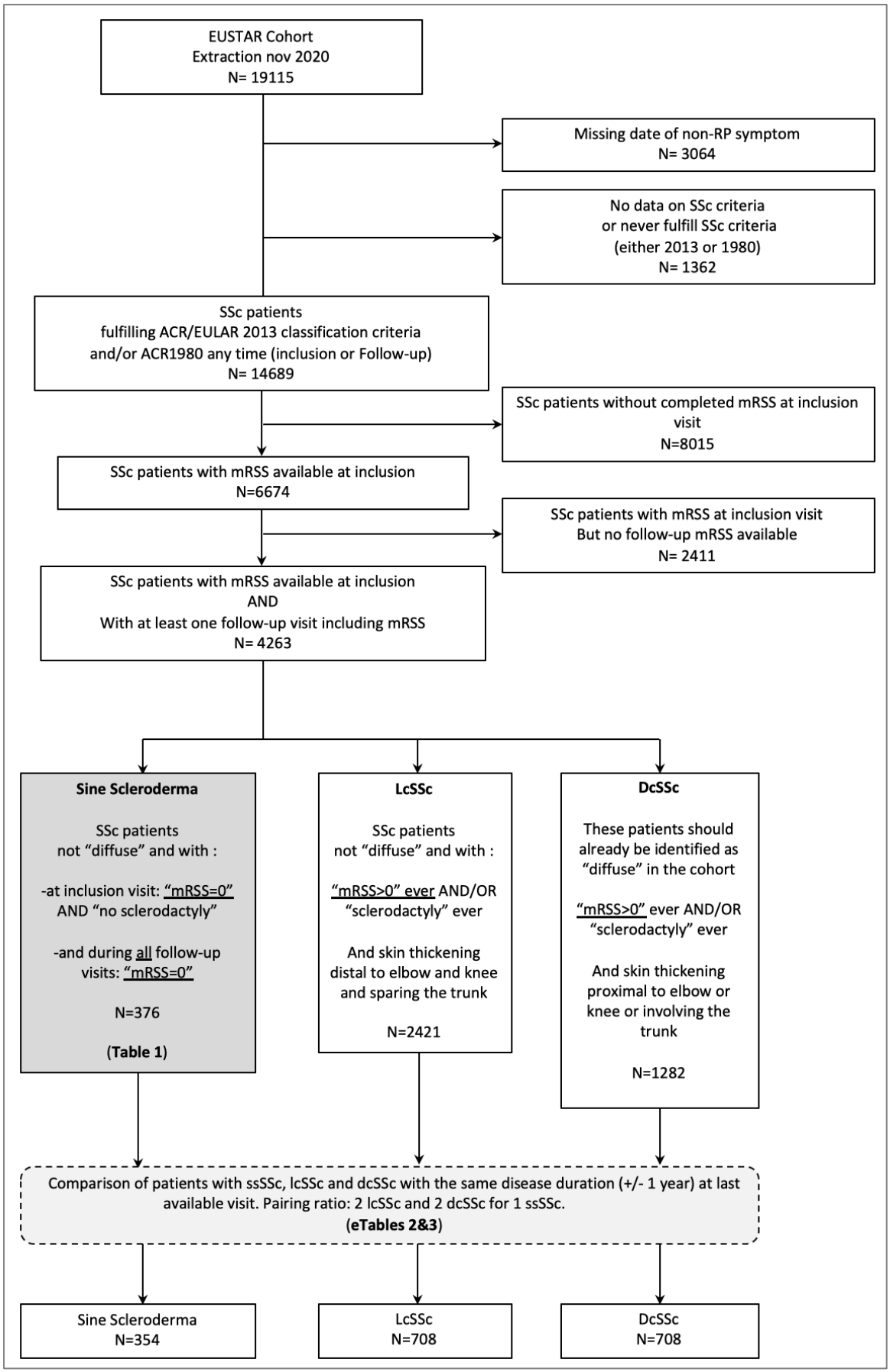
This supplemental material has been provided by the authors to give readers additional information about their work.

