Supplementary Material

Distinct cardiovascular phenotypes are associated with prognosis in systemic sclerosis: a cardiovascular magnetic resonance study.

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Supplementary Material

Supplementary Table 1: Clinical details of SSc patient cohort.

	SSc patients (n = 260)
Demographics and co-morbidities	
Female, n (%)	207 (80)
Age, years	59 (17-85)
BSA (m^2)	1.7 (1.3-2.3)
Interstitial lung disease (any severity), n (%)	117 (45)
Emphysema (any severity), n (%)	32 (12)
Hypertension, n (%)	77 (30)
Diabetes mellitus, n (%)	20 (8)
Ischaemic heart disease, n (%)	24 (9)
Atrial fibrillation, n (%)	24 (9)
Immunomodulatory therapy	
Mycophenolate mofetil, n (%)	105 (40)
Glucocorticoids, n (%)	102 (39)
Azathioprine, n (%)	13 (5)
Hydroxychloroquine, n (%)	63 (24)
Leflunomide, n (%)	2 (1)
Tacrolimus, n (%)	4 (2)
Rituximab, n (%)	9 (3)
Cyclophosphamide, n (%)	33 (13)
Nintedanib, n (%)	1 (0.5)
Tocilizumab, n (%)	5 (2)
Adalimumab, n (%)	1 (0.5)
Golimumab, n (%)	1 (0.5)
SSc details	
Limited/diffuse/sine SSc, n (%)	167/90/3 (64/35/1)
Overlap syndromes, n (%)	79 (30)
- Myositis, n (%)	44 (17)
- Inflammatory arthritis, n (%)	23 (9)
- Sjögren's syndrome, n (%)	18 (7)
- SLE, n (%)	13 (5)
Disease duration (diagnosis to CMR), years	9 (0-47)
Disease severity score	2 (1-4)
Autoantibody subset, %	
Anticentromere	88 (34)
Anti-topoisomerase I	56 (22)
Anti-RNA polymerase	21 (8)
Anti-U3 RNP	18 (7)
Anti-PM/Scl	11 (4)
Other*	25 (10)
ANA+ENA-	87 (33)
ANA-	13 (5)

SSc-associated PH	
Patients diagnosed with SSc-PH, n (%)	112 (43)
Group 1/2/3/4 PH, n (% of PH diagnoses)	78/24/8/2 (70/21/7/2)
Incident/Prevalent PH diagnosis, n (%)	26/54 (32.5/67.5)
Targeted therapies for Group 1 PH patients (n=78)	
PDE5I, n (% of Group 1 PH patients)	54
ERA, n (% of Group 1 PH patients)	38
SGCS, n (% of Group 1 PH patients)	2
Prostanoids (IV/oral), n (% of Group 1 PH patients)	4 (5) / 2 (3)

ANA, antinuclear antibody; BSA, body surface area; CMR, cardiovascular magnetic resonance; ENA, anti-extractable nuclear antigen; ERA = endothelin receptor antagonist; IV = intravenous; m, metres; PDE5I = phosphodiesterase type 5 inhibitors; PH, pulmonary hypertension; SGCS = soluble guanylate cyclase stimulators; SLE, systemic lupus erythematosus; SSc, scleroderma/systemic sclerosis.

Supplementary Table 2: CMR indications.

Tissue characterisation	Known / suspected PH	Volumes / function / mass	Myocardial ischaemia	Number
X				117
	X			80
		X		9
			X	3
X	X			24
X		X		9
X			X	12
	X	X		2
X	X		X	2
X		X	X	1
X	X	X	X	1

PH, pulmonary hypertension.

Supplementary Table 3: CMR parameters of SSc patient cohort.

CMR metrics	SSc patients (n = 260)
RVEDVi (mL/m^2)	73 (30-184)
RVESVi (mL/m^2)	28 (10-119)
RVSVi (mL/m^2)	43 (18-77)
RVEF (%)	61 (19-82)
RVMi (g/m^2)	28 (12-76)
LVEDVi (mL/m^2)	67 (27-131)
LVESVi (mL/m^2)	22 (6-83)
LVSVi (mL/m^2)	44 (14-83)
LVEF (%)	68 (15-87)
LVMi (g/m^2)	63 (29-140)
LAi area (cm/m^2)	13 (6-29)
RAi area (cm/m^2)	12 (4-30)
T1 (ms)	1071 (932-1290)
T2 (ms)	50 (40-59)
Major insertion point LGE, n (%)	44 (17)
Major non-insertion point LGE, n (%)	47 (18)
Pericardial effusion, n (%)	71 (27)

Values are median (range) or n (%). BSA, body surface area; cm, centimetres; CMR, cardiovascular magnetic resonance; EDV, end-diastolic volume; EF, ejection fraction; ESV, end-systolic volume; g, grammes; I, indexed for body-surface area; LA, left atrial; LV, left ventricular; M, mass; m, metres; mL, millilitres; ms, milliseconds; RA, right atrial; RV, right ventricular; SV, stroke volume.

Supplementary Table 4: CMR and clinical data for SSc patients with and without PH.

