

# The Lungs in Systemic Sclerosis (Systemic Scleroderma)

Systemic sclerosis (systemic scleroderma) is a chronic disease associated with skin thickening and changes to blood vessels, particularly those supplying the fingers and toes. The hardening of the skin is caused by the formation of scar tissue. This occurs because of an increase of collagen, a normal component of tissues, which in systemic sclerosis, is produced in excess. In systemic sclerosis, scar tissue or “fibrosis” can also form in internal organs such as the lungs, the gut, or the kidneys.

## What causes the scarring in systemic sclerosis?

Systemic sclerosis is considered to be an ‘autoimmune disease, - that is it arises because of over-activity of the body’s immune system which leads to excessive inflammation and ultimately results in the development of fibrosis (scar tissue). In health, the main role of the immune system is to protect and defend our bodies from infection (such as with bacteria or viruses) and other damaging substances that cause injury to the body.

However, the immune system can in some individuals occasionally overreact and go “over the top”, attacking the body itself. Why this over-activity of the immune system occurs in individuals with systemic sclerosis is unknown.

One of the important ways that the immune system deals with injury or infection is by causing inflammation. If the immune system is overactive as it is in systemic sclerosis and remains continuously ‘switched on’ then inflammation, rather than being helpful and protective, can instead cause damage to the body. Normally, following injury (e.g. a bad cut) or infection (e.g. pneumonia), inflammation helps the body repair itself by triggering the process that leads to healing.

Following a bad injury, a necessary part of the healing process is scarring. In systemic sclerosis, over-activity of the immune system leads to both excessive and continuous inflammation, which ultimately results in the development of scarring or “fibrosis”.

## What types of lung involvement are found in systemic sclerosis?

The most frequent type of lung involvement is an “interstitial lung disease” (also sometimes referred to as fibrosing alveolitis), in which the walls of the small air sacs (alveoli) that make up the lungs are thickened by a mixture of inflammation and scarring (or fibrosis). Because of the presence of scarring, this type of lung involvement is also called pulmonary fibrosis. When scarred lung is looked at under a microscope it is possible to recognise a number of different patterns of fibrosis. In general terms these different patterns of fibrosis occur in different diseases. In systemic sclerosis the most frequently occurring pattern of fibrosis is one called non-specific interstitial pneumonia (abbreviated as NSIP).

## How frequent is severe lung scarring in systemic sclerosis?

Although lung fibrosis is found in more than half of patients with systemic sclerosis, its degree varies very much from person to person. In the majority of individuals with systemic sclerosis, lung fibrosis is limited in extent, and only causes symptoms when individuals really exert themselves. In these individuals the fibrosis tends to remain stable over time and does not require specific treatment. However, in approximately one out of ten cases, lung fibrosis is more severe and/or has a tendency to worsen with time, because of continued development of scar tissue in the lungs. When this is the case, treatment to prevent fibrosis is required. Only in a very small number of individuals does lung fibrosis progress despite treatment.

## How is lung involvement diagnosed?

Symptoms of lung disease may include cough and shortness of breath, particularly when climbing stairs and walking uphill. However, these symptoms do not necessarily imply the presence of lung disease, as a cough can have other causes, such as chronic heartburn, and breathlessness on exertion can be related to joint and muscle pains. On the other hand, some individuals can be so limited by joint or muscle problems that they do not exert themselves to a degree that causes breathlessness even when they have lung involvement. Furthermore, when the lung disease is limited in extent, it may not be associated with any symptoms. Therefore, tests are needed to accurately assess the presence and extent of lung disease. These include lung function (breathing) tests, which allow an evaluation of how well the lungs work, and a CT scan of the chest, which picks up even mild degrees of lung involvement. Together, the lung function tests and the CT are used to estimate whether lung fibrosis, if present, is mild or extensive.

Another, less common, type of lung involvement is found when the inflammation and scarring are localised in the blood vessels of the lungs. When this is very extensive, it can cause an increase in the pressure of the big vessels feeding into the lungs from the heart. This is called “pulmonary hypertension”. More information on this can be found in the separate leaflet on “pulmonary hypertension in systemic sclerosis”.

