Sarcoidosis is a disease that occurs when areas of inflammation develop in different organs of the body. Very small clusters of inflammation, called granulomas, are seen with sarcoidosis. They may occur in the lungs, lymph nodes, eyes, skin or any area of the body. These granulomas may clear up on their own or cause permanent scarring.

The cause of sarcoidosis is unknown. Sarcoidosis occurs more often in Caucasians of European descent and in African-Americans. About 10 to 40 out of every 100,000 people develop sarcoidosis. 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, only vague symptoms of a general nature such as weight loss and fever, or symptoms associated with a specific organ, usually the lungs. More than one organ can be involved. Up to half of people with sarcoidosis have no symptoms when the illness is diagnosed. Thus, the diagnosis can be difficult to make.

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

Lungs
Inflammation in the lungs can cause shortness of breath, wheezing or cough (often a dry cough). In some people, the symptoms go away; in others there can be permanent scarring.

Lymph Nodes
Enlargement of various lymph nodes can occur, especially the lymph nodes in the chest.
**Eyes**
Inflammation of the eye can lead to watering, redness, and sensitivity to light. Blurred vision can also occur. In some cases there can be eye involvement with no obvious visual problems; therefore, it is important that the person be checked regularly by an eye doctor.

**Skin**
Skin involvement may appear as raised, pink or purplish areas or as painful nodules under the skin. These deeper nodules are often found on the legs and may be seen with arthritis from sarcoidosis.

**Bone**
Nodules in the bone can be painful and can cause pain in the hands and feet.

**Spleen and Liver**
Enlargement of the spleen or liver that a doctor can feel during a physical exam can occur.

**Heart**
Heart involvement occurs in a small percentage of people and can be difficult to diagnose. This can include heart rhythm abnormalities and can affect the ability of the heart muscle to pump blood.

**Brain and Nervous System**
Granulomas can develop in the brain and the nerves and cause many symptoms, including loss of sensation, loss of muscle strength, headaches, and dizziness. Only about one in 100 people with sarcoidosis are affected by neural sarcoidosis.

**Salivary Gland**
The salivary gland can also have granulomatous involvement. People with salivary involvement of their sarcoidosis may have trouble with a dry mouth.

**How is the diagnosis made?**
The first step in diagnosing sarcoidosis is a good evaluation. The inflamed microscopic granulomas seen in the affected organ with sarcoidosis are similar to those in other diseases such as tuberculosis, fungal diseases, berylliosis, and farmer's lung. Because of this, a careful evaluation is necessary to make a diagnosis. Only after the known causes of granulomas have been "ruled out" is the diagnosis of sarcoidosis made. An evaluation to detect sarcoidosis should include the following:

**Thorough Medical Examination**
A thorough medical examination can help "rule out" other diseases that may be similar to sarcoidosis.

**Chest X-Ray**
Doctors look at the x-ray for evidence of enlarged lymph nodes and small round spots in the lung caused by the clusters of inflammation. "Staging" can help the doctor determine the degree of lung involvement in sarcoidosis. A scale of 0-IV is commonly used with IV having the highest amount of lung involvement.

**Pulmonary Function Tests**
Standard breathing tests give an indication of the severity of lung disease. There is nothing unique about sarcoidosis on these tests, so they do not substitute for other, more specific tests. These tests can show obstruction of air flow out of the lungs, restriction of the lung's ability to take in air, and a decrease in the transport of oxygen from the lung into the blood stream. 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 should 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 can obtain this tissue. A bronchoscopy is an outpatient procedure in which the doctor places a narrow tube through the nose and into the airways. Sometimes the diagnosis is made by obtaining tissue samples from the skin, liver, or enlarged lymph nodes.

**Bronchoalveolar Lavage**
When a bronchoscopy is done, a small part of lung can be washed (lavaged) in order to obtain some cells of the immune system from the lung. By counting the types of cells in lavage fluid, it is possible to get an estimate of how inflamed the lungs are, and whether the type of inflammation is characteristic of sarcoidosis. This procedure is routinely used at a number of medical centers (including National Jewish Medical and Research Center), and may also provide a great deal of information for research into this disorder.

**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 nodes 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.

**Calcium Levels in the Blood and Urine**
People with sarcoidosis can have sensitivity to vitamin D. This results in too much calcium being absorbed through the intestines. This can cause a high level of calcium in the blood and urine. Exposure to the sun can 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. There are rarely symptoms seen with high levels of calcium in the blood. If symptoms are present they may include vague bone pain and frequent urination.

Kveim Test
The Kveim test is a test in which an extract of sarcoid affected tissue is injected under the skin. It is not used very often in a sarcoidosis evaluation.

**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, but some will get worse despite treatment. The goal of treatment is to maintain good lung function, lessen symptoms, and prevent organ damage.

**Corticosteroids** - Corticosteroids, which work to reduce inflammation, are the main treatment. Generally, prednisone (a tablet) is given daily or every other day, depending on the symptoms. It can decrease symptoms, improve lung function, decrease S.A.C.E. levels, reduce granuloma formation, and possibly, lessen scarring of the lungs. Prednisone can be associated with a number of side effects. Because of this, people on corticosteroids should be carefully monitored by a doctor. The potential benefits from treatment usually outweigh the risks from the medication side effects.

**Methotrexate** - When a person with sarcoidosis cannot take corticosteroids, there are other medicines which are used. Methotrexate is an anti-inflammatory medicine and is often used as a second-line medicine. It may be used with corticosteroids or after stopping corticosteroids.

**Other medicines and treatments** - Other medicines are used if corticosteroids and methotrexate are not effective. These other medicines are not used often, since their effect on sarcoidosis is not as well understood. They also can have side effects. These medicines include Azathioprin, Hydroxychloroquine, Chlorambucil, Cyclophosphamide and Pentoxifylline.

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

**Research at National Jewish Health**
National Jewish Health is currently involved with ongoing research regarding sarcoidosis. Our scientists and doctors are interested in the following areas of study:

- How genes and environmental exposures may interact to cause sarcoidosis in some people,
- What new medicines can better treat sarcoidosis,
- If there is more than one cause of sarcoidosis and
- What impact sarcoidosis has on the body over many years.

This research may be applied to develop better diagnosis and treatment for people with sarcoidosis.

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