

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 phenomenon (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.

If your client has a history of Scleroderma/CREST, please answer the following:

1. Please note type of scleroderma:

localized scleroderma- morphea or linea _____

limited scleroderma/CREST _____

progressive systemic sclerosis- diffuse scleroderma _____

2. Please list date of first diagnosis:

3. Is your client on any medications?

yes, please give most recent test results

4. Please check if your client has had any of the following:

weight loss _____ biliary cirrhosis _____
heart disease _____ liver enzyme abnormality _____
lung disease _____ kidney disease _____
Raynaud's disease trouble swallowing _____

5. Have all polyps been removed?

yes, please give most recent test results

**6. Please note date of last follow-up
colonoscopy:** _____

**6. Has your client smoked cigarettes or any other tobacco products in the last
5 years?**

yes, please give details

7. Does your client have any other major health problems (ex: cancer, etc.)?

yes, please give details
