

(1) Electrocardiogram (ECG) This records the electrical activity within the heart. It is this electrical activity which co-ordinates the heart beat.

(2) Exercise ECG. Similar to a resting ECG, this test assesses the heart's electrical activity during exercise which may reveal more subtle abnormalities. Occasionally a 24 hour tape of the heart rhythm is needed to detect changes in the heart's electrical activity during a 24 hour period.

(3) Echocardiogram. This technique uses sound waves in a fashion similar to radar to visualise the movement of the heart valves and muscle.

(4) Chest radiograph. This test visualises the heart as well as the lungs.

(5) Heart scans using isotopes such as thallium are also used occasionally to assess heart muscle function.

(6) Cardiac catheterization. During this investigation, a small tube is passed under local anaesthetic, from a vein or artery into the heart where pressures can be measured and if necessary, dye can be injected to visualise the blood vessels within the lung or the coronary arteries which feed the heart muscle.

Patients with a cardiac problem may require regular specialist unit follow up.

Treatment

For many patients the cardiac problem may be mild but for a minority treatment such as anti-anginal drugs e.g. sublingual or GTN spray, isosorbide, nifedipine may be required. If the heart is more severely affected diuretics (water tablets), digoxin, captopril or enalapril may be required. In patients who have pulmonary hypertension (an elevation of the pressure of the blood vessels that supply the lungs) drugs such as calcium channel blockers e.g. diltiazem or iloprost is needed. Please see our leaflet on Pulmonary Hypertension in Scleroderma for more information.

Summary

The symptoms of lung and heart disease due to scleroderma may be similar to those of more common diseases of the lung and heart from which the general population are at risk. These conditions are common in people over 50 and may be the cause of the lung or heart symptoms in a scleroderma patient rather than the symptoms being due to scleroderma itself.

Only by a specialist's careful assessment can these common conditions be distinguished from scleroderma lung or heart disease and the best advice and treatment prescribed.



**Scleroderma
Society**

Supporting people with Scleroderma for over 25 years

Scleroderma of the Lung and Heart

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INTRODUCTION

Scleroderma can affect many organs in the body. Changes in the skin or in the blood vessels supplying the fingers and toes (Raynaud's phenomenon) often bring the patient to their doctor.

Full assessment at this time may reveal changes in other organs even before symptoms are experienced. The lung is very commonly involved in scleroderma but sophisticated tests are often required to identify this at the earliest stage. The heart is less commonly involved.

THE LUNG

Symptoms

Lung scleroderma patients may experience varying degrees of shortness of breath, occasionally with a dry cough. The reason for this is that the lungs have become stiffened by the scleroderma process resulting in a decreased capacity to convey oxygen to the blood. A second potential cause is that the blood vessels which feed the lungs have become affected by a process similar to the Raynaud's phenomenon in the hands and feet. Less commonly the lining around the lung (the pleura) becomes inflamed causing a sharp pain in the chest which is made worse by breathing deeply or coughing.

Investigations

Your doctor may arrange for a number of investigations to assess the lungs:

- (1) Chest X-Ray
- (2) CT Scan of the chest. This test allows

the doctor to obtain a three dimensional picture of your lungs using a computer linked chest x-ray apparatus. The x-rays obtained using this technique provide a much more sensitive picture of the lungs and can often detect mild degrees of inflammation before the routine chest radiograph is abnormal.

(3) Lung function tests. These tests assess how the lungs actually work. They involve breathing in and out of a number of machines which assess the size of the lungs and the capacity of the lungs to transfer oxygen to the blood stream. Occasionally the doctor may want to do lung function tests on exercise. This can, once again, bring out more subtle changes in the lung.

If you are referred to a specialist centre other tests may be carried out. These can include:-

(4) Various isotope lung scans are sometimes employed to try and define the presence or absence of active inflammation of the lungs. These include gallium-scan and DTPA scans.

(5) Bronchoscopy and lavage. This technique involves passing a flexible instrument into the bronchial tubes in order to obtain samples of the inflammatory cell within the lungs by washing them out (lavage).

(6) Lung biopsy. Patients with evidence of lung inflammation may undergo a small surgical procedure to obtain a small sample of the lung. This is necessary in order to gain a complete picture of the degree of inflammation which can be

important in deciding on which form of therapy is best for the individual patient. This procedure is performed under a general anaesthetic.

Following a full assessment, patients are then seen in the respiratory unit at regular intervals and measurements of chest x-ray, lung function, CT and other scans are performed to assess whether there has been any change in the degree of the lung problem.

Treatment

A variety of treatments are available for patients with lung involvement in systemic sclerosis. Many of these are similar to those which would be used purely for the skin aspect of the disease process. Others are more specifically designed to arrest the inflammation in the lung. The most commonly used drugs are low dose corticosteroids and immunosuppressant therapy (treatment which reduces the inflammation of the lungs which contribute to the disability).

None of these treatments is inevitably successful in all patients and, therefore, a number of new drugs are continually being developed and assessed. These include specifically targeted drugs which are being developed on the basis of our increasing understanding of the causes of lung scarring. At present these are in the early laboratory phase of testing but hold promise for the future.

Summary

Although the lung is often involved with scleroderma not everyone will have symptoms suggestive of lung disease. It is important to identify lung disease at the earliest stage so that treatment can be

commenced to attempt to prevent disease progression as occasionally the severity of the disease is such that patients become breathless on slight exertion and even at rest. Regular lung assessment is advisable.

THE HEART

Symptoms

Patients with heart disease due to scleroderma may experience chest pain on exertion (angina) breathlessness or swelling of the ankles. These symptoms reflect the deposition of scleroderma tissue within the small blood vessels which feed the heart tissue and within the muscle itself. This results in the heart being less able to function as a pump and back pressure produces a leakage of fluid into the lungs and dependent areas of the body such as the ankles.

Patients who have a lot of scarring in the blood vessels leading to the lungs, or in the lungs themselves, can cause the right side of the heart to have to work much harder to pump blood through the lungs. This can produce similar features to those which results from the blood vessels supplying the heart muscles being affected.

The heart may also beat irregularly, causing palpitations. If the outer lining of the heart (pericardium) becomes inflamed a pain similar to that of pleurisy may result.

Investigations

Your doctor may recommend the following tests to assess the heart:-