Not everybody with systemic sclerosis has lung involvement. Pulmonary fibrosis only occurs in about half of individuals with systemic sclerosis. Pulmonary hypertension only occurs in about one in ten people with systemic sclerosis. Importantly, when an individual with systemic sclerosis has lung involvement they usually have either lung fibrosis or pulmonary hypertension. It is quite rare to have the two in combination to a severe degree.

**Lung fibrosis is found in over 50% of patients with systemic scleroderma**

As part of the initial assessment for patients diagnosed with systemic sclerosis, an echocardiogram, an ultrasound scan which looks at the heart, is usually also performed. The echocardiogram is used to rule out any significant problem with the heart and to determine if there are any signs of pulmonary hypertension. In some individuals, a bronchoscopy will be considered. Bronchoscopy involves passing a flexible fiberoptic camera into the airways in order to obtain samples of the inflammatory cells within the lungs. Not everyone with systemic sclerosis needs a bronchoscopy, however if your physician feels that it could be useful, they will discuss bronchoscopy with you in more detail. One of the reasons to perform a bronchoscopy is to check for possible infection.

### How is systemic sclerosis diagnosed?

The diagnosis of systemic sclerosis is usually based on a combination of symptoms, examination findings and laboratory tests. Common symptoms and examination findings include: thickening of the skin, particularly on the hands, Raynaud's phenomenon (characterised by the tips of the fingers going white and/or blue particularly in the cold), joint pains, and problems with heartburn and indigestion due to acid coming up the gullet from the stomach. Although these symptoms suggest a diagnosis of systemic sclerosis, they can also be found in individuals who don't have the disease. Laboratory tests that are used to diagnose systemic sclerosis include a test for auto-antibodies – these provide evidence of over-activity of the immune system. Other tests that help make the diagnosis include kidney blood tests and a chest x-ray or CT scan to look for any evidence of lung disease or fibrosis.

### How is disease activity measured?

Lung function testing, repeated every six to twelve months, is the most useful test for measuring lung fibrosis and for detecting any worsening of the condition. A persistent worsening of lung function tests usually means that the fibrosis is progressing and is an indicator to your doctor that additional treatment may be required. An additional type of lung scan, called a DTPA clearance scan, may also be performed to predict whether the lung disease is active.

### How is systemic sclerosis-associated lung involvement treated?

The majority of patients with systemic sclerosis lung disease do not need treatment especially if the lung involvement is limited and remains stable over time. Treatment may be needed if the lung disease is significant (is causing a lot of symptoms or is showing up as a major limitation in lung function testing) or if it is getting progressively worse. When there is fibrosis that needs treatment, medications are used that reduce the inflammation that causes the fibrosis. Treatments that are used are not however able to remove fibrosis that has already occurred. For this reason the main aim of treatment for the lungs is to prevent worsening in breathing symptoms and lung function; thus the first aim of treatment is to stabilise lung function and prevent further progression. In some individuals the treatment of the inflammation in the lungs leads to noticeable improvements in symptoms and lung function. However, as the primary aim of therapy is to prevent fibrosis getting worse, stabilisation of symptoms and lung function alone is evidence of response to treatment and should not be viewed as a treatment failure.

Where treatment for systemic sclerosis-associated lung fibrosis is needed, a number of drugs may be used. Many of these are also used purely for the skin or joint problems caused by systemic sclerosis. Other treatments are more specifically designed to treat the lung disease.

The most commonly used treatment for the lungs is a combination of low dose steroids (usually 10 mg once daily) together with an immunosuppressant drug. The immunosuppressants that are most frequently used for systemic sclerosis, and both of which come in tablet form, are azathioprine and mycophenolate. These immunosuppressants reduce the over-activity of the immune system which plays a crucial part in the development of irreversible lung scarring. In individuals with extensive or rapidly worsening lung fibrosis, intravenous treatment (by drip infusion) with cyclophosphamide may be used. Cyclophosphamide is also an immuno-suppressant but when used intravenously it acts more quickly than other immunosuppressant medications. Other novel treatments are currently under investigation for lung fibrosis in systemic sclerosis.

Treatment of lung fibrosis needs to be continued for as long as there is evidence of on-going inflammatory activity due to systemic sclerosis. For most individuals with systemic sclerosis and lung fibrosis however, it is usually possible to gradually reduce and stop treatment once the lung disease is no longer active. It is important to realise however, that this process of waiting for inflammatory activity to subside and then gradually reducing and stopping treatment may take several years.

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