eMethods

We explored demographics, disease characteristics, cardio-pulmonary features, disease activity, and immunological findings among patients with ssSSc at inclusion visit. Quantitative variables were expressed using mean and standard deviation (SD) in case of Gaussian distribution, and median and first and third quartiles in case of non-Gaussian distribution. Qualitative variables were expressed as numbers and percentages. We compared these characteristics among ssSSc patients with lcSSc patients and dcSSc patients, respectively, at last available visit. Comparison between groups were assessed using T-test for quantitative variables with Gaussian distribution, Wilcoxon rank sum test for quantitative variables with non-Gaussian distribution, and Chi^2 or Fisher exact test as appropriate for qualitative variables. Logistic regressions were used to explore the association between variables at last available visit and ssSSc with DU vs. ssSSc without DU, ssSSc with Puffy fingers vs. ssSSc without Puffy Fingers, ssSSc with Telangiectasia vs. ssSSc without Telangiectasia, ILD vs. ssSSc without ILD. A univariate model was fitted for each of the variables; a multivariate model was fitted including all variables with $P<0.2$ in their univariate models and excluding variables with 1/3 of missing data. Odds Ratios and 95% CIs were reported for logistic regression. All analyses described above were conducted in SAS (version 9.4). Time to event analysis was conducted for all-cause mortality. We plotted survival curves by SSc group and did pairwise comparisons between ssSSc, lcSSc and dcSSc. Figures were plotted via R package "survival" and "survminer" (R version 4.0.2). We performed all tests with a significance level of $P<0.05$.



eFigure 1: Flow Chart and selection strategy

eTable 1: Clinical characteristics associated with Puffy fingers (ever) in sine scleroderma patients (ssSSc).

Characteristics of ssSSc patients at last visit (n=353)	Univariate modeling for Puffy fingers (ever versus never)		Multivariable modeling for Puffy fingers (ever versus never)	
	Odds Ratio (95% CI)	P-value	Odds Ratio (95% CI)	P-value
Age at last visit (years)	1.011 (0.996, 1.027)	0.1500	0.971 (0.947, 0.996)	0.0237
Gender (ref=Female)	0.839 (0.391, 1.799)	0.6513		
Disease duration since RP onset (years) (last visit)	1.009 (0.989, 1.029)	0.3954		
Raynaud's Phenomenon (ref=Never)	2.009 (0.125, 32.400)	0.6230		
Telangiectasia (ref=Never)	1.013 (0.635, 1.616)	0.9579		
Pitting scars (ref=Never)	1.040 (0.646, 1.674)	0.8720		
DU (ref=Never)	1.085 (0.661, 1.780)	0.7476		
Joint synovitis (ref=Never)	1.515 (0.838, 2.742)	0.1694	0.518 (0.189, 1.419)	0.2007
Tendon friction rubs (ref=Never)	0.689 (0.313, 1.520)	0.3565		
CK-elevation (ref=Never)	1.315 (0.627, 2.759)	0.4687		
Esophageal symptoms (ref=Never)	1.182 (0.728, 1.919)	0.4996		
Stomach symptoms (ref=Never)	0.942 (0.605, 1.466)	0.7901		
Intestinal symptoms (ref=Never)	1.196 (0.784, 1.826)	0.4063		
History of scleroderma renal crisis (ref=Never)	0.847 (0.235, 3.056)	0.7997		
Proteinuria (ref=Never)	0.689 (0.358, 1.327)	0.2655		
Lung fibrosis on X-Rays or HRCT or presence of ILD (ref=Never)	1.216 (0.786, 1.881)	0.3798		
Significant dyspnea (ref=Never)	0.668 (0.375, 1.191)	0.1715	2.757 (1.066, 7.133)	0.0365
ppDLCO (last visit)	1.006 (0.992, 1.020)	0.4314		
ppFVC (last visit)	1.021 (1.009, 1.033)	0.0007	1.002 (0.984, 1.019)	0.8645
ppTLC (last visit)	1.009 (0.994, 1.024)	0.2417		
sPAP >40mmHg (ref=Never)	1.393 (0.663, 2.926)	0.3818		
Pulmonary hypertension (ref=Never)	0.866 (0.468, 1.602)	0.6464		
Left ventricular ejection fraction (last visit)	0.989 (0.941, 1.040)	0.6792		
Diastolic heart dysfunction (ref=Never)	1.116 (0.714, 1.745)	0.6306		
Conduction block (ref=Never)	0.825 (0.463, 1.469)	0.5127		
EScSG disease activity index (2001) (last visit)	0.855 (0.632, 1.157)	0.3093		
EScSG disease activity index (2016) (last visit)	1.198 (0.785, 1.827)	0.4027		
ANA+ (ref=Negative)	1.445 (0.381, 5.476)	0.5881		
ACA+ (ref=Negative)	1.873 (1.207, 2.906)	0.0051	1.157 (0.541, 2.475)	0.7070
ATA+ (ref=Negative)	0.953 (0.537, 1.690)	0.8691		
RNA (ref=Negative)	1.152 (0.347, 3.830)	0.8169		
PmScl+ (ref=Negative)	0.925 (0.307, 2.782)	0.8891		
U1RNP+ (ref=Negative)	0.323 (0.122, 0.856)	0.0231	2.205 (0.529, 9.196)	0.2777
CRP >5mg/L (ref=Never)	0.530 (0.242, 1.157)	0.1108	0.827 (0.209, 3.266)	0.7864

