SYSTEMIC SCLEROSIS, SCLERODERMA

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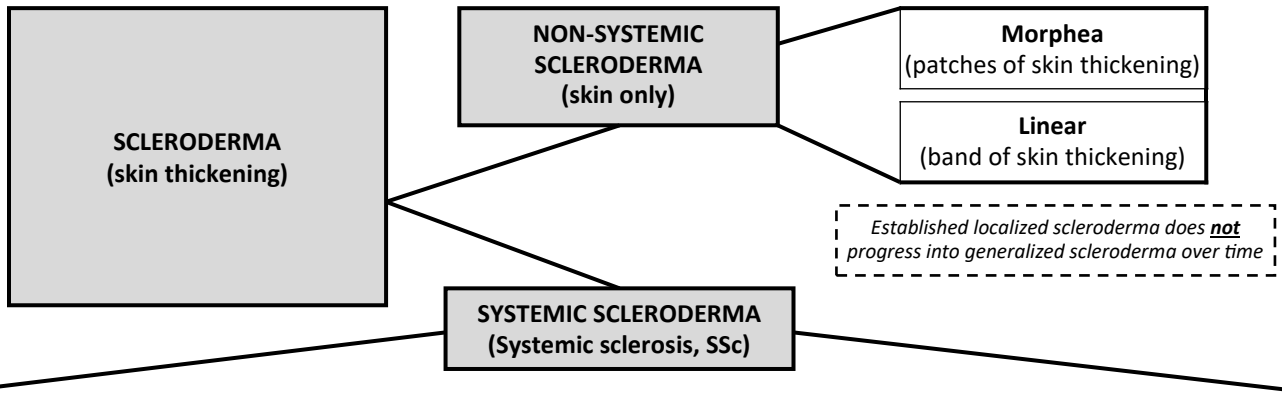
CLINICAL FRAMEWORK OF SYSTEMIC SCLEROSIS, SCLERODERMA



Limited cutaneous SSc (lcSSc)	Diffuse cutaneous SSc (dcSSc)
Puffy fingers leading to skin thickening hands, face, neck (to clavicles), feet but stays **distal** to elbows and knees.	Puffy fingers leading to skin thickening spreading **proximal** to elbows/knees (i.e.: "diffuse")
Slow onset: 5-10 years between new/worsening Raynaud and systemic manifestations Early disease: <10 years; Late disease: >10 years	**Fast** onset: ≤1 year between new/worsening Raynaud and systemic manifestations Early disease: <3 years; Late disease: >3 years
Prominent vascular manifestations (Raynaud, PAH, telangiectasias), "CREST" (calcinosis, Raynaud, esophageal dysmotility. Sclerodactyly, telangiectasias).	Can be rapidly progressive skin thickening. More arthralgia and tendon friction rubs in early disease. More systemic symptoms in early disease.
Low risk for renal crisis. Lower risk ILD early; can get ILD later, especially if anti-SCL70+ Higher risk PAH in late disease, especially if centromere ANA	Higher risk renal crisis (especially early and RNA polymerase III) Higher risk ILD early, especially if anti-SCL70+
Anti-centromere: Pulmonary arterial hypertension (PAH) Anti-RNP (anti-U1-ribonucleoprotein): Mixed CTD Anti-Pm-Scl: Overlap myositis Anti-Th/To: Interstitial lung disease (ILD), PAH Anti-RNA Polymerase III: GAVE	Anti-Scl70 (anti-topoisomerase I): Progressive ILD Anti-RNA Polymerase III: Scleroderma renal crisis, malignancy Anti-Fibrillarin (Anti-U3-ribonuceloprotein): PAH, myositis

Systemic sclerosis *sine* scleroderma

- Definition: Internal organ manifestations of scleroderma, no significant skin thickening, positive SSc antibody. Rare.
- May not be a truly distinct SSc subset and just a mild form of lcSSc: similar clinical and antibody profile to lcSSc
- Ex: Raynaud, dysphagia with esophageal dysmotility , ILD with SCL70+
- Occasionally, dcSSc patients may present with organ involvement first followed shortly after by skin thickening
- Ex: Raynaud and progressive ILD, followed weeks/months later with diffuse skin thickening

Classification of patients as lcSSc or dcSSc predict patients' expected pattern of organ involvement and disease course.

- ***"CREST" is itself not a diagnosis***; ***preferred terminology is lcSSc***. "CREST" describes a ***subset*** of lcSSc patients with combination of **C**alcinosis, **R**aynaud, **E**sophageal dysmotility, **S**clerodactyly, and **T**elangiectasia.
- Systemic sclerosis can also occur as an overlap syndrome with another systemic rheumatic disease (ex: RA, Sjogren's, SLE).

"The 15% Rule" in Scleroderma: The Frequency of Severe Organ Complications in SSc is around 5% or 15%			
Severe Organ Involvement		**Frequency, %**	**95% CI**
Renal Crisis	In dcSSc	12	5-19
	In SSc overall	3	3-4
Severe Cardiac	CHF, pericarditis, arrhythmia, cardiomyopathy in SSc	15	6-24
	Diastolic dysfunction in SSc	16	14-17
	Clinical CHF	5	1-9
	Arrythmia/conduction abnormality needing treatment	17	14-21
	Pericarditis: symptomatic or mod-large pericardial effusion	3	1-5
	Cardiomyopathy	3	1-6
Pulmonary hypertension	PH in SSc (suggestive via echo)	14	8-20
	PAH in SSc (via right heart catheterization)	15	12-17
	PH with pulmonary fibrosis in SSc	11	4-19
Pulmonary restriction	FVC % predicted <70%	15	12-17
Musculoskeletal	Myositis	13	10-17
	Inflammatory arthritis	12	9-16
Severe Raynaud	Prevalent digital ulcer in SSc	15	10-20
	Current digital ulcer (if one is present at any time)	2	1-3
	Digital amputation in SSc	12	8-16
	Hospitalization due to digital ischemic complication	13	6-21

CLASSIFICATION CRITERIA: ACR/EULAR 2013 CRITERIA

Classification criteria are not meant as diagnostic criteria to diagnose disease in a single specific patient. Classification criteria are a standardized way of recruiting a well-defined homogenous population of patients in research studies in order to ensure comparability across studies of a heterogenous disease.

Criteria are only to be applied if SSc is suspected.
A score of **9 or more** is strongly associated with a diagnosis of SSc

ITEM	SUB-ITEM	WEIGHT
Skin thickening of the fingers of both hands extending proximal to the meta-carpophalangeal joints *(sufficient criterion)*	-	9
Skin thickening of the fingers *(only count the higher score)*	Puffy fingers	2
	Sclerodactyly of the fingers (distal to the metacarpophalangeal joints but proximal to the proximal interphalan-geal joints)	4
Fingertip lesions *(only count the higher score)*	Digital tip ulcers	2
	Fingertip pitting scars	3
Telangiectasia	-	2
Abnormal nailfold capillaries	-	2
Pulmonary arterial hypertension and/or interstitial lung disease *(maximum score is 2)*	Pulmonary arterial hypertension	2
	Interstitial lung disease	2
Raynaud phenomenon	-	3
SSc-related autoantibodies (anticentromere, anti-topoisomerase I [anti-Scl-70], anti-RNA polymerase III) *(maximum score is 3)*	Anticentromere Anti-topoisomerase I Anti-RNA polymerase III	3

Adding the maximum weight (score) in each item category. Patients with a total score of ≥9 are classified as having definite SSc.

Criteria are not applicable to patients with skin thickening sparing the fingers or patients with scleroderma mimic