SARAA
South African Rheumatism & Arthritis Association

Scleroderma

What is Scleroderma?

Scleroderma is also known as Systemic Sclerosis. It is an autoimmune disease, where your immune system attacks your healthy cells. The word 'scleroderma' means 'hard skin'. Scleroderma affects the connective tissues of the body .The connective tissues of people with scleroderma have too much of a protein called collagen. Collagen is important to give connective tissue its strength, but excess collagen causes hardening and tightening of the affected area. Many different areas of the body can be affected. There are two types of scleroderma; Localised and Systemic. Localised scleroderma only affects the skin. The Systemic type affects skin, organs, and blood vessels.

What are the causes of Scleroderma?

The exact cause of Scleroderma is not known. It is believed to be a result of genetic factors combined with environmental and immune factors. Silica exposure, use of solvents such as vinyl chloride, benzene, paraffin carbon tetrachloride and radiotherapy may trigger the onset of scleroderma. Certain drugs such as appetite suppressants, carbidopa, bleomycin and cocaine are also thought to cause this disease.

Who gets Scleroderma?

This disease occurs mostly in women between the ages of 35 and 50, although it can affect people at any age. It does occur more frequently in women but there is an increase seen in men who work underground in mines. It is a very rare disease.

What are the symptoms of Scleroderma?

Skin:
Initially you may get swelling of the hands and fingers which extend all the way up the arms, and may involve the feet and legs. There will be a loss of hair and skin creases, and this may also be very itchy. The skin may then become thick, and hard and very dry later on. Sometimes there may be patches of lightening of the skin, as well ulcers on the fingertips. It may happen that it becomes very difficult to close your fingers or extend your elbows later on.

The skin on the face may also be affected, making it difficult to open your mouth widely, and you may develop problems with your gums and teeth,

Blood Vessels:
Raynaud's phenomenon: This term refers to color changes (blue, white and red) that occur in fingers (andsometimes toes), often after exposure to cold temperatures. It occurs when blood flow to the hands and fingers is temporarily reduced. This is one of the earliest signs of the scleroderma and it occurs in more than 90 percent of patients. Raynaud's can lead to finger swelling, color changes, numbness, pain, skin ulcers and gangrene on the fingers and toes. Enlarged red blood vessels on the hands, face and around nail beds (called "telangiectasias")

Lungs: different parts of the lung can be inflamed, causing a dry cough or shortness of breath.

Gut: Gastroesophageal reflux may lead to hoarseness, dysphagia and aspiration pneumonia . Bloating and a feeling of being full after a small meal is also common. Swelling of the tummy due to bacteria growing in the gut can cause diarrhea alternating with constipation.

Joints: sometimes pain and swelling can occur in the joints, and can affect the fingers, wrists, toes or any joint in the body. There can also be muscle weakness causing difficulty brushing your hair, climbing out of a chair and even climbing a few steps.

Hypertension: The blood pressure may suddenly increase due to a problem in the kidney called scleroderma renal crisis. This is very very rare, and is associated with using high doses of cortisone early on in the disease.

Erectile dysfunction: This is a common symptom in males.

How is Scleroderma diagnosed?

Your doctor will take a history and examine you. You will then do some blood tests and may have to go for further tests for the lungs, heart, and kidneys.
Blood tests: Antibodies against certain proteins called Antinuclear antibodies, topoisomerase, and anticentromere are tested, as well blood counts, kidney and liver function tests, and sometimes iron and vitamin B12 levels.

What are the treatment options for Scleroderma ?

Current treatment of systemic sclerosis is directed toward managing complications and provd=iding symptomatic relief. There are many disease modifying drugs being investigated and some are being used at present. Your rheumatologist will discuss these with you.

Skin: use aqueous creams, emollient creams, and avoid strong soaps. For the itchy skin, your doctor may sometimes use antihistamines, or cortisone.
Joint pains, fatigue, fevers: Your doctor may prescribe pain tablets or a mild dose of cortisone. Other organ involvement is not common but when it does occur, your physician or rheumatologist will consider using more aggressive immunosuppressive therapy such as methotrexate, azathioprine, cyclosporin, cyclophosphamide, mycophenolate mofetil, or a biologic called rituximab.

What is the long-term outlook of this disease?

As there is no cure for Sjögren's syndrome, it is natural to feel scared, frustrated, sad and sometimes angry.
The majority of patients with Sjögren's syndrome remain very healthy, without any serious complications. Patients should know that they face an increased risk for infections in and around the eyes and an increased risk for dental problems due to the long-term decrease in tears and saliva. Rarely, patients may have complications related to inflammation in other body systems. If this does occur, your rheumatologist will treat it accordingly.
What else can you do to help your condition?
Stay physically active, eat a healthy diet, stop smoking and reduce stress to help your overall health and wellbeing.

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Useful websites:
www.arthritis.org.za
www.sjogrens.org
www.uptodate.com
www.mayoclinic.com
www.rheuminfo.com