ACA: anti-centromere antibodies ; ANA: Antinuclear antibodies ;ATA: anti-topoisomerase I antibodies ; CK: creatinine-phospho-kinase ; CRP: C-reactive Protein ; DLCO: Diffusion capacities of carbon-monoxyde ; DU: digital Ulcers ; EScSG: European Systemic Sclerosis research group ; FVC: Forced Vital Capacity ; HRCT: High resolution computed tomography ; ILD: Interstitial lung disease ; MMF: Mycophenolate mofetil ; MTX: Methotrexate ; RNA pol III: anti RNA polymerase III antibodies ; RP: Raynaud's Phenomenon ; SD : standard deviation ; sPAP: systolique Pulmonary Arterial Pressure ; SSc : systemic sclerosis ; ssSSc: Systemic sclerosis sine scleroderma ; TLC: Total Lung capacities ; U1-RNP: Anti U1 ribonuclease protein antibodies

eTable 2: Comparison of the clinical characteristics of ssSSc with lcSSc and dcSSc with same disease duration at last available visit.

Characteristics at last visit n, (%) or mean (SD)	ssSSc N=354		lcSSc N=708			dcSSc N=708		
	Data available	Statistics	*Data available*	Statistics	P-value (ssSSc vs lcSSc)	*Data available*	Statistics	P-value (ssSSc vs dcSSc)
Demographics								
Age at last visit (years), mean (SD)	*354/354*	58.4 (13.9)	*708/708*	58.8 (13.0)	0.6193	*708/708*	55.4 (13.4)	0.0007
Male gender, n (%)	*354/354*	31 (8.8%)	*708/708*	111 (15.7%)	0.0018	*708/708*	182 (25.7%)	<.0001
Follow-up duration (years), mean (SD)	*354/354*	3.6 (3.2)	*708/708*	4.4 (3.6)	0.0002	*708/708*	4.3 (3.1)	<.0001
Disease duration since first non-RP symptom (years) (last visit), mean (SD)	*354/354*	10.3 (7.2)	*708/708*	10.3 (7.2)	0.9911	*708/708*	10.3 (7.2)	0.9968
Disease duration since RP onset (years) (last visit), mean (SD)	*329/354*	14.3 (10.8)	*657/708*	14.2 (10.4)	0.9714	*669/708*	12.5 (9.1)	0.0222
Disease characteristics								
Skin manifestations								
Raynaud's Phenomenon (ever), n (%)	*333/354*	331 (99.4%)	*664/708*	656 (98.8%)	0.5099	*682/708*	672 (98.5%)	0.3558
Telangiectasia (ever), n (%)	*336/354*	221 (65.8%)	*609/708*	455 (74.7%)	0.0036	*607/708*	484 (79.7%)	<.0001
DU (ever), n (%)	*333/354*	94 (28.2%)	*604/708*	321 (53.1%)	<.0001	*608/708*	415 (68.3%)	<.0001
Pitting scars (ever), n (%)	*332/354*	105 (31.6%)	*601/708*	391 (65.1%)	<.0001	*601/708*	463 (77.0%)	<.0001
Puffy fingers (ever), n (%)	*351/354*	224 (63.8%)	*703/708*	579 (82.4%)	<.0001	*700/708*	613 (87.6%)	<.0001
mRSS (last visit), mean (SD)	*320/354*	0.0 (0.0)	*551/708*	5.8 (6.0)	<.0001	*571/708*	14.1 (9.4)	<.0001
Other manifestations								
Joint synovitis (ever), n (%)	*354/354*	60 (16.9%)	*708/708*	172 (24.3%)	0.0063	*708/708*	218 (30.8%)	<.0001
Tendon friction rubs (ever), n(%)	*354/354*	26 (7.3%)	*706/708*	101 (14.3%)	0.0010	*706/708*	215 (30.5%)	<.0001
Muscle weakness (ever), n (%)	*354/354*	86 (24.3%)	*708/708*	195 (27.5%)	0.2579	*707/708*	315 (44.6%)	<.0001
CK-elevation (ever), n (%)	*296/354*	36 (12.2%)	*644/708*	79 (12.3%)	0.9636	*663/708*	146 (22.0%)	0.0003
Esophageal symptoms (ever), n (%)	*354/354*	265 (74.9%)	*708/708*	564 (79.7%)	0.0746	*708/708*	622 (87.9%)	<.0001