Variable	SSc without PH (n=148)	SSc-associated PH (n=112)	*P*-value
CMR metrics			
RVEDVi (mL/m^2)	70 (35-126)	80 (30-184)	**<0.001**
RVESVi (mL/m^2)	25 (10-69)	33 (10-119)	**<0.001**
RVSVi (mL/m^2)	44 (19-77)	43 (18-77)	0.63
RVEF (%)	63 (26-82)	56 (19-82)	**<0.001**
RV MVR (g/mL)	0.4 (0.2-0.9)	0.4 (0.2-0.8)	**<0.001**
RAi area (cm/m^2)	11 (4-30)	13 (6-22)	**<0.001**
LVEDVi (mL/m^2)	69 (39-123)	65 (27-131)	**0.037**
LVESVi (mL/m^2)	23 (9-83)	20 (6-82)	0.057
LVSVi (mL/m^2)	45 (14-83)	43 (18-71)	**0.030**
LVEF (%)	67 (15-87)	68 (28-86)	0.42
LV MVR (g/mL)	0.9 (0.4-2.4)	1.0 (0.6-2.2)	**0.013**
LAi area (cm/m^2)	13 (7-29)	13 (6-21)	0.93
COi (L/min/m^2)	3.2 (1.0-5.9)	3.0 (1.4-7.4)	0.055
T1 (ms)	1063 (932-1195)	1083 (983-1290)	**<0.001**
T2 (ms)	49 (41-59)	51 (40-59)	**0.0032**
ECV (%)	32 (23-43)	32 (25-48)	**0.0017**
Major insertion point LGE (%)	2	38	**<0.001**
Major non-insertion point LGE (%)	22	14	0.10
Pericardial effusion (%)	20	36	**0.0048**
Heart rate (bpm)	74 (46-112)	75 (48-113)	0.35
Haemodynamic data (from RHC within 1 year of CMR study)			
mPAP (mmHg)	18 (11-24)	31 (19-69)	**<0.001**
RAP (mmHg)	5 (1-15)	8 (0-30)	**<0.001**
PCWP (mmHg)	10 (4-16)	12 (5-22)	**<0.001**
Patient demographics			
Height (m)	1.6 (1.5-1.9)	1.6 (1.4-1.9)	**0.021**
Weight (kg)	64 (39-115)	65 (39-123)	0.998
Age (years)	58 (17-83)	64 (21-85)	0.056
Sex (female, %)	78	83	0.44
SSc details			
Limited (%)	56	73	**0.0041**
Diffuse (%)	43	24	**0.0023**
Overlap syndrome (%)	33	26	0.28
Inflammatory arthritis (%)	9	8	0.83
Sjögren's syndrome (%)	6	8	0.62
SLE (%)	6	3	0.40
Myositis (%)	20	12	0.13
Disease duration (years)	6 (0-47)	12 (0-42)	**0.011**
Disease severity score	2 (1-4)	3 (1-4)	**0.0064**

Autoantibody subset, %			
Anticentromere	24	46	**<0.001**
Anti-topoisomerase I	28	13	**0.0059**
Anti-RNA polymerase	10	5	0.18
Anti-U3 RNP	8	5	0.46
Anti-PM/Scl	6	0	**0.026**
Other*	8	11	0.40
ANA+ENA-	29	39	0.11
ANA-	7	3	0.16

Values are median (range) or n (%). Bold denotes statistically significant values ($P < 0.05$). ANA, antinuclear antibody; bpm, beats per minute; cm, centimetres; CMR, cardiovascular magnetic resonance; CO, cardiac output; ECV, extracellular volume; EDV, end-diastolic volume; EF, ejection fraction; ENA, anti-extractable nuclear antigen; ESV, end-systolic volume; g, grammes; I, indexed for body-surface area; kg, kilogrammes; L, litre; LA, left atrial; LGE, late gadolinium enhancement; LV, left ventricular; m, metres; min, minute; mL, millilitres; mmHg, millimetre of mercury; mPAP, mean pulmonary arterial pressure; ms, milliseconds; MVR, mass-to-volume ratio; PCWP, pulmonary capillary wedge pressure; PH, pulmonary hypertension; RA, right atrial; RAP, right atrial pressure; RHC, right heart catheter; RV, right ventricular; SLE, systemic lupus erythematosus; SSc, scleroderma/systemic sclerosis; SV, stroke volume.

Supplementary Table 5: CMR and clinical data for SSc patients with and without overlap syndromes.

