Systemic Sclerosis — EULAR Treatment

Organ-based recommendations covering Raynaud's/digital ulcers, pulmonary arterial hypertension, renal crisis, GI involvement, skin fibrosis, interstitial lung disease, and poor-prognosis disease.

2023 EULAR Update
Recommendations for the Treatment of Systemic Sclerosis
Del Galdo et al. · Ann Rheum Dis 2024

SOR — A · Strongest Evidence | B | C | D · Weakest Evidence

1 Raynaud's Phenomenon & Digital Ulcers

A 1a	**SSc-RP:** dihydropyridine calcium antagonists (usually oral nifedipine) first-line; PDE5 inhibitors also considered; IV iloprost for severe RP after oral therapy fails.
A 1a	**Digital ulcers:** PDE5 inhibitors and/or IV iloprost for treatment; **bosentan** to reduce number of new ulcers.

2 Pulmonary Arterial Hypertension

A 1a	**First-line:** combination of PDE5 inhibitor + endothelin receptor antagonist (ERA) at diagnosis of SSc-PAH.
A 1a	**Advanced (WHO class III–IV):** IV epoprostenol; other prostacyclin analogues/agonists (B, 1b) or riociguat (B, 1b) also considered.
C 2a	Anticoagulation (warfarin) for SSc-PAH is **not recommended**.

3 Scleroderma Renal Crisis

C 4	**ACE inhibitors** should be used immediately at diagnosis of scleroderma renal crisis.
C 3	Patients on **glucocorticoids** should have regular blood pressure monitoring to detect renal crisis.

4 Gastrointestinal Involvement

B 3	**PPIs** for SSc-GERD and prevention of esophageal ulcers/strictures.
C 1b	**Prokinetics** for symptomatic motility disturbances; **rotating antibiotics** (D, 2b) for small intestinal bacterial overgrowth.

5 Skin Fibrosis & Musculoskeletal Involvement

A/B 1a-b	**Skin fibrosis:** methotrexate, mycophenolate mofetil, and/or rituximab; **tocilizumab** (C, 1b) considered for early, inflammatory diffuse cutaneous SSc.
D 2b	**Musculoskeletal involvement:** methotrexate should be considered.

6 Interstitial Lung Disease & Poor-Prognosis Disease

A 1a	**SSc-ILD:** mycophenolate mofetil, cyclophosphamide, or rituximab; **nintedanib** alone or with MMF; **tocilizumab** (B, 1b) also considered.
A 1a	**Poor prognosis:** high-intensity immunosuppression (usually with cyclophosphamide) followed by **autologous HSCT** for selected early diffuse cutaneous SSc without advanced cardiorespiratory involvement.

Format note: Strength of Recommendation (SoR): A = strongest → D = weakest, per LoE (Oxford CEBM). Recommendations marked new/substantially revised vs. 2017 update include most PAH, skin, and ILD items. Condensed from Table 1.

Del Galdo F, Lescoat A, Conaghan PG, et al. EULAR recommendations for the treatment of systemic sclerosis: 2023 update. *Ann Rheum Dis.* 2024.