Stomach symptoms (ever), n (%)	*354/354*	123 (34.7%)	*707/708*	295 (41.7%)	0.0282	*708/708*	383 (54.1%)	<.0001
Intestinal symptoms (ever), n (%)	*354/354*	182 (51.4%)	*707/708*	326 (46.1%)	0.1031	*707/708*	354 (50.1%)	0.6802
History of scleroderma renal crisis (ever), n (%)	*354/354*	10 (2.8%)	*708/708*	16 (2.3%)	0.5744	*708/708*	35 (4.9%)	0.1062
Proteinuria (ever), n (%)	*336/354*	40 (11.9%)	*680/708*	96 (14.1%)	0.3298	*685/708*	126 (18.4%)	0.0083
Lung fibrosis on X-Rays or HRCT or presence of ILD (ever), n (%)	*331/354*	165 (49.8%)	*666/708*	380 (57.1%)	0.0313	*691/708*	518 (75.0%)	<.0001
DLCO (%pred) (last visit), mean (SD)	*214/354*	71.9 (19.7)	*420/708*	67.2 (19.7)	0.0045	*418/708*	60.0 (19.9)	<.0001
FVC (%pred) (last visit), mean (SD)	*241/354*	100.4 (22.2)	*463/708*	92.5 (20.7)	<.0001	*469/708*	82.4 (22.6)	<.0001
TLC (%pred) (last visit), mean (SD)	*175/354*	101.7 (20.8)	*355/708*	93.3 (19.0)	<.0001	*351/708*	83.9 (21.1)	<.0001
sPAP >40mmHg, echocardiography (ever), n (%)	*298/354*	32 (10.7%)	*586/708*	107 (18.3%)	0.0037	*604/708*	127 (21.0%)	0.0001
Pulmonary hypertension (ever), n (%)	*310/354*	47 (15.2%)	*633/708*	151 (23.9%)	0.0021	*649/708*	186 (28.7%)	<.0001
Left ventricular ejection fraction (%) (last visit), mean (SD)	*219/354*	61.0 (5.5)	*416/708*	61.3 (7.4)	0.2501	*433/708*	59.9 (8.2)	0.5450
Diastolic heart dysfunction (ever), n (%)	*327/354*	133 (40.7%)	*652/708*	293 (44.9%)	0.2042	*667/708*	275 (41.2%)	0.8669
Conduction block (ever), n (%)	*305/354*	51 (16.7%)	*641/708*	152 (23.7%)	0.0144	*671/708*	201 (30.0%)	<.0001
Disease activity at last available visit								
EScSG disease activity index (2001) (last visit), mean (SD)	*191/354*	0.7 (0.9)	*371/708*	1.3 (1.4)	<.0001	*425/708*	2.1 (1.8)	<.0001
EScSG disease activity index (2016) (last visit), mean (SD)	*354/354*	0.2 (0.6)	*708/708*	0.3 (0.9)	0.0297	*708/708*	0.5 (1.3)	0.0017
Immunological findings								
ANA+ (ever), n (%)	*352/354*	343 (97.4%)	*706/708*	693 (98.2%)	0.4422	*702/708*	678 (96.6%)	0.4485
ACA+ (ever), n (%)	*347/354*	214 (61.7%)	*688/708*	288 (41.9%)	<.0001	*680/708*	111 (16.3%)	<.0001
ATA+ (ever), n (%)	*348/354*	60 (17.2%)	*697/708*	278 (39.9%)	<.0001	*689/708*	419 (60.8%)	<.0001
RNA pol III+ (ever), n (%)	*308/354*	13 (4.2%)	*509/708*	26 (5.1%)	0.5643	*539/708*	68 (12.6%)	<.0001
PmScl+ (ever), n (%)	*290/354*	15 (5.2%)	*495/708*	17 (3.4%)	0.2346	*536/708*	39 (7.3%)	0.2430
U1RNP+ (ever), n (%)	*322/354*	19 (5.9%)	*619/708*	35 (5.7%)	0.8775	*615/708*	20 (3.3%)	0.0539
CRP >5mg/L (ever), n (%)	*314/354*	27 (8.6%)	*538/708*	50 (9.3%)	0.7329	*495/708*	56 (11.3%)	0.2150
Therapeutics								