Variable	SSc without overlap syndrome (n=181)	SSc with overlap syndrome (n=79)	*P*-value
CMR metrics			
RVEDVi (mL/m^2)	72 (30-142)	74 (37-184)	0.72
RVESVi (mL/m^2)	28 (10-101)	28 (10-119)	0.66
RVSVi (mL/m^2)	43 (18-76)	43 (19-77)	0.58
RVEF (%)	61 (19-82)	61 (26-82)	0.96
RV MVR (g/mL)	0.4 (0.2-0.8)	0.4 (0.2-0.9)	0.80
RAi area (cm/m^2)	12 (4-22)	12 (6-30)	0.67
LVEDVi (mL/m^2)	67 (27-122)	67 (44-131)	0.27
LVESVi (mL/m^2)	22 (6-83)	22 (7-83)	0.31
LVSVi (mL/m^2)	44 (18-79)	44 (14-83)	0.52
LVEF (%)	67 (28-87)	68 (15-86)	0.65
LV MVR (g/mL)	0.9 (0.4-2.4)	1.0 (0.4-1.6)	0.78
LAi area (cm/m^2)	13 (6-29)	13 (7-24)	0.55
COi (L/min/m^2)	3.1 (1.4-7.4)	3.4 (1.0-5.5)	**0.028**
T1 (ms)	1071 (932-1266)	1073 (946-1290)	0.43
T2 (ms)	50 (40-59)	50 (41-57)	0.22
ECV (%)	32 (25-48)	32 (23-47)	0.56
Major insertion point LGE (%)	18	17	1.00
Major non-insertion point LGE (%)	20	17	0.73
Pericardial effusion (%)	29	22	0.29
Heart rate (bpm)	74 (46-112)	76 (48-113)	0.16
Haemodynamic data (from RHC within 1 year of CMR study)			
PH (%)	45	37	0.28
mPAP (mmHg)	25 (11-69)	23 (12-52)	0.23
RAP (mmHg)	6 (0-25)	6 (2-30)	0.59
PCWP (mmHg)	10 (4-22)	12 (4-18)	**0.050**
Patient demographics			
Height (m)	1.6 (1.4-1.9)	1.6 (1.5-1.9)	0.78
Weight (kg)	64 (39-123)	65 (40-115)	0.37
Age (years)	62 (23-85)	55 (17-82)	**<0.001**
Sex (female, %)	77	88	**0.045**
SSc details			
Limited (%)	67	56	0.12
Diffuse (%)	31	44	**0.047**
Overlap syndrome (%)	-	100	-
Inflammatory arthritis (%)	-	29	-
Sjögren's syndrome (%)	-	22	-

SLE (%)	-	16	-
Myositis (%)	-	55	-
Disease duration (years)	9 (0-42)	8 (0-47)	0.81
Disease severity score	2 (1-4)	2 (1-4)	0.17
Autoantibody subset, %			
Anticentromere	39	23	**0.015**
Anti-topoisomerase I	21	23	0.74
Anti-RNA polymerase	10	2	**0.044**
Anti-U3 RNP	5	10	0.19
Anti-PM/Scl	1	11	**<0.001**
Other*	5	20	**<0.001**
ANA+ENA-	36	26	0.15
ANA-	6	6	1.00

Values are median (range) or n (%). Bold denotes statistically significant values ($P < 0.05$). ANA, antinuclear antibody; bpm, beats per minute; cm, centimetres; CMR, cardiovascular magnetic resonance; CO, cardiac output; ECV, extracellular volume; EDV, end-diastolic volume; EF, ejection fraction; ENA, anti-extractable nuclear antigen; ESV, end-systolic volume; g, grammes; I, indexed for body-surface area; kg, kilogrammes; L, litre; LA, left atrial; LGE, late gadolinium enhancement; LV, left ventricular; m, metres; min, minute; mL, millilitres; mmHg, millimetre of mercury; mPAP, mean pulmonary arterial pressure; ms, milliseconds; MVR, mass-to-volume ratio; PCWP, pulmonary capillary wedge pressure; PH, pulmonary hypertension; RA, right atrial; RAP, right atrial pressure; RHC, right heart catheter; RV, right ventricular; SLE, systemic lupus erythematosus; SSc, scleroderma/systemic sclerosis; SV, stroke volume.

Supplementary Table 6: Missingness of CMR variables.

Variable	Missingness, n (%)
RVEDVi	3 (1.2)
RVESVi	3 (1.2)
RVSVi	3 (1.2)
RVEF	2 (0.8)
RV MVR	4 (1.5)
RAi area	5 (1.9)
LVEDVi	3 (1.2)
LVESVi	3 (1.2)
LVSVi	3 (1.2)
LVEF	2 (0.8)
LV MVR	2 (0.8)
LAi area	4 (1.5)
T1	2 (0.8)
T2	6 (2.3)

EDV, end-diastolic volume; EF, ejection fraction; ESV, end-systolic volume; I, indexed for body-surface area; LA, left atrial; LV, left ventricular; MVR, mass-to-volume ratio; RA, right atrial; RV, right ventricular; SV, stroke volume.

Supplementary Table 7: Details of CMR indications and metrics according to CMR phenotype (post hoc *P*-values detailed in supplementary table 8).

