# (Progressive Systemic Sclerosis)



# **BASIC INFORMATION**

#### DESCRIPTION

A widespread connective tissue disease in which the skin and other body parts gradually degenerate, thicken and become stiff. It can involve the skin; joints; digestive system, especially the esophagus; heart; kidneys; lungs; blood vessels; fingers; and toes. It affects adults of both sexes, but is more common in women between ages 30 and 50.

#### FREQUENT SIGNS AND SYMPTOMS

- Fingers hardening and thickening of the skin, stiffness, poor circulation, numbness and fingertip ulceration.
- Digestive system swallowing difficulty, poor food absorption, bloating after eating, weight loss, heartburn and a feeling that food sticks in the chest.
- Skin hardening and thickening, especially in the face, which becomes tight and loses its elasticity.
- · Muscle aches.
- · Weakness and fatigue.
- · Joint pain, stiffness and swelling.
- · Anemia.

#### **CAUSES**

Unknown, but may be an autoimmune disorder. The connective tissue (the framework for all body tissues and blood vessels) thickens, becoming stiff and inflexible.

#### **RISK INCREASES WITH**

Unknown.

#### **PREVENTIVE MEASURES**

Cannot be prevented at present.

## **EXPECTED OUTCOMES**

The course of the disorder is variable and unpredictable. It is often slowly progressive and affects the heart, lungs and kidneys.

### **POSSIBLE COMPLICATIONS**

- · Poor wound healing and gangrene.
- · Bleeding tendencies.
- · Heart-rhythm disturbances.
- · Congestive heart failure.
- · Kidney failure.
- · High blood pressure.
- · Lung destruction.



### **TREATMENT**

# **GENERAL MEASURES**

• Diagnostic tests may include laboratory blood tests to detect anemia and measure antibodies; urinalysis to detect red cells in the urine; ECG (electrocardiogram: a method of diagnosing heart diseases by measuring electrical activity of the heart); X-rays of the hands, esophagus and chest; lung function tests; barium enema; and skin biopsy.

- Home care usually. Rarely, hospitalization for heart, lung or kidney complications or for surgical procedure (e.g., on the esophagus).
- Treatment program will vary depending on type and severity of symptoms.
- Because of poor circulation, wear warm clothing, especially socks and gloves. Avoid exposure to extreme cold. Be wary of air conditioning.
- · Protect yourself from burns and cuts.
- Sleep on 2 or 3 pillows, or raise the head of your bed 5 to 8 inches to prevent stomach acid from rolling back into the esophagus.
- Learn biofeedback techniques to increase circulation to the extremities.
- · Don't smoke.
- Seek psychotherapy or counseling to adjust to living with an incurable disease.
- · Use heat to relieve joint stiffness.
- Additional information available from the Scleroderma Association, P.O. Box 910, Lynnfield, MA 01940, (508) 535-6600 or the Scleroderma Federation, 1182 Teaneck Rd., Teaneck, NJ 07666, (201) 837-9826.

## **MEDICATIONS**

- You may take non-prescription antacids to relieve heartburn or indigestion, and aspirin or ibuprofen to relieve muscle aches and joint pain.
- · Use skin lotions, lubricants and bath oil to soften skin.
- Cortisone drugs to relieve inflammatory symptoms, antibiotics to fight infections, or drugs to lower blood pressure may be prescribed.
- Other medications may be prescribed depending on complications.

#### **ACTIVITY**

- · Be as active as your strength permits; avoid fatigue.
- Regular exercise (or movement) can help keep the skin flexible, maintain good blood circulation, and prevent fixed joints.

## DIET

Eat frequent, small meals to minimize bloating, heartburn and gastrointestinal discomfort. A soft diet is sometimes recommended. Use additional fluids to help with swallowing. A dietitian can help plan a nutritious diet.



# **NOTIFY OUR OFFICE IF**

- · You or a family member has symptoms of scleroderma.
- The following occur during treatment:
  Unexplained bruising or bleeding under:

Unexplained bruising or bleeding under the skin. Slow healing of a wound.

Any sign of infection (fever, pus in wound, etc.).