



SARCOIDOSIS

Sarcoidosis is a disease characterized by the presence of granulomas. Ball-like collections of white blood cells, granulomas may be associated with inflammation. The inflammation can damage organs within the body. Inflamed granulomas, most often occur within the lungs, but any area of the body may be affected. Inflamed granulomas may resolve on their own, resolve with treatment or cause permanent scarring. In about half of people, sarcoidosis resolves within two years of diagnosis. About two-thirds of people demonstrate resolution of the disease within a decade of diagnosis.

The cause of sarcoidosis is unknown. Sarcoidosis is a world-wide disease which occurs more often in Caucasians of European descent and in African-Americans. It is most common in young people between the ages of 20 and 40. Sarcoidosis is not contagious.

What are the Signs and Symptoms?

People with sarcoidosis may have no symptoms, vague symptoms of a general nature, or symptoms associated with a specific organ, usually the lungs. More than one organ may be involved. Up to half of people with sarcoidosis have no symptoms when the illness is diagnosed. In some cases, obtaining the diagnosis of sarcoidosis may be difficult.



Signs and symptoms associated with specific organ involvement can include the following:

General, Nonspecific Symptoms

Nonspecific symptoms may be present including fatigue, weakness, trouble sleeping, weight loss, and fevers.

Lungs

Inflammation in the lungs may cause shortness of breath, wheezing or cough (often a dry cough). In some people, the symptoms go away with or without treatment; in others there may be permanent scarring in the lungs.

Lymph Nodes

Enlargement of various lymph nodes can occur, especially the lymph nodes in the chest.

Eyes

Inflammation of the eye can lead to pain, redness, sensitivity to light, blurred vision and in severe cases

blindness. Many people may have dry eyes. Some people may have eye involvement without obvious visual problems. It is, therefore, recommended that all people with sarcoidosis visit the eye doctor every year.

Skin

Skin involvement may appear as raised, pink or purplish areas or as painful nodules under the skin. Swollen, painful red bumps on the arms or legs may be seen with arthritis in sarcoidosis.

Joint

Sarcoidosis may cause swelling, stiffness and pain within the joints. The ankles, knees, hands and wrists are most often affected.

Kidney

Inflamed granulomas rarely involve the kidneys. When kidney function is compromised by inflamed granulomas, the creatinine level in the blood will rise.

Spleen and Liver

The spleen and/or liver may be enlarged so it may be felt by the health care provider during the physical exam.

Heart

Heart involvement occurs in a small percentage of people and may be difficult to diagnose. Abnormalities involving the electrical system within the heart may compromise the ability of the heart muscle to pump blood.

Brain and Nervous System

Inflamed granulomas may develop within the brain and the nerves causing symptoms which include numbness in the extremities, muscle weakness, paralysis of the facial nerve and headaches.

Salivary Gland

The salivary gland may also be affected. People with salivary gland sarcoidosis may have a dry mouth.

How is the diagnosis made?

The first step in diagnosing sarcoidosis is a good evaluation. Inflamed granulomas may be seen under a microscope. These granulomas may have a similar

appearance to those in other diseases. A careful exam of the granulomas (obtained in a biopsy) by an experienced pathologist is important to exclude causes of granulomas other than sarcoidosis.

An evaluation to detect sarcoidosis should include the following:

Thorough Medical Examination

A thorough medical examination is important. This will include a detailed history and physical exam.

Chest X-Ray

Doctors look at the chest X-ray for evidence of enlarged lymph nodes and small round spots in the lung caused by granulomas. The chest X-ray may have patterns of disease known as stages. The stage of the chest X-ray (ranging from 0-IV) may provide further information about sarcoidosis. It is only the chest X-ray (not the disease) that is staged in sarcoidosis.

Pulmonary Function Tests

Standard breathing tests provide information about lung function. Breathing tests are important in evaluating and monitoring lung function, especially when sarcoidosis affects the lungs. These tests may show on or all of the following, obstruction of air flow out of the lungs, restriction of air flow into the lungs, and problems in the oxygen transport within the lungs. The most important types of breathing tests in sarcoidosis are spirometry, lung volumes, and diffusing capacity. In some cases, measurement of blood oxygen levels during an exercise test may also be done.

