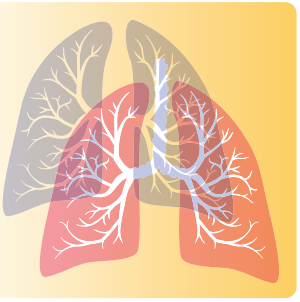
What is Sarcoidosis?

Part 1

Sarcoidosis (sar-coy-DOE-sis) is a disease of unknown cause in which inflammatory cells clump together and form tiny lumps of cells in various organs and tissues of the body. These lumps are called granulomas (gran-yu-LO-mas). Sarcoidosis most often affects the lungs and its hilar lymph nodes but can also involve other areas of the body including the eyes, skin, sinuses, liver, kidneys, brain and heart. Sarcoidosis varies in how active and how severe it is for each person and over time. The granulomas, when active, can cause short term and/or long term damage to the organ involved. This fact sheet provides a general overview of sarcoidosis. For more information about treatment of sarcoidosis, see "Treatment of Sarcoidosis" at www.thoracic.org/patients.



Chronic inflammation and damage leads to symptoms and occasionally, permanent loss of function of the involved tissue/organ . When sarcoidosis affects the lungs (pulmonary sarcoidosis), the disease can reduce the amount of air the lungs can hold and cause abnormal stiffness, called "restriction," of the lungs. This results in breathing problems that can interfere with daily activities.

What causes sarcoidosis?

The cause of sarcoidosis is unknown. The disease can be seen in people of every race, sex and age. However, sarcoidosis is more common in people who are:

- African-Americans.
- Of German, Irish, Scandinavian, Asian or Puerto Rican origin.

Sarcoidosis is not contagious and it is not a cancer. People with a family member who has sarcoidosis have a low risk (1 in 20) of also getting the disease.

What are the signs and symptoms of sarcoidosis?

Many people with sarcoidosis do not have any symptoms. Others have only vague symptoms that can be seen in many other illnesses, such as weight loss, fever, loss of appetite, depression, night sweats, and sleep problems. Signs and symptoms that may come from problems with a specific organ include:

- **Lungs:** Shortness of breath, wheezing or dry cough that may lessen or go away over time in some people, but remain in others.
- **Lymph nodes:** Enlarged and sometimes tender lymph nodes, most often in the neck and chest, but sometimes under the chin, arm pits or groin.
- **Eyes:** Burning, itching, tearing, redness, sensitivity to light, dryness, seeing black spots, blurred vision, reduced color vision, and, in rare cases, blindness.
- **Skin:** Bumps, ulcers, or rarely, flat areas of discolored skin that appear mostly near the nose or eyes or on the back, arms, legs and scalp. Painful and tender discreet reddened bumps called *erythema nodosum* can suddenly appear on the ankles and shins. This rash is most often seen in younger patients and can be associated with joint pains, fever and enlarged lymph nodes in the chest called *Löfgren's syndrome*.
- **Bones and Joints:** Bone lumps (nodules), causing pain in the hands and feet, and/or swelling of ankles or other joints.
- **Spleen and Liver:** There can be pain in the upper abdomen, under the ribs on the right (liver) or left (spleen).
- **Heart:** Shortness of breath with activity and swelling in the legs. One may have an irregular or fast heart beat at times, or pass out without warning.
- **The Nervous System:** Headaches, vision problems, numbness, weakness, or loss of movement of arms or legs, drooping of one side of the face, pain or a "pins and needles" feeling.
- **Fatigue** is a common problem, seen in more than half of patients.

How is sarcoidosis diagnosed?

Since sarcoidosis can affect one or more parts of the body, the signs and symptoms depend on the tissue/organs involved. Some people with the disease do not have any symptoms and it may be noticed by chance when they are being seen for other problems. Other people may be hard to diagnose because the symptoms they have are not very specific. But certain clinical features such as the erythema nodosum, rash or eye findings may lead a healthcare provider to suspect sarcoidosis.

There is no specific blood test to diagnose sarcoidosis. Sarcoidosis can mimic infection and several other diseases. Hence the diagnosis requires checking for other causes of symptoms and disproving infection, cancer and other diseases as a cause.

