



Sjögren's and the respiratory tract

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Background

Sjögren's Disease is the second most common multisystem auto-immune disease affecting around 0.5% of the female population. The characteristic findings are of dry eyes and mouth as a result of lymphocytic infiltration into the salivary glands. It is commonly associated with fatigue and other extra glandular problems including inflammation of blood vessels and rarely kidney or brain involvement.

The respiratory system encompasses the airways from nose down to the lungs and the lungs themselves.

Any part of the respiratory system can be affected by Sjögren's Disease. In large studies of people with Sjögren's Disease the numbers affecting range from 9-20% of people with Sjögren's. It is likely that a larger proportion of patients, perhaps up to 75%, have abnormalities of the respiratory system which are asymptomatic.

In contrast to Sjögren's Disease in general, which is more common in women, the biggest risk for lung involvement is being male. Smokers and those who have a long duration of disease are also more at risk. People with other systemic Sjögren's problems are also more likely to develop lung disease.

Investigations for lung disease

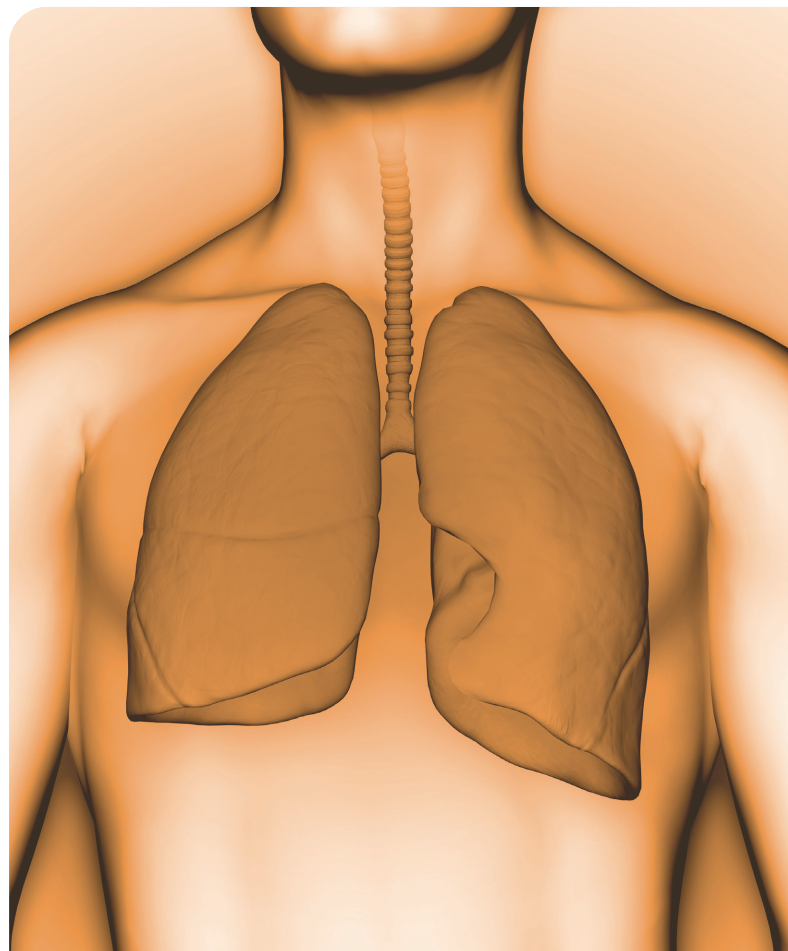
If you develop a cough or shortness of breath it is important to mention it to the team looking after you. Depending on the examination findings you may need some/all of the following investigations.

Chest X-ray

This is usually the first investigation. It usually involves wearing a gown but is quick and not painful! Chest X-rays are good at showing infection or fluid but a CT scan may still be needed.

CT Scan

A CT Scan shows the lung in more detail. Multiple X-rays are used to build up a detailed picture of the lungs. It is necessary to lie down on the examination table which is moved in and out of the scanner. Your head remains outside the scanner. Films are taken whilst breathing in and out and holding breath. The process takes about 30 minutes in total and shouldn't be uncomfortable. It can be difficult to lie still. Occasionally an injection of contrast may be needed which would involve a small cannula in the hand.



Lung Function Tests

These are used to measure how efficiently lungs are working. The amount of air going in and out is measured together with a test called gas diffusion which measures how efficiently oxygen gets into your blood stream.

