Scleroderma/CREST

Diffuse scleroderma, also called progressive systemic sclerosis (PSS), is a generalized disorder of connective tissue characterized by inflammation and scarring of the skin, esophagus, intestinal tract, heart, lungs, and kidneys. Primary biliary cirrhosis can occur. The leading cause of death is renal failure. The cause is obscure. Raynaud's phenomenon occurs in 95% of PSS and can precede other symptoms by years. The course of scleroderma is unpredictable and variable in severity. In disease limited to mild skin involvement, scleroderma is only slowly progressive, and affected individuals may live a normal life span. The prognosis for scleroderma is poorer when cardiac, pulmonary, or renal manifestations are present. The course of the disease may take many years, but some experience a rapid progression to death. Treatment of scleroderma depends on the locale, extent, and severity of the disease and includes aspirin, other non steroidal anti- inflammatory agents, steroids, and penicillamine.

CREST syndrome is scleroderma characterized by:

- **C:** Calcinosis cutis (calcium formation in the tips of the fingers or toes)
- **R:** Raynaud's phenomonen (cold induced blanching of the fingers or toes followed by blue color then redness)
- **E:** Esophageal mobility problems which interfere with swallowing
- **S:** Sclerodactyly (thickening of the skin on the fingers)
- **T:** Telangiectasis (small vessels are seen on the skin)

Limited scleroderma involves the skin exclusively and is characterized by skin that appears tightly drawn, bound, and fixed to underlying structures, especially in the fingers where contractures may occur. The skin on the face may be involved in limited scleroderma. Ulcers may develop over bony prominences and on the finger tips.

Morphea is a localized form of systemic scleroderma that is characterized by the formation of white or pink patches, bands, or lines which are firm. Other manifestations of this disease rarely occur.

Underwriting Considerations for Scleroderma/CREST:

Progressive systemic sclerosis is usually a decline	
Limited scleroderma/CREST	
Within 1 year of diagnosis	Decline
2-5 years of diagnosis	Table D
After 5 years	Table C
Involvement beyond CREST	Decline
Localized scleroderma- morphea or linea	0

Minor credits may be given in the over 65 age group or if there is no disease activity for 5 years.

To get an idea of how a client with Scleroderma/CREST would be viewed in the underwriting process, feel free to use the Ask "Rx" pert underwriter on the reverse side for an informal quote.