Scleroderma (Systemic Sclerosis)

Scleroderma is a continual (*chronic*) disease that keeps getting worse (*progressive*). It is a disease of the connective tissue. It is generally listed as one of the arthritic or rheumatic diseases. It is a condition leading to hardening and thickening of the skin and connective tissues.

This disease is associated with over-activation of the system which usually protects the body against cancers and invading infections (*immune system*). This causes damage to cells that line small blood vessels, which in turn leads to the over-production of scar tissue.

Scleroderma means "hard skin." It is also known as systemic sclerosis when it also affects other organs in the body. The disease process is caused by the overproduction of collagen. Collagen is a connective tissue. The cause of this overproduction is not known. It affects women four times more frequently than men. It affects adults more than children. In some cases it seems to run in families. It has been caused by toxins and is seen following bone marrow transplants. It cannot be passed to another person (*not contagious*). It varies from mild to life threatening. It can affect almost all organs of the body. It is a disease in which the symptoms may either be:

- Visible, as when the skin is affected.
- Invisible, as when only internal organs are involved.

THERE ARE DIFFERENT KINDS OF SCLERODERMA:

LOCALIZED SCLERODERMA

- More common in children.
- Usually found in only a few places on the skin or in the muscles.
- Rarely, if ever, does localized scleroderma develop into the systemic form of the disease.

SYSTEMIC SCLEROSIS

This may affect the connective tissue in many parts of the body, including:

- Skin.
- Tube which carries food from your mouth to your stomach (*esophagus*).
- Gastrointestinal tract.
- Lungs.
- Kidneys.
- Heart
- Other internal organs.
- Blood vessels.
- Muscles.
- Joints (causing arthritis).

May include one or more of the following:

- Raynaud's Phenomenon (abnormal sensitivity to cold in the arms/hands and legs/feet). This cold sensitivity causes the fingers to become white or blue or red and often painful. This can also be brought on by stress or change in temperature.
- Swelling of the hands and feet.
- Pain and stiffness of the joints.
- Thickening and discoloration of the skin.
- Joint contractures, permanent shortening of muscle tissue.
- Digestive system and gastrointestinal tract problems which lead to:
  - Difficulty swallowing and absorbing the food.
  - Acid reflux.
  - Heartburn.
  - Bloating.
  - Diarrhea.
  - Constipation.
- Sjogren's Syndrome, dry mucus membranes leading to dry eyes and dry mouth.
- Oral, facial and dental problems.
- Kidney, heart, and lung involvement leading to difficulty breathing and high blood pressure.
- Nonspecific symptoms such as extreme fatigue, generalized weakness, weight loss, sleep disturbances, and vague aching of muscles, joints and bones.

- Less commonly, you may get breathlessness, upset stomach, sore eyes, and depression.

DIAGNOSIS

The diagnosis may require seeing:

- An arthritis specialists (*rheumatologists*).
- A skin specialists (*dermatologists*).

Diagnosis of scleroderma is based on clinical history and physical findings. Diagnosis may be delayed in those without significant skin thickening. Laboratory, x-ray and lung (*pulmonary*) function tests determine the extent and severity of internal organ involvement.

Diagnosis may also require blood studies and a variety of specialized tests depending on which organs are affected. Diagnosis may be very difficult, particularly in the early stages before there are significant skin changes. This difficulty is because many of the symptoms are common to, or may overlap with those of other diseases. Symptoms may overlap with other connective tissue diseases such as:

- Rheumatoid arthritis.
- Lupus.
- Polymyositis.

TREATMENT

- The treatment program varies depending on the type and severity of the symptoms.
- There is no known cure for scleroderma. No treatment has been scientifically proven to change the overall course of the disease, although d-penicillamine is commonly used for this purpose and may be of some value.
- To help soften the skin, light therapy, topical corticosteroids, calcipotriol (made from Vitamin D) and methotraxate are used. These medications may be used in combination as well as alone.
- There are a number of effective organ-specific treatments for scleroderma. These treat the symptoms of the disease but do not treat the disease itself.
- Calcium channel blockers may help Raynaud's phenomenon. To help prevent Raynaud’s phenomenon or keep it from getting worse avoid:
  - Cold.
  - Stress.
  - Nicotine.
  - Caffeine.
  - Decongestant medications may help
- Declining kidney (*renal*) function and high blood pressure (*hypertension*) may be treated with drugs.
- Esophageal damage from reflux of stomach contents can be treated with acid-reducing drugs.
- Antibiotics, special diets and medication can improve absorption of nutrients in people who have abnormalities of their intestines.
- Musculoskeletal pain may respond to nonsteroidal anti-inflammatory agents.
- Lung inflammation may be treated with cyclophosphamide.

Your caregiver will help decide with you what treatment will be best for you.

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