Variable	NF-AC (n=128)	BVF (n= 14)	RVF (n= 27)	NF-LC (n= 40)	NF-SC (n= 51)	*P*-value
CMR indication						
Myocardial ischaemia (%)	8	14	3	5	5	0.70
Known / suspected PH (%)	42	0	62	42	41	**0.0017**
Tissue characterisation (%)	71	100	25	60	56	**<0.001**
Volumes / function / mass (%)	7	7	14	7	9	0.6834
CMR metric						
RVEDVi (mL/m^2)	70 (51-94)	105 (58-184)	92 (63-142)	92 (70-131)	58 (30-85)	**<0.001**
RVESVi (mL/m^2)	25 (11-59)	62 (35-119)	64 (31-101)	34 (14-67)	22 (10-47)	**<0.001**
RVSVi (mL/m^2)	44 (32-59)	40 (19-65)	38 (18-61)	58 (45-77)	35 (21-45)	**<0.001**
RVEF (%)	64 (35-82)	37 (26-52)	39 (19-57)	63 (47-82)	62 (45-77)	**<0.001**
RV MVR (g/mL)	0.4 (0.2-0.7)	0.3 (0.2-0.5)	0.5 (0.2-0.8)	0.3 (0.2-0.6)	0.4 (0.2-0.9)	**<0.001**
RAi area (cm/m^2)	12 (5-18)	12 (9-17)	15 (10-22)	14 (11-30)	10 (4-16)	**<0.001**
LVEDVi (mL/m^2)	67 (49-99)	103 (93-131)	50 (36-87)	83 (61-123)	52 (27-70)	**<0.001**
LVESVi (mL/m^2)	22 (7-58)	66 (48-83)	20 (6-44)	24 (11-46)	16 (6-29)	**<0.001**
LVSVi (mL/m^2)	45 (34-58)	40 (14-55)	33 (18-58)	57 (45-83)	37 (21-50)	**<0.001**
LVEF (%)	68 (40-86)	37 (15-53)	59 (32-86)	71 (53-87)	68 (57-83)	**<0.001**
LV MVR (g/mL)	0.9 (0.4-1.6)	0.8 (0.4-1.2)	1.0 (0.7-1.8)	0.9 (0.6-1.4)	1.1 (0.7-2.4)	**<0.001**
LAi area (cm/m^2)	14 (7-29)	13 (10-18)	11 (7-20)	15 (12-24)	11 (6-18)	**<0.001**
COi (L/min/m^2)	3.1 (2.1-4.9)	3.2 (1.0-4.2)	2.4 (1.4-5.5)	3.8 (2.7-7.4)	3.0 (1.7-4.1)	**<0.001**
T1 (ms)	1066 (946-1180)	1087 (1008-1127)	1111 (983-1290)	1089 (990-1202)	1050 (932-1195)	**<0.001**

T2 (ms)	50 (44-59)	48 (40-52)	52 (46-59)	53 (45-58)	48 (41-59)	**<0.001**
ECV (%)	32 (23-43)	30 (25-38)	34 (28-47)	34 (25-48)	29 (25-40)	**<0.001**
Major insertion point LGE (%)	9	15	79	10	15	**<0.001**
Major non-insertion point LGE (%)	17	69	8	18	15	**<0.001**
Pericardial effusion (%)	18	21	59	42	21	**<0.001**
Heart rate (bpm)	70 (46-100)	75 (53-113)	79 (49-109)	66 (48-103)	82 (50-112)	**<0.001**

Values are median (range) or n (%). Bold denotes statistically significant values ($P < 0.05$). bpm, beats per minute; cm, centimetres; CMR, cardiovascular magnetic resonance; CO, cardiac output; ECV, extracellular volume; EDV, end-diastolic volume; EF, ejection fraction; ESV, end-systolic volume; g, grammes; i, indexed for body-surface area; L, litre; LA, left atrial; LGE, late gadolinium enhancement; LV, left ventricular; m, metres; min, minute; mL, millilitres; ms, milliseconds; MVR, mass-to-volume ratio; RA, right atrial; RV, right ventricular; SV, stroke volume.

Supplementary Table 8: Post hoc *P*-values for details of CMR metrics according to CMR phenotype.

Variable	NF-AC vs BVF	NF-AC vs RVF	NF-AC vs NF-LC	NF-AC vs NF-SC	BVF vs RVF	BVF vs NF-LC	BVF vs NF-SC	RVF vs NF-LC	RVF vs NF-SC	NF-LC vs NF-SC
CMR indication										
Myocardial ischaemia	1.0	1.0	1.0	1.0	0.97	0.97	0.97	1.0	1.0	1.0
Known / suspected PH	**0.0049**	0.12	1.0	1.0	**0.0011**	**0.0069**	**0.0069**	0.19	0.16	1.0
Tissue characterisation	**0.030**	**<0.001**	0.19	0.10	**<0.001**	**0.012**	**0.0048**	**0.014**	**0.027**	0.83
Volumes / function / mass	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0
CMR metrics										
RVEDVi	**6.1e-05**	**4.0e-11**	**3.8e-14**	**5.4e-13**	0.97	0.72	**3.1e-07**	0.54	**5.7e-12**	**9.6e-15**
RVESVi	**1.8e-08**	**3.8e-13**	**1.7e-05**	**0.00035**	0.88	**3.3e-06**	**2.8e-08**	**1.8e-08**	**4.3e-12**	**5.9e-09**
RVSVi	0.076	**0.00090**	**4.0e-17**	**4.3e-16**	0.69	**1.8e-07**	0.076	**2.2e-13**	**0.034**	**1.2e-15**
RVEF	**6.7e-09**	**1.9e-13**	0.78	0.60	0.77	**9.6e-08**	**3.6e-08**	**1.6e-10**	**3.4e-11**	0.58
RV MVR	**0.026**	**1.4e-05**	0.59	**0.038**	**6.5e-05**	0.15	**0.0053**	**4.3e-05**	**0.0053**	**0.038**
RAi area	0.20	**4.6e-07**	**4.6e-07**	**3.8e-05**	**0.0074**	0.058	**0.0021**	0.19	**2.0e-08**	**1.1e-10**
LVEDVi	**1.8e-09**	**4.7e-05**	**1.3e-11**	**5.5e-15**	**1.1e-10**	**3.7e-07**	**1.8e-08**	**1.0e-10**	0.89	**5.5e-15**
LVESVi	**3.7e-09**	0.55	**0.048**	**1.6e-06**	**2.8e-10**	**6.2e-12**	**3.2e-08**	0.14	**0.035**	**1.5e-06**
LVSVi	**0.024**	**4.8e-09**	**5.2e-16**	**5.2e-16**	0.057	**4.2e-09**	0.14	**3.5e-14**	0.068	**1.7e-15**
LVEF	**1.6e-08**	**0.015**	0.24	0.40	**1.6e-05**	**1.2e-07**	**6.2e-08**	**0.0078**	**0.0078**	0.73
LV MVR	0.051	**4.9e-05**	0.67	**3.8e-09**	**0.0010**	0.067	**9.3e-05**	**0.0014**	0.33	**1.5e-05**
LAi area	0.80	**1.5e-05**	**1.5e-05**	**1.2e-06**	**0.0011**	**0.019**	**0.0049**	**2.3e-09**	0.26	**5.1e-10**
COi	0.54	**6.9e-05**	**8.6e-06**	**0.015**	0.064	**0.0025**	0.54	**2.5e-07**	**0.011**	**1.6e-07**
T1	0.30	**4.4e-05**	**0.0047**	0.066	**0.042**	0.30	0.080	0.080	**4.4e-05**	**0.00051**
T2	**0.038**	**0.0019**	**2.0e-05**	**4.3e-06**	**0.0014**	**0.00018**	0.98	0.57	**2.6e-06**	**1.3e-07**
ECV	0.20	**0.019**	**0.032**	**0.0031**	**0.013**	**0.018**	0.46	0.50	**0.00027**	**0.00027**