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Your healthcare provider will do a history and physical exam to look for signs of sarcoidosis and rule out other diseases. A number of other studies may be done including:

- A **chest X-ray** to look for enlarged lymph nodes and small round spots, called granulomas, in the lungs
- **Pulmonary function tests** to measure how well the lungs work. (For more information, see ATS Patient Information Series: Lung Function Testing at www.thoracic.org/patients)
- **Bronchoscopy and bronchoalveolar lavage**
  A bronchoscopy is a test in which a flexible tube is put into the airways and mucus fluid samples can be suctioned out. This fluid is called bronchoalveolar lavage (BAL) and the cells in it can be examined under the microscope. (For more information, see ATS Patient Information Series: Flexible Bronchoscopy at www.thoracic.org/patients)
- **A tissue biopsy—**taking a small piece of tissue to examine under a microscope to look for signs of disease. This can be obtained through surgery or bronchoscopy (endobronchial or transbronchial biopsies). A special device with ultrasound guided biopsy via bronchoscopy (EBUS) may also be used to get samples from the lung.
- An **eye exam** with a specially lighted tool allows the doctor to look inside the eye for possible signs of sarcoidosis.
- **Blood tests** can show reflect abnormal function of the involved organs such as the liver, kidney, bone marrow and calcium levels.
- A **CT scan** of the chest may show enlarged lymph nodes and scars in the lungs that a regular chest X-ray may not.
- An **electrocardiogram (EKG)** is a test that records a tracing of the electrical activity of the heart. It shows the rate of heart beats. It may show how regularly the heart beats and may show if there is any strain on the heart, irregular rhythm, heart blocks.
- An **echocardiogram** may detect abnormal heart function and/ or increased pressure in the arteries of the lungs (pulmonary hypertension) that can occur in some cases.
- An **MRI** of the heart is a type of imaging scan that can detect heart involvement by sarcoidosis earlier than an EKG. It can also be helpful to identify areas in the heart to biopsy, if needed.
- **PET scan** of the may detect sarcoidosis in various parts of the body and may predict response to treatment. A dedicated cardiac PET scan can be done to detect sarcoidosis in the heart.

Definitive diagnosis requires the provider to put together information from the history, physical exam, and laboratory results that show granulomas (characteristic features of sarcoidosis).

How serious is sarcoidosis?

The course of sarcoidosis varies greatly among people and over time for an individual. In many cases, sarcoidosis is mild and self-limited. A short time after appearing, the granulomas may stop growing or shrink. Symptoms may go away within a few years without treatment.

Sarcoidosis starts with active, ongoing inflammation. Granulomas (lumps) form and grow. Symptoms develop, and scar tissue can form in the organs where the granulomas are growing. In most patients, the inflammation decreases, and the granulomas stay the same size or shrink within a few years. But any old scars will remain and can still cause symptoms. However, for some patients, sarcoidosis can become chronic, lasting life-long.

The severe form of sarcoidosis slowly worsens over a period of years, and can cause permanent organ damage. Treatment can help, but the disease may still leave scar tissue in the lungs, skin, eyes, sinuses, heart or other organs. Between 20 and 30 percent of people with pulmonary sarcoidosis end up with permanent lung damage.

The scarring from sarcoidosis is often in upper portions of the lungs. The lungs are at risk of infection due to fungus or bacteria, including mycobacteria. A 'fungus ball' can form and a person can develop bleeding (cough up blood).

There are two reasons to treat sarcoidosis: concern for developing organ damage (including respiratory failure) and/ or to improve quality of life. Treatment is aimed at maintaining good function of the organ involved, reducing symptoms, improving quality of life, and preventing organ damage. Talk with your healthcare provider about how sarcoidosis is affecting your health and what treatment you may need.

Author: Ganesh Raghu, MD
Reviewers: Marianna Sockrider, MD, DrPH; Hrishikesh S. Kulkarni, MD; Ginger Spitzer; Robert Baughman, MD

Rx *Action Steps*

- ✔ Talk with your healthcare provider about whether you need treatment for sarcoidosis and what options you have.
- ✔ Do not smoke and try to avoid being around tobacco smoke.
- ✔ Take action to stay healthy and watch for early changes with sarcoidosis.

Healthcare Provider's Contact Number:

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Additional Lung Health Information

Foundation for Sarcoidosis Research
http://www.stopsarcoidosis.org

American Thoracic Society
www.thoracic.org/patients

National Heart, Lung and Blood Institute
http://www.nhlbi.nih.gov/health/health-topics/topics/sarc
(also available in Spanish)

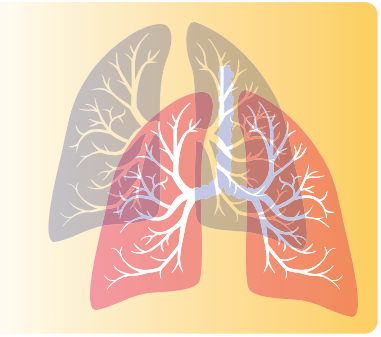
American Lung Association
http://www.lung.org/lung-health-and-diseases/lung-disease-lookup/sarcoidosis/learn-about-sarcoidosis.html