Immunomodulatory therapies (ever)*, n (%)	*338/354*	167 (49.4%)	*611/708*	392 (64.2%)	<.0001	*612/708*	459 (75.0%)	<.0001
MMF (ever), n (%)	*338/354*	15 (4.4%)	*611/708*	51 (8.3%)	0.0234	*612/708*	85 (13.9%)	<.0001
MTX (ever), n (%)	*338/354*	47 (13.9%)	*611/708*	169 (27.7%)	<.0001	*612/708*	208 (34.0%)	<.0001
Corticosteroids (ever), n (%)	*338/354*	127 (37.6%)	*611/708*	227 (37.2%)	0.8976	*612/708*	311 (50.8%)	<.0001
Cyclophosphamide (ever), n (%)	*338/354*	28 (8.3%)	*611/708*	86 (14.1%)	0.0086	*612/708*	169 (27.6%)	<.0001
Calcium channel inhibitors (ever), n (%)	*338/354*	222 (65.7%)	*611/708*	388 (63.5%)	0.5025	*612/708*	414 (67.6%)	0.5373
Sildenafil (ever), n (%)	*338/354*	41 (12.1%)	*611/708*	73 (11.9%)	0.9340	*612/708*	81 (13.2%)	0.6260
Bosentan (ever), n (%)	*338/354*	38 (11.2%)	*611/708*	102 (16.7%)	0.0234	*612/708*	170 (27.8%)	<.0001
Iloprost (ever), n (%)	*338/354*	32 (9.5%)	*611/708*	124 (20.3%)	<.0001	*612/708*	157 (25.7%)	<.0001

ACA: anti-centromere antibodies ; ANA: Antinuclear antibodies ;ATA: anti-topoisomerase I antibodies ; CK: creatinine-phospho-kinase ; CRP: C-reactive Protein ; DLCO: Diffusion capacities of carbon-monoxyde ; DU: digital Ulcers ; EScSG: European Systemic Sclerosis research group ; FVC: Forced Vital Capacity ; HRCT: High resolution computed tomography ; ILD: Interstitial lung disease ; MMF: Mycophenolate mofetil ; MTX: Methotrexate ; RNA pol III: anti RNA polymerase III antibodies ; RP: Raynaud's Phenomenon ; SD : standard deviation ; sPAP: systolique Pulmonary Arterial Pressure ; SSc : systemic sclerosis ; ssSSc: Systemic sclerosis sine scleroderma ; TLC: Total Lung capacities ; U1-RNP: Anti U1 ribonuclease protein antibodies