Major insertion point LGE	0.85	**7.8e-11**	0.85	0.58	**0.00081**	0.85	1.0	**3.9e-07**	**7.1e-07**	0.85
Major non-insertion point LGE	**0.0012**	0.74	0.82	0.82	**0.0012**	**0.0041**	**0.0012**	0.77	0.82	0.82
Pericardial effusion	0.81	**0.00046**	**0.018**	0.81	0.091	0.31	1.0	0.31	**0.0062**	0.091
Heart rate	0.25	**0.0092**	0.25	**5.9e-05**	0.56	0.15	0.38	**0.0092**	0.51	**0.00042**

Bold denotes statistically significant values ($P < 0.05$).

CMR, cardiovascular magnetic resonance; CO, cardiac output; ECV, extracellular volume; EDV, end-diastolic volume; EF, ejection fraction; ESV, end-systolic volume; i, indexed for body-surface area; LA, left atrial; LGE, late gadolinium enhancement; LV, left ventricular; MVR, mass-to-volume ratio; RA, right atrial; RV, right ventricular; SV, stroke volume.

Supplementary Table 9: Haemodynamics and demographics in CMR phenotypes (post hoc *P*-values detailed in supplementary table 10).

Variable	NF-AC (n=128)	BVF (n= 14)	RVF (n= 27)	NF-LC (n= 40)	NF-SC (n= 51)	*P*-value
PH details (haemodynamic data from RHC within 1 year of CMR study)						
PH (%)	35	50	92	42	33	**<0.001**
PH Group, n (Group 1/2/3/4)	30/12/4/0	1/5/0/1	20/2/3/0	14/3/0/0	13/2/1/1	-
mPAP (mmHg) all patients	23 (11-56)	24 (15-35)	47 (17-69)	22 (12-61)	25 (12-57)	**<0.001**
mPAP (mmHg) PH patients	30 (22-56)	27 (27-35)	51 (25-69)	33 (20-61)	31 (19-57)	**<0.001**
RAP (mmHg) all patients	6 (0-15)	8 (4-21)	10 (3-30)	6 (1-17)	5 (1-11)	**<0.001**
RAP (mmHg) PH patients	8 (0-12)	14 (8-21)	10 (3-30)	6 (3-17)	5 (1-11)	**0.0073**
PCWP (mmHg) all patients	11 (4-22)	13 (9-20)	11 (5-16)	10 (5-16)	9 (4-18)	0.73
PCWP (mmHg) PH patients	13 (5-22)	20 (14-20)	11 (5-16)	10 (6-16)	11 (5-18)	**0.0091**
PH targeted therapies	37	21	55	55	35	0.061
Patient demographics and clinical details						
Height (m)	1.6 (1.5-1.9)	1.7 (1.4-1.9)	1.6 (1.5-1.8)	1.7 (1.4-1.9)	1.6 (1.5-1.9)	**0.0024**
Weight (kg)	63 (39-123)	68 (40-89)	66 (45-110)	66 (42-102)	65 (43-97)	0.99
Age (years)	58 (21-85)	55 (31-77)	60 (25-79)	58 (21-78)	64 (17-78)	0.64
Sex (female, %)	17	50	18	32	11	**0.0091**

Values are median (range) or n (%). Bold denotes statistically significant values ($P < 0.05$). CMR, cardiovascular magnetic resonance; kg, kilogrammes; m, metres; mmHg, millimetre of mercury; mPAP, mean pulmonary arterial pressure; PCWP, pulmonary capillary wedge pressure; PH, pulmonary hypertension; RAP, right atrial pressure; RHC, right heart catheter.

Supplementary Table 10: Post hoc *P*-values for details of haemodynamics and demographics according to CMR phenotype.

