Systemic sclerosis (scleroderma) patients should be classified as having one of three variants: diffuse scleroderma; limited scleroderma; or scleroderma in overlap. A number of clinical features are useful in these distinctions, including the following:

1. **Diffuse scleroderma**: rapidly progressive skin thickening; scleroderma affecting the upper extremities proximal to the elbows and/or the trunk; palpable tendon friction rubs; and serum anti-Scl 70 antibody

2. **Limited scleroderma**: skin changes limited to the distal extremities and changing very slowly over time; prominent telangiectasias and calcinosis; and serum anticentromere antibodies

3. **Overlap syndrome**: associated features of polymyositis and/or other connective tissue diseases

The natural history of these subgroups is considerably different. Patients with diffuse scleroderma tend to have rapidly progressive skin thickening with the early appearance of arthritis and joint contractures, pulmonary, myocardial and renal involvement and a reduced survival. In contrast limited scleroderma patients have a slowly evolving disorder which may span several decades before the development of more distinctive visceral sequelae such as pulmonary arterial hypertension or biliary cirrhosis.

There is clinical and laboratory evidence in support of two pathophysiologic processes in systemic sclerosis: vascular and immunologic. In the former case, vasodilators and other vasoactive compounds could be considered reasonable therapeutic choices, whereas in the latter situation, interventions such as corticosteroids and immunosuppressive drugs have been proposed. The new calcium-channel blockers and other recently introduced vasodilators have been helpful in treating the Raynaud's phenomenon and digital ulcerations, but not systemic disease. In the latter case, D-penicillamine has been the most useful agent in our experience and that of others. In comparison to untreated patients with early diffuse scleroderma, those given have shown significant improvement of skin thickening and finger flexion, reduced frequency of a variety of visceral sequelae, most notably the kidney, and improved survival. New, potent antihypertensive agents such as captopril and minoxidil, along with improved techniques of hemodialysis and transplantation, have dramatically changed the outlook for patients developing "scleroderma kidney", formerly a uniformly fatal illness.