

The Lungs in Scleroderma

Scleroderma of the lung

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 brings the patient to their doctor. Full assessment at this time may reveal changes in other organs even before symptoms are experienced. Up to half of scleroderma patients have evidence of lung disease, but sophisticated tests are often required to identify this at the earliest stage.

How is the lung affected?

The lung can be affected in scleroderma in two ways.

Firstly, damage to the delicate membranes that line the lungs can lead to scarring or fibrosis of lung tissue termed interstitial lung disease, similar to changes in the skin in scleroderma.

Secondly, the blood vessels can become thickened, narrowed and scarred without any other part of the lung becoming affected. This may progress to cause pulmonary hypertension, which usually shows itself from five years onwards in patients with limited cutaneous systemic sclerosis.

Although the lung is often involved in 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.

Occasionally the severity of the disease is such that patients become breathless on slight exertion and even at rest. Regular lung assessment is advisable. Studies of lung disease of either type in systemic sclerosis have suggested that the earlier the disease is detected, the more likely there is to be a good response to treatment.

Lung disease in scleroderma usually has a long, slow progression and patients have to learn to adjust to what they can and can't do. It is advisable to do regular exercises and try to maintain a good basic level of physical fitness.

Pulmonary Hypertension

In pulmonary hypertension (PHT), the blood vessels which supply the lungs narrow in a similar way as they do in the hands and feet in Raynaud's. This makes it harder for the heart to pump blood through the lungs and may eventually lead to heart failure. This is because there is high blood pressure in the blood vessels in the lungs. Blood returns from all over the body to the right side of the heart where the right ventricle pumps it to the lungs, through arteries which branch and rebranch into very small vessels. In the lungs, the blood takes up oxygen, then flows back to the left side of the heart, which pumps it around the body. This abnormally high blood pressure is caused by changes in the small blood vessels in the lungs, resulting in an increased resistance to blood flowing through these vessels. The increased resistance in turn places a strain on the right side of the heart, causing it to work much harder than usual to move adequate amounts of blood through the lungs.

Pulmonary hypertension is a rare condition but occurs in about 1 in 7 patients with scleroderma. It is more commonly seen in patients who have limited cutaneous systemic sclerosis (previously known as the CREST syndrome), but does also occur in the diffuse subset. Common symptoms include shortness of breath or feeling tired, especially on exertion or exercise. Other symptoms include blackouts or a very fast heart beat. Eventually, if left untreated PHT is fatal.

Treatments

New treatments for PHT have been developed and tested over the past 10 years. We now have three broad groups of treatments, prostacyclins (such as iloprost), endothelin antagonists (e.g. bosentan) and PDE5 inhibitors (e.g. sildenafil). The last two groups of treatments are given as tablets and have made the treatment of PHT much more straightforward.

Is exercise advisable?

Because the body works more efficiently when trained, exercise is helpful in scleroderma because it allows the heart and lungs to work more efficiently. Obviously the type of exercise is important and if there is severe lung damage, it is not possible to exercise strenuously, but provided the correct balance is struck, exercise can be extremely helpful in overall management.

Lung Fibrosis

Studies of lung fibrosis in systemic sclerosis have suggested that the earlier the disease is detected, the more likely there is to be a good response to treatment. Fibrosis affects the alveoli, the tiny air sacs in the lungs. These are responsible for transferring oxygen from the air to the blood and removing the waste product, carbon dioxide, from the blood. The cells which normally defend the body against infection, invade the lungs. While the scarring helps the injured areas to heal, it also results in thickening or fibrosing of the alveoli wall. This means that the areas damaged by scleroderma fibrosis lose some of the elasticity and are therefore less able to perform their normal function. The body's cells that normally protect us from infection appear to be involved in the damage. Breathlessness is the most common symptom, which may be noticeable when taking exercise or when walking up hills or upstairs. There may also be a dry irritating cough.

Treatment

It is important to have an early diagnosis so that treatment can start as soon as possible. Treatment will depend on the results of your tests. Steroids may be used or immunosuppressant treatment, which aims to suppress the immune system. This treatment may leave you more susceptible to infection so regular blood tests are important.

Genetic inheritance

It has been suggested that lung fibrosis occurs in individuals who have inherited a susceptibility to this condition. Just as the genes which we inherit determine the colour of our eyes and hair, there is now some evidence that genetic inheritance can predispose to lung fibrosis and PHT in a patient who already has systemic sclerosis.

It is therefore critical to identify those individuals for whom there is an increased likelihood of lung fibrosis, so that treatment may be administered at an early stage in the disease and the fibrotic process may be slowed. Genetic studies are on-going to define these predispositions more precisely.

Screening

The World Health Organisation recommends that all patients with scleroderma should be screened annually for lung complications, even if they have no symptoms.

Investigations

There are a number of investigations to assess for lung fibrosis and PHT:

Chest X-ray

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, which provides a much more sensitive picture of the lungs.

Lung function tests - these tests assess how the lungs actually work and involve breathing in and out of a number of machines to assess the size and capacity of the lungs to transfer oxygen into the blood stream. This can also be done whilst exercising.

At a specialist centre certain other tests may be carried out including:

Isotope lung scans to try and define the presence or absence of active inflammation of the lungs.

Bronchoscopy and lavage - a technique which 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.

Lung biopsy - patients with evidence of lung inflammation may undergo a surgical procedure, performed under a general anaesthetic 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.

ECG and Echocardiogram - a test that uses a type of ultrasound to measure the structure and function of the heart.

Right Heart Catheter - this involves passing a small tube through a vein into the heart to measure directly the pressures in the heart chambers and major blood vessels leading from the heart to the lungs. 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.

The Heart

The heart can be affected in scleroderma both directly and also due to other complications of the disease that put strain on an otherwise normal heart, including scleroderma, renal crisis or pulmonary hypertension. Although many patients with even limited scleroderma have some sort of functional heart problem, most patients, including those with diffuse scleroderma, do not have severe heart involvement. It only affects about 1 in 20 patients with scleroderma, in a similar way to other organs of the body.

Direct involvement of the heart comes in two main forms, one is inflammation of the membrane around the heart which can become inflamed causing a condition called pericarditis, which can be entirely incidental and just be picked up on the electro cardiogram or can cause breathlessness. Sometimes fluid accumulates around the heart (a pericardial effusion) and sometimes this causes problems with the heart's pumping activity. This is quite easily treated by releasing the fluid. Secondly, one can get either inflammation or weakness of the heart muscle due to either the disease itself or the effects of the scleroderma on the small vessels. This is treated in the same way as other conditions for weakness of the heart, with drugs that have been developed and are highly effective. If the heart muscles do not pump efficiently, this can occasionally affect the rhythm of the heart so the heart beat can become irregular.



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