

Systemic Sclerosis

1980 CRITERIA FOR THE CLASSIFICATION OF SYSTEMIC SCLEROSIS

Table 2. Glossary of clinical terms used in description or classification of systemic sclerosis

1. Typical sclerodermatous skin changes: tightness, thickening, and non-pitting induration, excluding the localized forms of scleroderma (morphea or linear scleroderma)

1. Sclerodactyly: above-indicated changes limited to (fingers and toes)
2. Proximal scleroderma: above-indicated changes proximal to the metacarpophalangeal or metatarsophalangeal joints, affecting other parts of the extremities, face, neck, or trunk (thorax or abdomen); usually bilateral, symmetrical and almost always *including* sclerodactyly

2. Other skin manifestations attributable to systemic sclerosis or comparison disorders

1. Digital pitting scars or loss of substance from the finger pad: depressed areas at tips of digits or loss of digital pad tissue as a result of digital ischemia rather than trauma or exogenous causes
2. Bilateral finger or hand edema: firm but pitting edema, especially involving fingers (includes puffy sausage-like swelling of fingers) or the dorsal aspect of the hands
3. Abnormal skin pigmentation: hyperpigmentation often containing areas of punctate or patchy hypopigmentation or depigmentation ("pepper and salt")
4. Raynaud's phenomenon: at least two-phase color change in fingers and often toes consisting of pallor, cyanosis, and/or reactive hyperemia in response to cold exposure or emotion, as determined by patient's history or physician's observation

3. Visceral manifestations

1. Bibasilar pulmonary fibrosis: bilateral reticular pattern of linear or lineonodular densities which are most pronounced in basilar portions of the lungs on standard chest roentgenogram; may assume appearance of diffuse mottling or "honeycomb lung," and should not be attributable to primary lung disease
2. Lower (distal) esophageal dysphagia: substernal discomfort on swallowing or sensation of food holdup in the retrosternal location
3. Lower (distal) esophageal dysmotility: hypoperistalsis or aperistalsis, as demonstrated by either cine esophagram or fluoroscopy or by manometric study, often accompanied by evidence of decrease in lower esophageal sphincter tone with reflux of gastric contents into the esophagus
4. Colonic sacculations: wide-mouthed diverticula of colon located along the antimesenteric border; found on barium enema examination; these sacculations may also occur in ileum and jejunum

* The text explains the derivation of this "combination" variable which was not on the SCCS form.

Subcommittee for Scleroderma Criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. Preliminary criteria for the classification of systemic sclerosis (scleroderma). Arthritis Rheum 1980;23:581---90.