*= Abatacept, Abatacept (iv or sc), Enbrel, Gololimumab, Humira, Infliximab, JAK kinase inhibitors, Rituximab, TNF-alpha antagonist, Tnf alpha antagonist, Tocilizumab, Tocilizumab (iv or sc), Other biologic therapy, Azathioprine, Cyclosporine A, Cyclophosphamide, D-penicillamine, Chloroquine/Hydroxychloroquine, Imatinib, Leflunomide, Methotrexate, Mycophenolate mofetyl, Prednisone, Sulfasalazine

eTable 3: Comparison of the clinical characteristics of ssSSc (with Lung fibrosis/ILD) with lcSSc with Lung fibrosis/ILD) and dcSSc with Lung fibrosis/ILD).

Characteristics at last visit n, (%) or mean (SD)	ssSSc with Lung fibrosis/ILD N=165		lcSSc with Lung fibrosis/ILD N=380			dcSSc with Lung fibrosis/ILD N=518		
	Data available	Statistics	*Data available*	Statistics	P-value (ssSSc vs lcSSc)	*Data available*	Statistics	P-value (ssSSc vs dcSSc)
Demographics								
Age at last visit (years), mean (SD)	165/165	61.2 (12.9)	380/380	60.0 (12.8)	0.3194	518/518	55.8 (13.4)	<0.0001
Male gender, n (%)	165/165	19 (11.5%)	380/380	71 (18.7%)	0.0384	518/518	132 (25.5%)	0.0002
Follow-up duration (years), mean (SD)	165/165	4.0 (3.5)	380/380	4.8 (3.7)	0.0088	518/518	4.4 (3.2)	0.0398
Disease duration since first non-RP symptom (years) (last visit), mean (SD)	165/165	10.8 (7.8)	380/380	10.6 (7.0)	0.7440	518/518	10.7 (7.2)	0.8681
Disease duration since RP onset (years) (last visit), mean (SD)	149/165	14.8 (10.9)	356/380	13.7 (9.1)	0.5433	497/518	12.8 (8.9)	0.0796
Cardio-pulmonary parameters								
Significant dyspnea (ever), n (%)	146/165	33 (22.6%)	354/380	97 (27.4%)	0.2660	497/518	173 (34.8%)	0.0055
DLCO (%pred) (last visit), mean (SD)	103/165	65.3 (20.6)	230/380	61.1 (18.2)	0.0651	306/518	56.2 (19.2)	<0.0001
FVC (%pred) (last visit), mean (SD)	117/165	92.8 (22.6)	265/380	86.7 (20.6)	0.0103	349/518	78.9 (22.3)	<0.0001
TLC (%pred) (last visit), mean (SD)	92/165	98.3 (21.2)	195/380	88.8 (19.7)	0.0002	258/518	81.0 (21.0)	<0.0001
sPAP >40mmHg, echocardiography (ever), n (%)	150/165	17 (11.3%)	324/380	74 (22.8%)	0.0031	451/518	112 (24.8%)	0.0005
Pulmonary hypertension (ever), n (%)	147/165	31 (21.1%)	345/380	111 (32.2%)	0.0130	480/518	168 (35.0%)	0.0015
Immunological findings								
ANA+ n (%)	164/165	161 (98.2%)	380/380	374 (98.4%)	1.0000	515/518	499 (96.9%)	0.5868
ACA+ , n (%)	162/165	84 (51.9%)	369/380	97 (26.3%)	<0.0001	508/518	66 (13.0%)	<0.0001
ATA+ , n (%)	163/165	36 (22.1%)	376/380	205 (54.5%)	<0.0001	510/518	351 (68.8%)	<0.0001
RNA pol III+ , n (%)	143/165	7 (4.9%)	275/380	17 (6.2%)	0.5916	400/518	46 (11.5%)	0.0224

