

▶ Serological Markers for Systemic Sclerosis

The most accepted subsets of Systemic Sclerosis (SSc), based on clinical and laboratory features, are diffuse cutaneous disease (dcSSc), limited cutaneous disease (lcSSc) and CREST.

▶ Diffuse Cutaneous SSc (dcSSc)

- ▶ Onset of Raynaud's phenomenon within one year of skin changes (puffy or hidebound)
- ▶ Truncal and acral skin involvement
- ▶ Presence of tendon friction rubs
- ▶ Early and significant incidence of interstitial lung disease, oliguric renal failure, diffuse gastrointestinal disease and myocardial involvement
- ▶ Absence of anti-centromere antibody
- ▶ Nailfold capillary dilatation and capillary destruction
- ▶ Anti-Scl-70 antibody positive (30% of patients).

▶ Limited Cutaneous Ssc (lcSSc)

- ▶ Raynaud's phenomenon for years (occasionally decades)
- ▶ Skin involvement limited to hands, face, feet and forearms (acral) or absent
- ▶ A significant late incidence of pulmonary hypertension with or without interstitial lung disease, trigeminal neuralgia, skin calcifications, telangiectasia and liver disease
- ▶ A high incidence of anti-centromere antibody (70-80%)
- ▶ Dilated nailfold capillary loops, usually without capillary dropout.

Anti-Nuclear Antibodies: More than 95% of patients with Scleroderma

Anti-Centromere Antibodies: Primarily in patients with lcSSc; a useful marker for a mild variant of SSc called CREST Syndrome. The antibodies recognize primarily centromere B, a major centromere protein. Prevalence: 40-98%

Anti-Scl 70 Antibodies: High proportion of individuals with dcSSc. Increased skin thickness and renal involvement. Prevalence: 15%

Anti-U1 RNP: Associated with SSc as overlap with SLE or polymyositis (Mixed Connective Tissue Disease [MCTD]). Prevalence: 8%

Anti-Th/To: Relatively specific to lcSSc, associated with pulmonary hypertension and pulmonary fibrosis. Prevalence: 4-10% of all SSc, 8-19% for lcSSc, 5% for dcSSc and 3% for Raynaud's Syndrome

Anti-U3 RNP (Fibrillar): High frequency among African Americans with SSc and associated with skeletal and cardiac muscle disease and pulmonary arterial hypertension. Prevalence: 6-8% of all SSc, 5% in dcSSc, 10% in CREST

Anti-PM/Scl: Subset of patients with high prevalence of skeletal myositis. Prevalence: 8%

Anti-RNA Polymerase III: Nucleolar or speckled ANA pattern, specific for SSc. Prevalence: 20%

- ▶ **Anti-RNA Polymerase III and Anti-Fibrillar are usually associated with a worse prognosis.**
- ▶ **Anti-PM/Scl and Anti-Th/To are usually associated with a better prognosis.**

See reverse side for additional information



Serologic Markers of Scleroderma



▶ Good Prognosis

- ▶ **Anti-Centromere Antibody**
Limited Disease Pulmonary Hypertension
Caucasian Ethnicity
- ▶ **Anti-U1 RNP Antibody**
Overlap Syndromes
African American Ethnicity
- ▶ **Anti-PM/Scl Antibody**
Polymyositis Overlap
Limited Skin Disease
Caucasian Ethnicity

▶ Poor Prognosis

- ▶ **Anti-Topoisomerase I (Scl-70) Antibody**
Diffuse > Limited Skin Disease
Pulmonary Fibrosis
Caucasian, Hispanic and African American Ethnicity
- ▶ **Anti-Th/To Antibody**
Diffuse > Limited Skin Disease
Pulmonary Hypertension
Pulmonary Fibrosis
- ▶ **Anti-U3 RNP (Fibrillarin) Antibody**
Diffuse > Limited Skin Disease
- ▶ **Anti-RNA Polymerase III**
Diffuse Skin Disease
Pulmonary Hypertension
Renal Crisis