SYSTEMIC SCLEROSIS
A rare, progressive, immune-mediated disease
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Reported incidence between 3.7 and 23 cases per million

Reported prevalence of 2 individuals per 10,000

Strong sex bias with a ratio of 3:1 more women than men affected

Mortality rate is greater than in any other rheumatic disease

SYSTEMIC SCLEROSIS CLINICAL SUBTYPES
A rare, progressive, immune-mediated disease

Without skin involvement (<5% patients)

SYSTEMIC SCLEROSIS SINE SCLERODERMA

With skin involvement (≥95% patients)

LIMITED CUTANEOUS SYSTEMIC SCLEROSIS
DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS

SYSTEMIC SCLEROSIS OVERLAP SYNDROME
Manifestations of 1 of the 3 subtypes, with clinical and investigational features of another rheumatic disease

SYSTEMIC SCLEROSIS SINE SCLERODERMA
An infrequent subtype with internal organ but no skin involvement

These images depict examples of some of the manifestations observed in patients with systemic sclerosis. This is for educational purposes only and should not be used as the basis for systematic assessment and follow-up. Patient clinical presentations may vary.
Raynaud’s phenomenon is a common feature of systemic sclerosis sine scleroderma.

SYSTEMIC SCLEROSIS SINE SCLERODERMA
An infrequent subtype with internal organ but no skin involvement

Digital ulcers are common.


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SYSTEMIC SCLEROSIS SINE SCLERODERMA
An infrequent subtype with internal organ but no skin involvement

Clinical presentation

Capillary abnormalities typical of systemic sclerosis, observed in the cuticles.


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Pulmonary arterial hypertension (PAH) is frequent.


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Thrombotic microangiopathy in the kidney can lead to renal crisis.

Anticentromere autoantibodies are associated with pulmonary arterial hypertension. Anti-Scl-70 (anti-DNA-topoisomerase I) autoantibodies are associated with digital ulcers.

LIMITED CUTANEOUS SYSTEMIC SCLEROSIS

A subtype with restricted skin but major internal organ involvement

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Typically there is long history of Raynaud’s phenomenon.

Sclerodactyly occurs late and is limited to fingers and toes. Extensive calcinosis cutis may occur.


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**LIMITED CUTANEOUS SYSTEMIC SCLEROSIS**

A subtype with restricted skin but major internal organ involvement

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**Clinical presentation**

Facial and mucosal membrane telangiectasias are a common clinical presentation.


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Esophageal dysmotility, which results in gastroesophageal reflux, is a frequent gastrointestinal feature.


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LIMITED CUTANEOUS SYSTEMIC SCLEROSIS
A subtype with restricted skin but major internal organ involvement

Clinical presentation

Severe pulmonary arterial hypertension (PAH) is frequent.


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Anticentromere autoantibodies are detected in 70%–80% of patients and are associated with a high risk of pulmonary hypertension. Anti-Th/To autoantibodies have also been detected in a subset of patients.

DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS
A subtype with extensive internal organ and skin involvement

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Typically there is a short history of Raynaud’s phenomenon.


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Early distal skin thickening can progress to involve proximal extremities as well as the trunk.

- Peak skin involvement occurs at 12–18 months
- Skin thickness may decrease late in the disease

Digital ulcers may be present.

25%–30% of patients develop progressive interstitial lung disease.

Early gastric antral vascular ectasia (watermelon stomach) may occur.


Gastrointestinal complications in the small and large intestine, including blind loop syndrome (small intestinal bacterial overgrowth), malabsorption syndrome, and intestinal pseudo-obstruction, are common.


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Anti-Scl-70 (anti-DNA topoisomerase I) autoantibodies are associated with diffuse systemic sclerosis, progressive lung fibrosis, digital ulcers, and hand disability. Anti-RNA polymerase III autoantibodies are associated with diffuse systemic sclerosis, scleroderma renal crisis, and hand disability.


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