PmScl+ , n (%)	135/165	10 (7.4%)	261/380	9 (3.4%)	0.0806	398/518	28 (7.0%)	0.8845
U1RNP+ , n (%)	154/165	8 (5.2%)	337/380	17 (5.0%)	0.9440	459/518	16 (3.5%)	0.3441
CRP >5mg/L (ever), n (%)	156/165	19 (12.2%)	300/380	29 (9.7%)	0.4068	369/518	48 (13.0%)	0.7948
Therapeutics								
Immunomodulatory therapies (ever)*, n (%)	162/165	89 (54.9%)	334/380	241 (72.2%)	0.0001	447/518	350 (78.3%)	<0.0001
MMF (ever), n (%)	162/165	13 (8.0%)	334/380	42 (12.6%)	0.1301	447/518	67 (15.0%)	0.0246
MTX (ever), n (%)	162/165	20 (12.3%)	334/380	84 (25.1%)	0.0010	447/518	144 (32.2%)	<0.0001
Corticosteroids (ever), n (%)	162/165	69 (42.6%)	334/380	158 (47.3%)	0.3231	447/518	254 (56.8%)	0.0019
Cyclophosphamide (ever), n (%)	162/165	27 (16.7%)	334/380	74 (22.2%)	0.1545	447/518	147 (32.9%)	<0.0001
Calcium channel inhibitors (ever), n (%)	162/165	90 (55.6%)	334/380	214 (64.1%)	0.0678	447/518	320 (71.6%)	0.0002
Sildenafil (ever), n (%)	162/165	14 (8.6%)	334/380	48 (14.4%)	0.0704	447/518	62 (13.9%)	0.0845
Bosentan (ever), n (%)	162/165	18 (11.1%)	334/380	65 (19.5%)	0.0195	447/518	145 (32.4%)	<0.0001
Iloprost (ever), n (%)	162/165	14 (8.6%)	334/380	67 (20.1%)	0.0013	447/518	121 (27.1%)	<0.0001

ACA: anti-centromere antibodies ; ANA: Antinuclear antibodies ;ATA: anti-topoisomerase I antibodies ; CK: creatinine-phospho-kinase ; CRP: C-reactive Protein ; DLCO: Diffusion capacities of carbon-monoxyde ; DU: digital Ulcers ; EScSG: European Systemic Sclerosis research group ; FVC: Forced Vital Capacity ; HRCT: High resolution computed tomography ; ILD: Interstitial lung disease ; MMF: Mycophenolate mofetil ; MTX: Methotrexate ; RNA pol III: anti RNA polymerase III antibodies ; RP: Raynaud's Phenomenon ; SD : standard deviation ; sPAP: systolique Pulmonary Arterial Pressure ; SSc : systemic sclerosis ; ssSSc: Systemic sclerosis sine scleroderma ; TLC: Total Lung capacities ; U1-RNP: Anti U1 ribonuclease protein antibodies

*= Abatacept, Abatacept (iv or sc), Enbrel, Gololimumab, Humira, Infliximab, JAK kinase inhibitors, Rituximab, TNF-alpha antagonist, Tnf alpha antagonist, Tocilizumab, Tocilizumab (iv or sc), Other biologic therapy, Azathioprine, Cyclosporine A, Cyclophosphamide, D-penicillamine, Chloroquine/Hydroxychloroquine, Imatinib, Leflunomide, Methotrexate, Mycophenolate mofetyl, Prednisone, Sulfasalazine

eTable 4: Clinical characteristics associated with ILD and/or Lung Fibrosis ever (i.e. history of ILD and/or Lung Fibrosis at inclusion and/or ILD and/or Lung Fibrosis during follow-up) in sine scleroderma patients (ssSSc).

