# SYSTEMIC SCLEROSIS

An Illustrated Guide to Manifestation and Management in Asian Skin



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## Classification and clinical features

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#### CLASSIFICATION

The clinical picture of systemic sclerosis (SSc) may be variable at presentation. Some patients may present at the initial phase of the disease evolution with minimal skin sclerosis or only Raynaud's phenomenon, whereas others present at a later stage with the unmistakable florid, clinical picture. The classification system for SSc categorizes the entity into two subsets. The basis of this categorization is the extent of fibrosis on extremities; the limited cutaneous SSc (ISSc) and diffuse cutaneous SSc (dSSc). This categorization is of immense importance in terms of systemic involvement and disease outcome; discussed in subsequent sections. The assessment of cutaneous sclerosis is done by pinching a fold of skin on the distal extremities with

two fingers of the examiner's hand (Figure 3.1) and the extent upwards is assessed.

### Limited cutaneous SSc

The cutaneous sclerosis is limited to the distal extremities beyond elbows and knees, and the face is involved. Trunk is spared in this subtype of the disease. A subset of patients may have sclerosis of the fingers alone for a long duration. Systemic involvement is present.

#### Diffuse cutaneous SSc

The skin sclerosis progresses proximal to the elbows, knees, and on the trunk.¹ Facial involvement may be absent in the initial stage of the disease. Systemic