ELF EUROPEAN LUNG FOUNDATION

ERS EUROPEAN RESPIRATORY SOCIETY
every breath counts

Sarcoidosis

This factsheet is about sarcoidosis symptoms, diagnosis and treatment. It also explains how sarcoidosis affects people. It is for anyone affected by the condition whether you are newly diagnosed or have been living with this condition for a long time. As the lungs are the most commonly affected organ, this factsheet is mainly about pulmonary (lung) sarcoidosis. You can find out more about how sarcoidosis affects other organs by following the links included in the Further Information section.

What is sarcoidosis?

Sarcoidosis is an inflammatory condition. In people with sarcoidosis, patches of red, swollen tissue (granulomas) form in the body. These granulomas are made up of inflammation cells which clump together. The cause of these granulomas is still not known.

Sarcoidosis affects many organs in the body, but the lungs are most commonly affected. It can also affect the skin, eyes, heart, muscles, joints, bones, liver, kidneys and brain (and more). Chest physicians (pulmonologists) are most often involved in diagnosing and managing this disease.

Many people with sarcoidosis do not need treatment, and the disease may go away on its own. There are treatments that can help to slow the disease and improve symptoms. However, currently no treatment is available to cure this disease.

Sarcoidosis is a rare condition and it is not known for sure how many people have sarcoidosis. This is because many people may not know they have it, and sometimes it can be mistaken for other diseases. The number of cases varies from country to country, with around 2-40 people per 100,000 affected. Scientists estimate that 344,000 people are diagnosed each year worldwide.

What causes sarcoidosis?

Scientists do not know the cause of sarcoidosis, but there is research being carried out to try and answer this question.

What we do know is:

- Sarcoidosis is not infectious (you cannot catch it and you cannot pass it to another person).
- Sarcoidosis is not a form of cancer.
- Sarcoidosis does run in families, but there is only a 5-10% chance someone else in your family will have the disease.

Most people with sarcoidosis get better without specific treatment within 12-18 months and go on to lead normal lives. This is because the body's immune system can heal the condition over time.

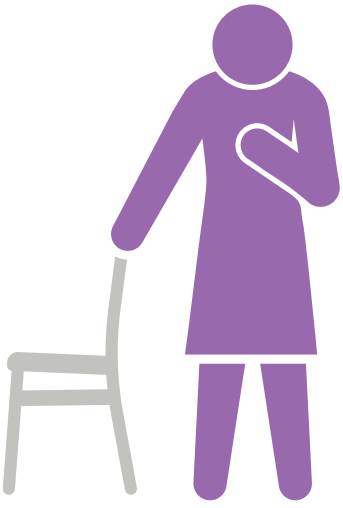
Sometimes, for reasons that are not understood, the immune system does not heal the granulomas and scar tissue forms (fibrosis). This can result in damage to the affected body part that requires long-term treatment and monitoring.

What are the symptoms and how is it diagnosed?

Diagnosing sarcoidosis can be difficult. Symptoms are very varied and no single test gives a definite diagnosis for this disease.

The most common symptoms are:

- Dry cough
- Shortness of breath
- Chest pain
- Fatigue
- A flu-like illness with fever, tiredness and joint pains
- A painful red rash, usually on the arms or legs
- Eye irritation and sight problems
- Swollen glands, which can be felt in the neck or around the face



People with sarcoidosis tell us that difficulties with concentration and remembering facts can also be a symptom.

It may take a long time for doctors to make a diagnosis as they must rule out other conditions that look like sarcoidosis, and no two people will have the same symptoms. This can be a very slow and frustrating process for people wanting to know what is wrong with them.

Sometimes sarcoidosis is discovered by chance, for example when a chest X-ray is done for other reasons.

Testing for sarcoidosis usually includes:

- A chest X-ray to find out whether the lungs and lymph glands (part of the immune system) are affected.
- Blood tests, including liver and kidney function, calcium and other blood values of the immune system. This can include an ACE level (Angiotensin Converting Enzyme) which may be higher in someone with sarcoidosis.
- Tests to find out how severe the disease is, and which parts of the body are affected, including:
    - Heart tracing (ECG).
    - Lung function tests (spirometry, single breath gas transfer). See ELF spirometry factsheet
    - Eye examination

However, a normal ACE or chest X-ray does not rule out sarcoidosis while an elevated ACE level does not diagnose sarcoidosis.

