



# SYSTEMIC SCLEROSIS

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Scleroderma may present as a limited disease affecting only the skin and nearby tissues (**morphea**) or it can present as a systemic disease with internal organ involvement. Systemic sclerosis (SSc) can be categorized into diffuse and limited cutaneous sclerosis determined by how extensive the skin involvement is.

The characteristics of SSc include Raynaud's phenomenon which is a vasospastic disorder causing discoloration of the fingers and toes due to an abnormality in circulation. In addition the musculoskeletal, renal, pulmonary, cardiac and gastrointestinal systems are all involved with fibrotic or vascular problems. Patients will present with fatigue, arthralgia, myalgia, pain, and skin discoloration. The skin involvement is always present in SSc with variable extent and severity of skin thickening and hardening. The fingers, hands, and face are generally the earliest areas of the body involved. Edematous swelling and erythema may precede atrophy of the skin, also termed morphea. Pruritus and edema in the early stages are evident, sclerodactyly, digital ulcers, pitting at the fingertips, telangiectasia and calcinosis cutis are all features of systemic sclerosis.

Patients with limited cutaneous SSc present with skin sclerosis on the hands, the distal forearm, face and neck. They often also present with severe Raynaud's phenomenon and cutaneous telangiectasia and can exhibit manifestations of **CREST syndrome** which includes calcinosis cutis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia.

Patients with diffuse cutaneous SSc have sclerotic skin on different areas of the body than those with limited cutaneous SSc. The location of sclerotic skin is usually on the chest, abdomen, upper arms or shoulders and patients with this diffuse SSc are more likely to have or to manifest severe internal organ damage due to ischemic injury or fibrosis than those with limited cutaneous SSc.

The skin involvement can be diagnosed with a scoring system that estimates skin thickness, hardness, and fixation to underlying structures. The scoring system is called the Rodnan skin score. This semi-quantitative score rates the severity of the above findings from 0 which is normal, to 3 which is the most severe, in 17 separate areas of the body.

Treatment of SSc varies according to each individual's internal organ involvement and skin manifestations. For pruritus, camphor and menthol as well as **PUVA** light treatment may be used. For patients with calcinosis, surgery can help and in patients with Raynaud's phenomenon the most effective means of treatment is to avoid cold temperatures and smoking cessation. In terms of pharmacologic treatment, calcium channel blockers and intravenous prostaglandins can be used. In patients with GI involvement a proton pump inhibitor and H2 blockers can help. For those with lung involvement cyclophosphamide, calcium channel blockers and prostaglandins have been used with variable success. In terms of renal disease, an ACE inhibitor or angiotensin II inhibitor have shown to be helpful. In addition, **mycophenolate mofetil** has been shown to benefit patients with diffuse progressive cutaneous systemic sclerosis.

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