Variable	NF-AC vs BVF	NF-AC vs RVF	NF-AC vs NF-LC	NF-AC vs NF-SC	BVF vs RVF	BVF vs NF-LC	BVF vs NF-SC	RVF vs NF-LC	RVF vs NF-SC	NF-LC vs NF-SC
PH details (haemodynamic data from RHC within 1 year of CMR study)										
PH	0.56	**3.8e-07**	0.58	0.86	**0.0092**	0.84	0.56	**8.4e-05**	**1.4e-06**	0.56
mPAP all patients	0.99	**1.1e-07**	0.91	0.69	**0.014**	0.91	0.87	**9.5e-05**	**7.5e-05**	0.91
RAP all patients	0.12	**0.0075**	0.78	**0.023**	0.96	0.12	**0.023**	**0.023**	**0.00063**	0.12
PCWP all patients	0.20	0.59	0.29	0.17	0.17	0.16	0.16	0.59	0.41	0.66
PH targeted therapies	0.54	0.16	0.16	0.96	0.16	0.16	0.65	1.0	0.16	0.16
Patient demographics and clinical details										
Height	**0.040**	0.32	0.074	0.090	**0.040**	0.48	**0.020**	0.090	0.90	**0.026**
Weight	0.96	0.96	0.96	0.96	0.96	0.96	0.96	0.96	0.96	0.96
Age	0.71	0.91	0.91	0.71	0.71	0.71	0.71	0.91	0.73	0.71
Sex	**0.045**	0.79	0.11	0.55	0.14	0.48	**0.042**	0.44	0.55	0.067

Bold denotes statistically significant values ($P < 0.05$).
CMR, cardiovascular magnetic resonance; mPAP, mean pulmonary arterial pressure; PCWP, pulmonary capillary wedge pressure; PH, pulmonary hypertension; RAP, right atrial pressure; RHC, right heart catheter.

Supplementary Table 11: Clinical details and auto-antibody profiles of CMR phenotypes (post hoc *P*-values detailed in supplementary table 12).

Variable	NF-AC (n=128)	BVF (n= 14)	RVF (n= 27)	NF-LC (n= 40)	NF-SC (n= 51)	*P*-value
Clinical details						
Limited (%)	65	42	59	67	66	0.50
Diffuse (%)	34	57	37	32	31	0.50
Overlap syndrome (%)	32	42	29	27	25	0.75
Inflammatory arthritis (%)	10	7	3	10	7	0.93
Sjögren's syndrome (%)	7	0	7	5	7	0.94
SLE (%)	5	7	3	7	1	0.66
Myositis (%)	16	35	18	12	15	0.41
Disease duration (years)	7 (0-47)	12 (2-29)	8 (0-26)	12 (0-45)	11 (0-36)	0.35
Disease severity score	2.0 (1.0-4.0)	3.0 (2.0-4.0)	3.0 (2.0-4.0)	3.0 (1.0-4.0)	2.0 (1.0-4.0)	**<0.001**
Immunomodulatory therapies (%)	69	85	53	62	60	0.22
History of IHD (%)	7	14	14	15	5	0.26
Autoantibody subset, %						
Anticentromere	37	0	23	40	36	**0.020**
Anti-topoisomerase I	21	21	19	17	26	0.89
Anti-RNA polymerase	7	7	11	10	6	0.90
Anti-U3 RNP	6	14	3	12	4	0.35
Anti-PM/Scl	5	0	3	2	4	0.98
Other*	8	14	19	7	8	0.43
ANA+ENA-	33	35	26	37	34	0.94
ANA-	6	8	4	0	9	0.39

* Includes anti–U1 RNP, anti-Th/To, anti-Ku, anti-Jo-1, anti-PL-7, anti-PL-12, anti-SRP1, and anti-TIF1G.

Values are median (range) or n (%). Bold denotes statistically significant values (*P* < 0.05). ANA, antinuclear antibody; CMR, cardiovascular magnetic resonance; ENA, anti-extractable nuclear antigen; IHD, ischaemic heart disease; SLE, systemic lupus erythematosus; SSc, systemic sclerosis/scleroderma.

