

# Systemic Sclerosis and the Heart

## What is systemic sclerosis?

Systemic sclerosis is a disease in which many parts of the body become permanently thick and hard, and lose their ability to work properly. The part of the body most often affected is the skin. When the disease occurs only in the skin it is called scleroderma. Systemic sclerosis may also affect the joints, muscles, blood vessels, kidneys, and lungs.

## What is the cause?

Systemic sclerosis appears to be an autoimmune problem. This means the body mistakenly attacks its own tissue. Over 90% of people with systemic sclerosis appear to have antibodies in their blood that attack their cells and tissues. Doctors do not know why this happens.

Each organ affected by the disease, including the skin, goes through a sequence of destructive changes: from inflammation, to hardening, and then to complete scarring. In severe cases some organs eventually stop working at all.

## What are the symptoms?

Changes in the skin are among the most common symptoms. The fingers and other patches of skin become hard and numb. These areas of skin may appear smooth and slightly swollen. The skin may become tight, making it hard to move the joints, especially the wrists and elbows. This tightness may occur around the mouth as well.

Another very common symptom is Raynaud's phenomenon. Raynaud's phenomenon can be brought on by cold, emotion, or stress. When this happens, the fingers and sometimes toes, ears, and nose are very sensitive to cold temperatures. They react to cold by turning pale, then blue, and eventually red. They feel tingly or painful. Such episodes are usually short but may last more than an hour.

Other possible symptoms of scleroderma include:

- Trouble swallowing pills or solid foods, such as meat
- Heartburn
- Weakness
- Shortness of breath or trouble taking deep breaths because the skin of the chest has tightened or the lungs are becoming scarred

## How does it affect the heart?

Lung disease caused by systemic sclerosis can result in pulmonary hypertension, which can lead to right heart failure. Heart failure is a buildup of fluid in the lungs and other parts of the body caused by poor pumping by the heart.

Sometimes small scars form in the heart muscle. Over time, these small scars make the heart muscle stiff and less able to squeeze (contract) normally. You may also have abnormal heart rhythms.

The blood vessel scarring of systemic sclerosis may cause a heart attack. Some people with systemic sclerosis get pericarditis, an inflammation of the tissue sac surrounding the heart. When the kidneys are also involved, fluid may build up in the pericardial sac and may require drainage. Systemic sclerosis rarely affects the heart valves.

## **How is it diagnosed?**

Your healthcare provider will ask about your symptoms and do a physical exam. He or she will examine your skin.

Possible tests include:

- Skin biopsy
- Blood tests, such as tests for ANA (antinuclear antibody) and ACA (anticentromere antibody), which look for signs that the body is rejecting its own tissues
- Special X-rays to see how well you can swallow

Other tests may be done to check organs such as your liver, kidneys, heart, or lungs.

## **How is it treated?**

Treatment doesn't cure the disease but helps to ease the symptoms. Treatment may include medicines to relieve pain, slow the rate of skin thickening, suppress the body's immune response, or lower blood pressure. You may need physical therapy for joint problems. You may need dialysis for kidney failure.

Treatment of systemic sclerosis includes frequent checks of your blood pressure. Prompt treatment of any increases in blood pressure with medicine may help prevent or slow damage to the kidneys. It may also help prevent other complications of high blood pressure, such as heart disease and stroke.

Heart failure and heart attacks are treated with medicines and rest. You may need coronary angioplasty or bypass surgery. Drugs called calcium channel blockers may temporarily improve the function of your heart and will often improve the symptoms of Raynaud's phenomenon. Another medicine for Raynaud's is nitroglycerin ointment, which is rubbed onto the skin.

Many drugs are being studied to see if they will prevent or control the effects of scleroderma. Other treatments under study include light therapy (ultraviolet A) and bone marrow transplants (BMT). The goal of BMT is to replace old, problem-causing cells with new, healthy cells. Unfortunately, BMT has some serious side effects and complications.

## **How long do the effects last?**

The course and severity of the disease varies a lot from person to person. Unfortunately, in most cases the symptoms get worse with time. However, much research is being focused on autoimmune diseases. Treatments may be found soon that will help slow or stop the course of systemic sclerosis.

## **How can I help take care of myself?**

- Follow your healthcare provider's instructions for taking prescribed medicines.
- Stay as active as possible with regular exercise or physical therapy, as recommended by your healthcare provider.
- Tell your healthcare provider about any new symptoms right away.
- Avoid overwork and getting overly tired. Get plenty of rest.
- Avoid cold and air conditioning.
- Do not smoke. Avoid being around smoke.
- Ask your provider about heat therapy for stiff joints.
- Avoid fingersticks for blood tests.
- Use lotions, ointments, and bath oils to prevent skin dryness and cracking.
- Learn skills to cope with stress and tiredness, such as meditation and relaxation techniques.

Developed by RelayHealth.

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