Doctors may decide to do extra tests depending on your symptoms. This could include laboratory tests, urine tests, a heart ultrasound (echo), a brain scan (MRI) or a chest scan (CT).

Often, doctors need tissue samples or biopsies to confirm the diagnosis. These are usually taken from the glands in the lung, lung tissue, glands in the neck or chest, or from the skin. If a biopsy is needed it will usually be done under local anaesthetic.

> *"It can be difficult for patients, as we live in constant doubt about different signs and symptoms that occur, and do not know if they are related to the disease or not."* (patient quote)

How will I be treated?

Your doctor will discuss with you if treatment is needed and what the options are. Many people do not need treatment and can simply be checked regularly.

There are a few different drugs used to treat sarcoidosis, which often work by reducing symptoms.

Sometimes sarcoidosis medicines treat a single body part, such as eyedrops or skin creams. Other medicines treat the whole

body from the inside and can be particularly useful for treating granulomas in the lungs, brain and heart – including:

- Prednisolone and prednisone (steroids) - the most common drugs used for sarcoidosis.
- Methotrexate and azathioprine are often used to help people come off prednisolone (this is known as 'steroid-sparing').
- Infliximab and similar drugs can sometimes work when prednisolone or steroid sparing drugs do not work.

Sarcoidosis responds very well to steroids, but they do not cure the disease. Sarcoidosis may get worse again after the steroids are stopped. Steroids can also cause side-effects, such as weight gain, diabetes, thinning of the bones (osteoporosis) and skin, and mood swings. Other drugs can have other side-effects. It is a good idea to talk to your doctor about the risks and benefits of any treatment prescribed to you.

If you begin steroid treatment, your doctor will help you to find the lowest dose that can control your symptoms by gradually lowering the dose over time. If it cannot be lowered, a second drug such as methotrexate or azathioprine may be added.

Monitoring of your condition will depend on how sarcoidosis affects you. Most people are seen every 3 months or so for the first 2 years.

Besides medication, other support such as exercise, pulmonary rehabilitation and psychological support may be offered. Talk to your doctor about the options that could help you.

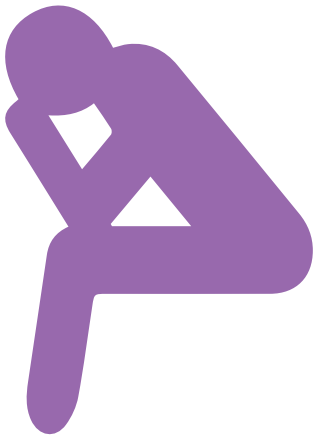
How does sarcoidosis progress?

Most people who are diagnosed with sarcoidosis get better without specific treatment and go on to lead normal lives. But around 1 in 4 will have long-term chronic symptoms that require ongoing treatment.

Daily living and support

Living with sarcoidosis can affect you emotionally as well as physically. People have found that it has a significant impact on all aspects of their lives. This may be because of fatigue, chronic pain, sleeping problems, difficulty working as usual and depression. Some of these effects do not improve much with treatment.

Having sarcoidosis is different for every person. It is important that you get the support you need. You can talk to your doctor about:



- How you are feeling (physically and mentally).
- Other types of support (including psychological support and help to stop smoking).
- How to maintain your quality of life.
- Improving your diet and activity levels.

Remember that you are not alone in what you are going through. There are patient organisations and support groups where you can find more information. See the Further Information section.

> *"Scans, tests and examinations are crucial in the treatment, but my well-being and quality of life determine how much I suffer from sarcoidosis. And that is the most important thing."* (patient quote)

Further Information

- **ELF website: www.europeanlunginfo.org/sarcoidosis** links to more information including websites of national patient organisations in your own language.
- **World Association of Sarcoidosis and Other Granulomatous Disorders (WASOG):** www.wasog.org. Takes a multidisciplinary approach to interstitial lung diseases including sarcoidosis and brings together physicians and patients through information, education and research.

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The European Lung Foundation (ELF) was founded by the European Respiratory Society (ERS) in 2000 with the aim of bringing together patients, the public and respiratory professionals to positively influence lung health.

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