

X-Plain Sarcoidosis

Reference Summary

Introduction

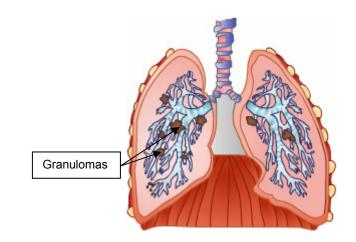
Sarcoidosis is a disease that develops gradually, producing symptoms such as small lumps that come and go throughout life. It can appear in almost any body organ, but often starts in the lungs. About 140,000 Americans suffer from sarcoidosis. The majority of patients with sarcoidosis can expect to live a normal and active life. Some cases of sarcoidosis are severe or fatal.

This reference summary will help you understand sarcoidosis, its symptoms, causes, diagnosis, and treatment options.

Granulomas

Sarcoidosis is a disease where *granulomas* appear in various organs of the body. Granulomas are lumps that form when cells of the immune system clump together. The immune system is responsible for fighting germs that invade the body.

The immune system is made of special blood cells that destroy germs, such as viruses and bacteria. These blood cells are called white blood cells.



There are 2 types of white blood cells.

- 1. T-cells
- 2. B-cells

T-cells identify a foreign material or germ and attack it themselves.

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When B-cells identify a foreign material or germ, they secrete special chemicals called *antibodies*. These antibodies stick on the foreign material and destroy it. When T-cells and B-cells identify a foreign organism, they release chemicals that recruit other blood cells to help destroy the foreign organism.

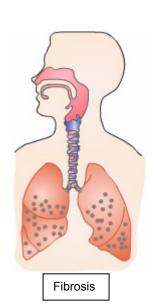
The reaction of the immune system to foreign materials and germs causes an inflammation. The inflamed site looks red and swollen. Sometimes, all the cells that come to the infection site clump and form a small bump. This bump is called a granuloma.

Sarcoidosis

Granulomas that appear in people with sarcoidosis can form in any organ or tissue of the body. The lungs are usually the first site affected. Granulomas can also affect the lymph nodes, the skin, the brain, the spinal cord, the kidneys, and the eyes.

In the majority of cases, granulomas clear up and heal in 2-3 years, without the patient knowing or doing anything about them. In a few cases, the granulomas do not disappear and the tissue becomes scarred, which is called *fibrosis*.

If granulomas keep forming and causing scarring, sarcoidosis is called *chronic*, which means ongoing and long-lasting. If an important organ, such as the heart is scarred extensively by sarcoidosis, the damage can be serious and sometimes fatal. This is rare.



Most sarcoidosis patients live normal lives. About 60% recover on their own, 30% have permanent damage, and 10% have serious damage that can be fatal.

Sarcoidosis usually affects people between 20-40 years of age. White women are as likely as white men to get it, but black women get sarcoidosis 2 times as often as black men.

Causes

Sarcoidosis is due to an abnormal reaction of the immune system, the part of the body that fights infection. Scientists do not currently know what causes this abnormal reaction of the immune system.

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Symptoms

The symptoms of sarcoidosis depend on which organ and body parts are affected. In addition, symptoms vary from patient to patient.

If the lungs are affected, patients usually have shortness of breath and a cough that will not go away.

If the eye is affected, the symptoms depend on which parts of the eye are involved. In some cases, there are no symptoms at all and in others, eye inflammation, cataracts, glaucoma, and blindness may result.

Small, raised patches on the face usually mark skin sarcoidosis. Occasionally, the patches are large and purplish in color. Patches may also appear on limbs, face, and buttocks.

If joints are affected, sarcoidosis can lead to *arthritis*. Arthritis is an inflammation of joints that causes pain and stiffness. Joints that are commonly affected by sarcoidosis include the ankles, elbows, wrists, and hands.

Granulomas caused by sarcoidosis can appear in the brain, spinal cord, and facial and optic nerves. This could lead to symptoms associated with the nerves involved. For instance, if the facial nerve is involved, sarcoidosis can lead to facial paralysis.