Supplementary Table 12: Post hoc *P*-values for SSc details and auto-antibody profiles according to CMR phenotype.
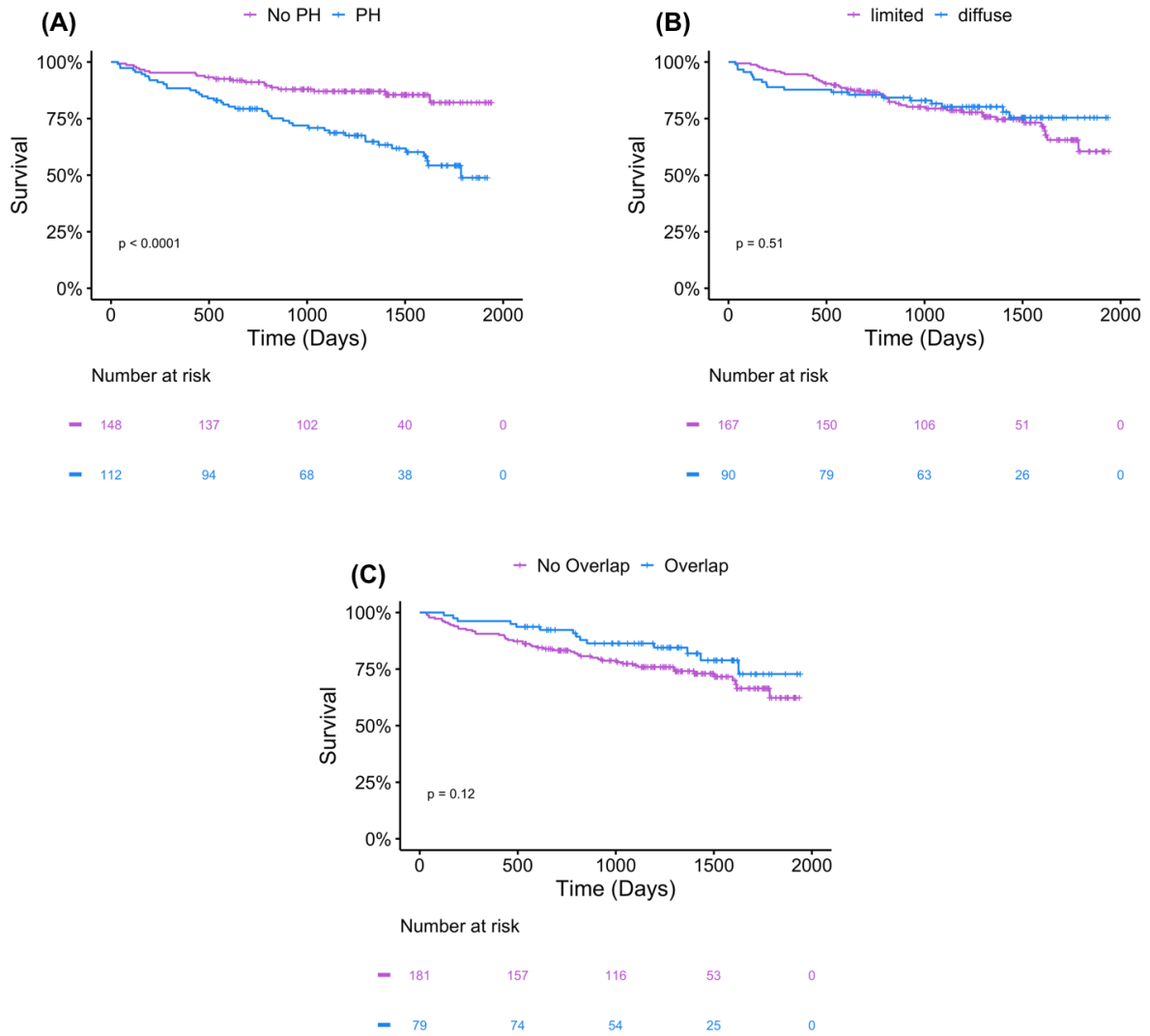
Variable	NF-AC vs BVF	NF-AC vs RVF	NF-AC vs NF-LC	NF-AC vs NF-SC	BVF vs RVF	BVF vs NF-LC	BVF vs NF-SC	RVF vs NF-LC	RVF vs NF-SC	NF-LC vs NF-SC
SSc details										
Limited	0.48	0.94	0.96	0.96	0.86	0.48	0.48	0.94	0.94	1.00
Diffuse	0.47	1.0	1.0	1.0	0.80	0.47	0.47	1.0	1.0	1.0
Overlap syndrome	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0
Inflammatory arthritis	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0
Sjögren's syndrome	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0
SLE	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0
Myositis	0.45	0.87	0.87	1.0	0.67	0.45	0.45	0.87	0.87	0.87
Disease duration	0.57	0.70	0.57	0.57	0.57	0.98	0.98	0.57	0.57	0.98
Disease severity score	**0.0040**	**0.00030**	**0.017**	0.23	0.90	0.23	**0.0066**	0.15	**0.00086**	**0.016**
Immunomodulatory therapies	0.59	0.45	0.63	0.58	0.45	0.45	0.45	0.70	0.70	0.83
History of IHD	0.49	0.49	0.49	1.0	1.0	1.0	0.49	1.0	0.49	0.49
Autoantibody subset										
Anticentromere	**0.021**	0.31	0.95	1.00	0.19	**0.021**	**0.021**	0.31	0.43	0.95
Anti-topoisomerase I	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0
Anti-RNA polymerase	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0
Anti-U3 RNP	0.55	1.0	0.55	1.0	0.55	1.0	0.55	0.65	1.0	0.55
Anti-PM/Scl	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0
Other*	1.0	0.87	1.0	1.0	1.0	1.0	1.0	0.87	0.87	1.0
ANA+ENA-	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0	1.0
ANA-	0.93	1.0	0.86	0.93	1.0	0.86	1.0	0.93	0.93	0.86