These tests involve blowing in and out through a mouthpiece often with a nose clip on. Other tests include breath holding. Certain tests called plethysmography require sitting inside a small airtight booth while pressure and air flow measurements are collected. The booth usually has see-through sides. These tests can be difficult to perform particularly if coughing is a problem. The technicians are very good at explaining what needs to be done and helping you to get the best results.

Cough

The most common symptom of respiratory disease is a dry cough. It is likely that this is caused by a combination of dry airways and a lymphocytic infiltrate into the trachea, bronchi or bronchioles.

Treatment of Cough

Dry airways may benefit from the use of pilocarpine (a drug which non-specifically stimulates secretions by stimulating muscarinic receptors on the surface of secretory cells) or nebulised saline solution which can help with hydration. Drinking plenty and keeping hydrated may also help.

Pneumonia

Recurrent chest infections may occur in people with Sjögren's. It is thought that this is multifactorial and may be related to abnormal sputum production, difficulties clearing sputum, reflux and treatment with immunosuppression.

Prompt recognition and treatment of infections with antibiotics is essential to prevent lung damage (Bronchiectasis) which can also be seen in Sjögren's (7-40% on CT).

Interstitial Lung Disease

This is the most serious complication of the lungs in Sjögren's Disease but symptomatic disease is unusual affecting less than 1/5 people with Sjögren's.

Interstitial lung disease (ILD) means respiratory disease affecting the interstitium (the tissue and space around the air sacs of the lungs). It includes alveolar epithelium, pulmonary capillary endothelium, basement membrane, perivascular and perilymphatic tissues.

ILD usually presents with shortness of breath plus cough. In the early stages, a chest X-ray may be normal. CT scans and lung function tests are usually used to make the diagnosis. Different patterns of disease on CT scan predict

long term prognosis and also response to treatment.

The most common pattern is known as NSIP (non-specific interstitial pneumonitis) and also occurs in other connective tissue diseases such as Systemic Lupus and Rheumatoid Arthritis.

Treatment is based on the management of interstitial lung disease in other conditions and usually includes corticosteroids and may include other immunosuppressants such as Azathioprine or Mycophenolate mofetil.

In rapidly progressive cases, Cyclophosphamide (chemotherapy) or Rituximab may be tried.

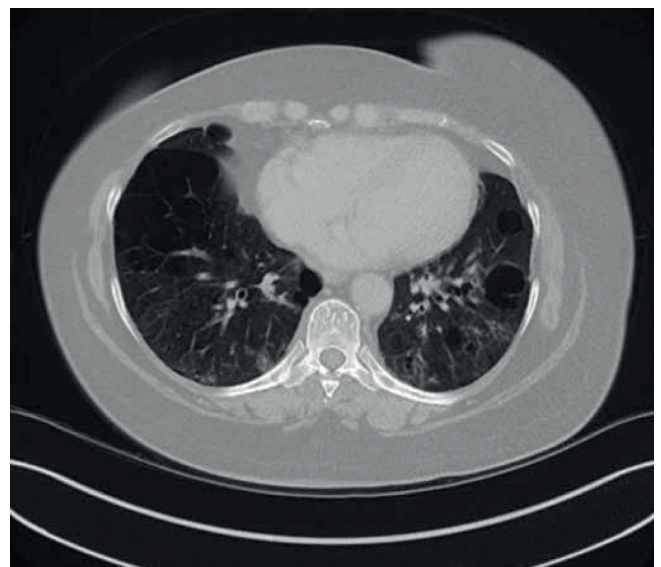
Inhalers

These are not prescribed specifically for ILD but many patients particularly those with co-existent asthma or chronic obstructive pulmonary disease (COPD) do have benefit from bronchodilators (inhalers which contain medication to relax the muscles within the airways opening them up and making breathing easier). The results from lung function tests may help decide whether you might benefit from an inhaler.

Sometimes a condition called Lymphocytic Interstitial Pneumonitis is seen and this condition only occurs in Sjögren's Disease. Large cysts are seen on the CT scan together with interstitial changes. This condition usually responds very well to steroids.

Summary

Respiratory Disease is common in Sjögren's but serious disease is rare. Management includes self-help with good hydration and more serious conditions may require the use of steroids or other immuno suppressant drugs.



CT Scan showing characteristic appearances of Lymphocytic Interstitial Pneumonia.