If the liver, kidney, or heart are affected by sarcoidosis, they may malfunction. Symptoms are related to the part affected and to the severity of the disease.

Sarcoidosis may also have general symptoms that are not specific to the affected organs, such as weight loss, fatigue, night sweats, and fever.

Diagnosis

Sarcoidosis is relatively difficult to diagnose because symptoms may resemble those of other diseases. The doctor usually confirms the diagnosis of sarcoidosis by eliminating other diseases.

The doctor first takes the patient's medical history and does a physical examination. X-rays and blood tests are usually the first procedures the doctor will do.

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A chest x-ray can give the first indication of sarcoidosis in the lungs. Advanced radiological techniques, such as MRI^1 and CAT^2 scans, can be used for other organs.

Analysis of the blood can also give an indication of sarcoidosis. This includes the number and types of blood cells, blood proteins, serum calcium levels, and a special chemical associated with sarcoidosis, angiotensin converting enzyme or ACE.

Several diagnostic tests are specific to affected organs. For example, a *pulmonary function test*, or PFT, can provide clues to the diagnosis of lung sarcoidosis. This test helps measure how well the lungs are functioning.

When a doctor suspects sarcoidosis in a body organ, he or she takes a sample of the tissue to be examined under a microscope. The type of cells in the sample can tell the doctor if there are inflammatory cells and/or granulomas.

The site of inflammation can also be found by taking pictures of the inside of the body after injecting radioactive materials. This is called a gallium or a thallium scan.

Treatment

Since sarcoidosis can disappear without treatment, the doctor may recommend observing the disease and not treating it. Treatment depends on the organ involved and how much inflammation there is. The purpose of treatment is to keep the organ working and to relieve symptoms.

Corticosteroid medication treats the inflammation and stops the formation of granulomas. Prednisone is a common corticosteroid medication. Corticosteroid treatment usually results in improvement. Symptoms often start up again, however, when it is stopped. Treatment may be necessary for several years, sometimes as long as the disease remains active. Corticosteroids can have side effects. If your doctor prescribes corticosteroids, make sure to discuss the potential side effects with your doctor. Corticosteroids should not be stopped without the supervision of a doctor.

Other medications are available to treat sarcoidosis. Hydroxychloroquine, which is used to also treat malaria, can be helpful in about one third of the patients, especially those with skin involvement or high levels of calcium in the blood. One of its side effects is that it can damage the eyes, therefore eye examinations should be done every six months. Methotrexate, azathioprine and cyclophosphamide are medications

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that affect the immune system and tend to help with sarcoidosis. They do however have some significant side effects including: causing abnormalities in unborn children and increasing the risk of some cancers.

Frequent checkups are important so that the doctor can monitor the illness and, if necessary, adjust treatment or stop it. There is currently no treatment to reverse fibrosis or scarring that might be present in advanced sarcoidosis.

If blood tests show high calcium levels, the patient may need to avoid calcium-rich foods, vitamin D, or sunlight. Examples of food rich in calcium are dairy products, oranges, and canned salmon with bones.

As a last resort, a lung transplant may be done.

Summary

Sarcoidosis is a disease due to inflammation of body tissues. Small lumps called sarcoid granulomas may form that can leave scars in the affected organs.

In most cases, granulomas disappear and do not come back. If they do not disappear, they may cause symptoms, depending on the affected organs.

Thanks to medical advances, effective treatment is available for those who have sarcoid granulomas that do not disappear on their own.



Most patients with sarcoidosis can expect to live normal and active lives!

¹ MRI, or Magnetic Resonance Imaging, is a technique that allows the doctor to create pictures of areas inside the body by using a magnet linked to a computer.

² CAT Scan or Computed Axial Tomography scan is a radiological test that provides pictures of structures inside the body by taking multiple X-ray images.