* Includes anti–U1 RNP, anti-Th/To, anti-Ku, anti-Jo-1, anti-PL-7, anti-PL-12, anti-SRP1, and anti-TIF1G.
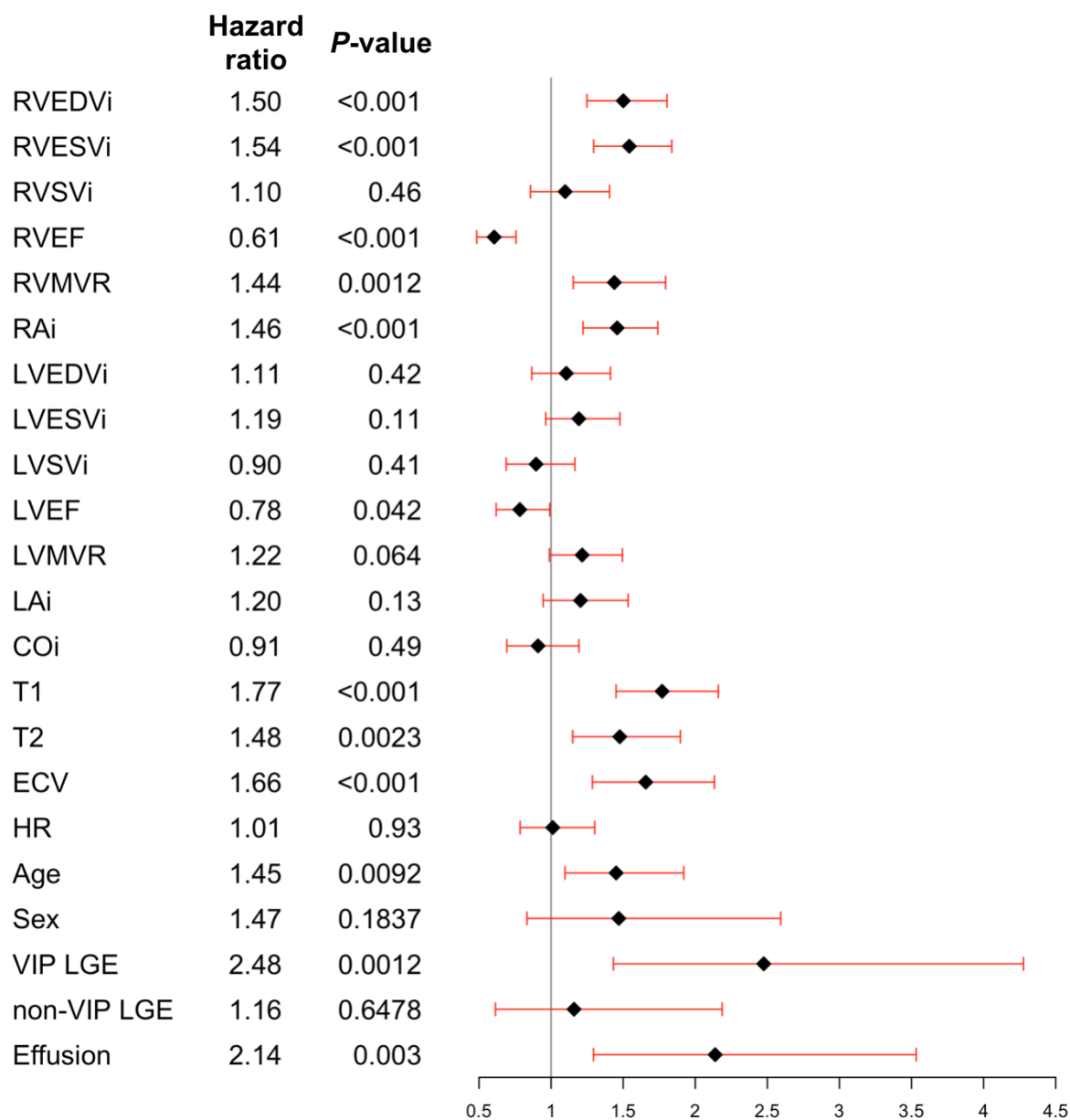Bold denotes statistically significant values ($P < 0.05$).
ANA, antinuclear antibody; CMR, cardiovascular magnetic resonance; ENA, anti-extractable nuclear antigen; IHD, ischaemic heart disease; SLE, systemic lupus erythematosus; SSc, systemic sclerosis/scleroderma.

Supplementary Figure 1: Kaplan-Meier curves for all-cause mortality stratified by: (A) presence or absence of SSc-associated PH; (B) limited versus diffuse cutaneous SSc; (C) presence or absence of overlap syndrome.

Statistically significant values: *P*<0.05.

Supplementary Figure 2: Forest plot summarising simple Cox regression analysis of the association between CMR variables and all-cause mortality.

	Hazard ratio	P-value
RVEDVi	1.50	<0.001
RVESVi	1.54	<0.001
RVSVi	1.10	0.46
RVEF	0.61	<0.001
RVMVR	1.44	0.0012
RAi	1.46	<0.001
LVEDVi	1.11	0.42
LVESVi	1.19	0.11
LVSVi	0.90	0.41
LVEF	0.78	0.042
LVMVR	1.22	0.064
LAi	1.20	0.13
COi	0.91	0.49
T1	1.77	<0.001
T2	1.48	0.0023
ECV	1.66	<0.001
HR	1.01	0.93
Age	1.45	0.0092
Sex	1.47	0.1837
VIP LGE	2.48	0.0012
non-VIP LGE	1.16	0.6478
Effusion	2.14	0.003

bpm, beats per minute; CO, cardiac output; ECV, extracellular volume; EDV, end-diastolic volume; EF, ejection fraction; ESV, end-systolic function; HR, hazard ratio; i, indexed for body-surface area; LA, left atrial; LGE, late gadolinium enhancement; LV, left ventricular; M, mass; RA, right atrial; RV, right ventricular; SV, stroke volume; VIP, ventricular insertion point.

Hazard ratios are reported per standard deviation change in metric for continuous data. Statistically significant values: *P*<0.05.