American Lung Association of Canada
http://www.lung.ca/lung-health/lung-disease/sarcoidosis-1

Treatment of Sarcoidosis

Part 2

Sarcoidosis (sar-coy-DOE-sis) is a disease of unknown cause in which inflammatory (white blood) cells clump together and form tiny lumps of cells in various organs and tissues of the body. These lumps are called granulomas (gran-yu-LO-mas). Sarcoidosis most often affects the lungs and its surrounding lymph nodes but can also involve other areas of the body including the eyes, skin, sinuses, liver, kidneys, brain and heart. Sarcoidosis varies in how active and how severe it is for each person and over time. The inflammation in sarcoidosis may resolve spontaneously, even without treatment, or it may persist, causing symptoms and organ damage that may get worse over time. The granulomas, when active, can cause short term and long term damage to the organ involved. This fact sheet reviews treatment of sarcoidosis. For more information about sarcoidosis see 'What is Sarcoidosis' at www.thoracic.org/patients.



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The severe form of sarcoidosis slowly worsens over a period of years, and can cause permanent organ damage. Treatment can help, but the disease may still leave scar tissue in the lungs, skin, eyes, sinuses, heart or other organs. Between 20 and 30 percent of people with pulmonary sarcoidosis end up with permanent lung damage.

The scarring from sarcoidosis is often in upper portions of the lungs. The lungs are at risk of infection due to fungus or bacteria, including mycobacteria. A 'fungus ball' can form and a person may cough up blood (hemoptysis) as a result of the fungus.

There are two reasons to treat sarcoidosis: concern for developing organ damage (including respiratory failure) and/or to improve quality of life.

Treatment is aimed at maintaining good function of the organ involved, reducing symptoms, improving quality of life, and preventing organ damage. Talk with your healthcare provider about how sarcoidosis is affecting your health and what treatment you may need.

What medicines are used to treat sarcoidosis?

Medications to treat inflammation in sarcoidosis include:

- **Corticosteroids.** The most common corticosteroid prescribed for sarcoidosis is prednisone. Initially, higher doses may be used to control the inflammation. Once your symptoms are better, your healthcare provider may decrease the dose slowly.
- **Methotrexate.** This medicine may be given with or in place of corticosteroids.
- **Azathioprine, hydroxychloroquine, mycophenolate** are other drugs that may be used if corticosteroids and methotrexate are not effective. Thalidomide and minocycline may be used in skin sarcoidosis.

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- **Infliximab** and **adalimumab** are newer treatments that have proved useful for chronic sarcoidosis when standard treatments have failed.

Research continues to try to find new treatment for sarcoidosis. Other drugs are currently being tested in clinical trials.

Medicines commonly used to treat sarcoidosis may cause side effects. Side effects range from those that are mild, to those that are severe and potentially dangerous. If you are taking one or more medicines for sarcoidosis, you will need to be monitored closely by your healthcare provider.

Other medicines used for symptoms in sarcoidosis

Some medications may help improve the symptoms from sarcoidosis without treating the sarcoidosis itself. Sarcoidosis associated fatigue may be treated with neurostimulants such as methylphenidate and modafinil. Small fiber neuropathy may be treated with medicines such as gabapentin.

Is there any role for lung transplantation in sarcoidosis?

Some people with advanced lung disease because of sarcoidosis may be eligible to receive a lung transplant. Many factors go into deciding if you need a transplant and will be able to tolerate the surgery. Your healthcare provider can refer you to a lung transplant center for further testing and information. For additional information on lung transplantation, go to www.thoracic.org/patients/.

What else can I do to stay as healthy as possible with sarcoidosis?

Many people with sarcoidosis can lead normal lives, and are able to carry on with their usual social, intellectual, artistic and athletic activities. There are several important steps a person with active or inactive sarcoidosis can take to keep healthy:

- If your sarcoidosis is active, have frequent medical check-ups so your doctor can monitor your illness and adjust your treatment if needed.
- If you are not taking any medications and have no symptoms from your sarcoidosis, you should visit your healthcare provider for a checkup at least once a year for at least the first few years.
- See a medical eye specialist (an ophthalmologist) for an eye exam each year.
- Don't smoke. While smoking doesn't cause sarcoidosis, it can make your lung function worse. For help quitting smoking, go to www.thoracic.org/patients.
- Avoid dust, chemicals, fumes and other substances that can harm your lungs.
- Exercise and be as active as you can, but don't strain yourself. A pulmonary rehabilitation program is a good way to gradually increase your exercise capacity. Research has shown that a supervised pulmonary rehabilitation program may reduce fatigue and improve a person's quality of life. For more information, see ATS Patient Information Series "Pulmonary Rehabilitation at www.thoracic.org/patients".

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