Characteristics at last visit N=352	Univariate modeling for ILD and/or Lung Fibrosis (ever)		Multivariable modeling for ILD and/or Lung Fibrosis (ever)	
	Odds Ratio (95% CI)	P-value	Odds Ratio (95% CI)	P-value
Age at last visit (years)	1.037 (1.020, 1.054)	<.0001	1.048 (1.009, 1.089)	0.0148
Gender (ref=Female)	1.727 (0.797, 3.745)	0.1664	4.801 (0.416, 55.361)	0.2086
Disease duration since RP onset (years) (last visit)	1.018 (0.998, 1.038)	0.0751	1.003 (0.968, 1.038)	0.8847
Raynaud's Phenomenon (ref=Never)	1.018 (0.063, 16.417)	0.9898		
Telangiectasia (ref=Never)	0.975 (0.618, 1.536)	0.9117		
Previous or Current (ref=Never)	0.813 (0.505, 1.309)	0.3946		
Pitting scars (ref=Never)	1.016 (0.640, 1.613)	0.9477		
Puffy fingers (ref=Never)	1.216 (0.786, 1.881)	0.3799		
Joint synovitis (ref=Never)	1.355 (0.777, 2.366)	0.2846		
Tendon friction rubs (ref=Never)	1.331 (0.593, 2.984)	0.4883		
CK-elevation (ref=Never)	1.851 (0.917, 3.738)	0.0858	4.194 (1.137, 15.471)	0.0314
Esophageal symptoms (ref=Never)	0.941 (0.581, 1.525)	0.8056		
Stomach symptoms (ref=Never)	1.022 (0.659, 1.586)	0.9221		
Intestinal symptoms (ref=Never)	1.404 (0.923, 2.136)	0.1127	2.427 (1.075, 5.478)	0.0328
History of scleroderma renal crisis (ref=Never)	1.199 (0.317, 4.541)	0.7894		
Proteinuria (ref=Never)	1.277 (0.652, 2.501)	0.4756		
Significant dyspnea (ref=Never)	3.764 (1.925, 7.357)	0.0001	2.503 (0.711, 8.806)	0.1529
ppDLCO (last visit)	0.963 (0.948, 0.979)	<.0001		
ppFVC (last visit)	0.967 (0.954, 0.980)	<.0001	0.964 (0.943, 0.985)	0.0007
ppTLC (last visit)	0.980 (0.965, 0.995)	0.0083		
sPAP >40mmHg (ref=Never)	1.273 (0.630, 2.573)	0.5019		
Pulmonary hypertension (ref=Never)	2.499 (1.321, 4.730)	0.0049	0.625 (0.189, 2.069)	0.4417
Left ventricular ejection fraction (last visit)	0.965 (0.918, 1.013)	0.1498		
Diastolic heart dysfunction (ref=Never)	1.924 (1.237, 2.991)	0.0037	1.364 (0.553, 3.366)	0.5006
Conduction block (ref=Never)	2.493 (1.338, 4.645)	0.0040	1.031 (0.350, 3.039)	0.9556
EScSG disease activity index (2001) (last visit)	1.802 (1.272, 2.551)	0.0009		
EScSG disease activity index (2016) (last visit)	1.190 (0.807, 1.754)	0.3798		
ANA+ (ref=Negative)	1.767 (0.416, 7.510)	0.4406		
ACA+ (ref=Negative)	0.429 (0.275, 0.671)	0.0002	1.581 (0.578, 4.324)	0.3721
ATA+ (ref=Negative)	2.318 (1.282, 4.194)	0.0054	3.246 (0.850, 12.390)	0.0850
RNA (ref=Negative)	1.182 (0.388, 3.601)	0.7684		
PmScl+ (ref=Negative)	3.382 (0.911, 12.554)	0.0686	>999.999 (<0.001, >999.999)	0.9702
U1RNP+ (ref=Negative)	0.740 (0.284, 1.926)	0.5374		
CRP >5mg/L (ref=Never)	2.199 (0.932, 5.185)	0.0718	0.940 (0.219, 4.031)	0.9341

ACA: anti-centromere antibodies ; ANA: Antinuclear antibodies ;ATA: anti-topoisomerase I antibodies ; CK: creatinine-phospho-kinase ; CRP: C-reactive Protein ; DLCO: Diffusion capacities of carbon-monoxyde ; DU: digital Ulcers ; EScSG: European Systemic Sclerosis research group ; FVC: Forced Vital Capacity ; HRCT: High resolution computed tomography ; ILD: Interstitial lung disease ; MMF: Mycophenolate mofetil ; MTX: Methotrexate ; RNA pol III: anti RNA polymerase III antibodies ; RP: Raynaud's Phenomenon ; SD : standard deviation ; sPAP: systolique Pulmonary Arterial Pressure ; SSc : systemic sclerosis ; ssSSc: Systemic sclerosis sine scleroderma ; TLC: Total Lung capacities ; U1-RNP: Anti U1 ribonuclease protein antibodies