

Supplementary Table S1. Definitions of SSc-related organ involvement.

Gastrointestinal involvement	Gastrointestinal involvement is defined as gastrointestinal involvement documented by either imaging or functional study and/or patient-reported oesophageal symptoms (dysphagia, reflux) or stomach symptoms (early satiety, vomiting) or intestinal symptoms (diarrhoea, bloating, constipation) or malabsorption syndrome or paralytic ileus occurring concurrently with other disease symptoms and not explained by another cause (1).
Interstitial lung disease	Interstitial lung disease (ILD) evidenced by ground-glass opacities, honeycombing, reticular abnormalities, or traction bronchiectasis on high-resolution computed tomography (HRCT) of the chest.
Pulmonary hypertension and pulmonary arterial hypertension	Pulmonary hypertension defined by a mean pulmonary artery pressure (mPAP) at rest greater than 20 mmHg, and pulmonary arterial hypertension characterized by a pulmonary artery wedge pressure (PAWP) of ≤ 15 mmHg and pulmonary vascular resistance (PVR) > 2 Wood units (WU) on right heart catheterisation (2).
SSc-associated primary heart involvement (SSc-pHI)	Patients with clinical symptoms and an ejection fraction of $< 50\%$ on echocardiography were considered to have primary heart involvement (3).
Scleroderma renal crisis	Scleroderma renal crisis (SRC) was defined based on the most recently validated definition by Hudson <i>et al.</i> (4) A. Hypertensive SRC: Requires both A1 and A2 to be fulfilled. A1. New-onset hypertension is defined as any of the following: 1. Systolic blood pressure ≥ 140 mmHg, or 2. Diastolic blood pressure ≥ 90 mmHg, or 3. Increase in systolic blood pressure by ≥ 30 mmHg from baseline, or 4. Increase in diastolic blood pressure by ≥ 20 mmHg from baseline AND A2. Any one of the following five features: 1. Increase in serum creatinine by $\geq 50\%$ from baseline OR serum creatinine $\geq 120\%$ of the upper limit of normal for the local laboratory 2. Proteinuria: $\geq 2+$ on dipstick, confirmed by a protein-to-creatinine ratio exceeding the upper limit of normal 3. Haematuria: $\geq 2+$ on dipstick or ≥ 10 red blood cells per high-power field (HPF), not due to menstruation 4. Thrombocytopenia: platelet count $< 100,000/\text{mm}^3$ 5. Haemolysis, as evidenced by: ◦ Red blood cell fragments (schistocytes) on blood smear, or ◦ Increased reticulocyte count 6. Hypertensive encephalopathy B. Normotensive SRC: Requires both B1 and B2 to be fulfilled. B1. Either of the following: 1. Increase in serum creatinine by $\geq 50\%$ from baseline OR 2. Serum creatinine $\geq 120\%$ of the upper limit of normal for the local laboratory AND B2. Any one of the following five features: 1. Proteinuria: $\geq 2+$ on dipstick, confirmed by protein-to-creatinine ratio exceeding the upper limit of normal 2. Haematuria: $\geq 2+$ on dipstick or ≥ 10 RBCs/HPF, not due to menstruation 3. Thrombocytopenia: platelet count $< 100,000/\text{mm}^3$ 4. Haemolysis is defined as anaemia, not due to other causes, with either: ◦ Schistocytes or other red cell fragments on blood smear ◦ Increased reticulocyte count 5. Renal biopsy findings consistent with scleroderma renal crisis (<i>i.e.</i>, microangiopathy)

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2. HUMBERT M, KOVACS G, HOEPER MM *et al.*: 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Respir J* 2023; 61(1). <https://doi.org/10.1183/13993003.00879-2022>
3. BRUNI C, BUCH MH, FURST DE *et al.*: Primary systemic sclerosis heart involvement: A systematic literature review and preliminary data-driven, consensus-based WSF/HFA definition. *J Scleroderma Relat Disord* 2022; 7(1): 24-32. <https://doi.org/10.1177/23971983211053246>
4. HUDSON M, BARON M, TATIBOUET S *et al.*: Exposure to ACE inhibitors prior to the onset of scleroderma renal crisis-results from the International Scleroderma Renal Crisis Survey. *Semin Arthritis Rheum* 2014;43(5):666-72. <https://doi.org/10.1016/j.semarthrit.2013.09.008>