Tissue biopsy

A microscopic examination of tissue samples from the lungs or other affected organs is needed to be absolutely sure of the diagnosis. A bronchoscopy is often done to obtain tissue. A bronchoscopy is an outpatient procedure in which the doctor places a narrow scope (tube) with a camera at its end through the nose and into the airways.

A non-invasive skin test call the Kveim-Sitzbach Test can be used to confirm the diagnosis of sarcoidosis. The test is diagnostic in up to 80 percent of people. Sometimes the diagnosis of sarcoidosis is made by

obtaining tissue samples from involved organs such as the skin, liver, or enlarged lymph nodes.

Bronchoalveolar Lavage

When a bronchoscopy is done, a small part of the lung tissue can be washed (lavaged) in order to obtain some cells of the immune system from the lung. Examination of the immune cells determines whether there is inflammation present within the lung and whether the inflammation is characteristic of sarcoidosis.

Eye Examination

A slit lamp examination by an eye doctor (ophthalmologist) is an important part of an eye examination to detect inflammation.

S.A.C.E. (Serum Angiotensin Converting Enzyme Level)

S.A.C.E. is a blood test. If the level of this blood test is high, it can indicate the presence of sarcoidosis. It can sometimes be used to monitor if sarcoidosis is improving or worsening. Unfortunately, other diseases can cause increased S.A.C.E. blood levels also, so this test cannot be used alone to make the diagnosis.

CT Scan

A CT Scan is a detailed type of X-ray. The CT scan may make it possible to see lymph node nodules and scars in the lung when regular chest X-rays sometimes cannot.

Gallium Scan

The Gallium scan is a type of X-ray scan. A small amount of radioactive material is injected into a person's blood, which travels to areas of the body that are inflamed. An X-ray camera scans the body to see where the radioactive material has collected. Although not needed in all cases, gallium scanning can help show sarcoidosis in many organs, including the lungs, salivary glands, bone, spleen, and liver. Many people with sarcoidosis may have inflamed granulomas. The gallium scan may determine the location and extent of inflammation in the body.

Calcium Levels in the Blood and Urine

Inflamed granulomas make their own vitamin D. In some people, the vitamin D produced by the

granulomas is not properly regulated, which results in too much vitamin D. The overproduction of vitamin D may lead to high calcium levels in the blood or urine. Elevated urine calcium levels may result in kidney stones. Exposure to the sun may further stimulate this process. It may be necessary to collect a urine sample for 24 hours to measure the calcium level as well as collect a blood sample for calcium.

What is the treatment?

Not everyone with sarcoidosis will need treatment. Up to one half of the people diagnosed with sarcoidosis improve without treatment. Those who do not improve are often placed on medicine to reduce inflammation. Many people will recover. A minority of people will worsen despite treatment. The goal of treatment is to maintain good lung function, lessen symptoms and prevent organ damage.

Corticosteroids - Corticosteroids, which suppress inflammation, are the main treatment. Generally, prednisone (a tablet) is given daily or every other day, depending on the symptoms. Corticosteroids often alleviate symptoms and/or improve or maintain lung function. In most cases corticosteroids reduce the long-term risk of organ scarring. Prednisone may be associated with a number of side effects. Because of this, people on corticosteroids should be carefully monitored by their doctor.

Methotrexate - Methotrexate is a very effective medication for the treatment of sarcoidosis. It may be used in place of or in combination with corticosteroids.

Other medicines and treatments - Other medicines are used if corticosteroids and methotrexate are not effective. These include Azathioprin, Leflunomide, Hydroxychloroquine, Mycophenolate, Colchicine, Minocycline and Infliximab.

Oxygen therapy may be part of a treatment plan for people with severe pulmonary sarcoidosis or pulmonary hypertension. Oxygen can help reduce heart and lung side effects of low oxygen levels. For people who develop chronic, progressive sarcoidosis, pulmonary rehabilitation may also be helpful.

One important thing a person can do to improve the outcome of sarcoidosis is to see a doctor when the symptoms first appear. This can help prevent irreversible damage to the lungs, eyes, heart, and other organs. Also, people with sarcoidosis should continue to follow up with their doctor after they have been diagnosed to monitor if the disease is progressing. People with sarcoidosis can, like others, develop lung disease from smoking. Therefore, it is also important to avoid smoking.

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