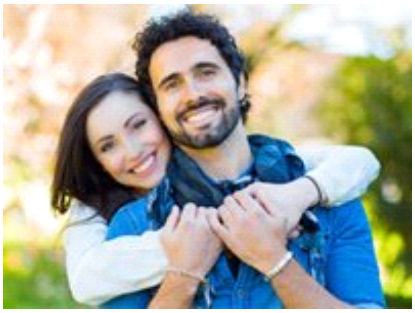
Sarcoidosis



Sarcoidosis is a chronic disease that can affect any organ in the body, but most commonly affects the lungs. Very small (microscopic) clusters of red and swollen tissue also called inflammatory cells or “granulomas” are seen in the organs affected with sarcoidosis. These “granulomas” may clear up on their own. Other times, the granulomas can be present and not cause major problems, although they can cause organ dysfunction, which can lead to scarring. There is no cure for sarcoidosis at this time. The disease can be managed and treated to minimize organ dysfunction (when the organ does not function properly).

Sarcoidosis Care at National Jewish Health

National Jewish Health is recognized by the World Association for Sarcoidosis and Other Granulomatous Diseases (WASOG) as a WASOG Sarcoidosis Clinic. This designation provides formal recognition of our team’s commitment to meet the needs of sarcoidosis patients and success in keeping up-to-date with ongoing new developments in the field. National Jewish Health is currently involved in multiple areas of ongoing sarcoidosis research. Results of this research will be used to develop better ways to diagnose, as well as treat people living with sarcoidosis.

Causes

Sarcoidosis is likely to have more than one cause, since not one single cause has been found. Sarcoidosis can be triggered by infections or other environmental exposures. Sarcoidosis is most common in young people between the ages of 20 and 40. However, it can affect any age group and race. About 10 to 40 of 100,000 people develop sarcoidosis. Sarcoidosis is not contagious.

What Are the Signs and Symptoms of Sarcoidosis?

Sarcoidosis symptoms can vary greatly, and up to half of people with sarcoidosis have no symptoms when the illness is diagnosed. A person with sarcoidosis may have:

- No symptoms (asymptomatic)
- Only vague symptoms of a general nature, such as weight loss, night sweats, fatigue and low grade fever. High fever is less common, but can still occur.
- Symptoms caused by the affected organ.

The lungs are the most common organ affected by sarcoidosis. However, any organ can be affected. More than one organ can be involved. Signs and symptoms associated with specific organ involvement can include:

Lungs

Inflammation in the lungs can cause shortness of breath, wheezing or cough (often a dry cough). In some people, the symptoms may resolve. In others, there can be continued inflammation that can lead to permanent scarring and persistent symptoms.

Lymph Nodes

Enlargement of various lymph nodes can occur, especially the lymph nodes in the chest, although these may not cause symptoms. Sometimes, lymph node enlargement and inflammation can be associated with pain.

Eye

Inflammation of the eye can lead to redness, pain, dry eyes and sensitivity to light, and can also cause some more serious issues with blurred vision or vision loss. It is important that an eye doctor perform an eye exam regularly to determine if there is eye involvement.

Skin

Sarcoidosis skin involvement may appear as raised, pink or purplish areas or as painful nodules under the skin.

Bone

Bone involvement is often detected incidentally (something extra found by a test). Sometimes it can cause pain, but very rarely fractures.

Spleen and Liver

Enlargement of the spleen or liver that a doctor can feel during a physical exam can occur. These organs can be involved frequently, but rarely cause serious issues and rarely require treatment.

Heart

Heart involvement is thought to occur in up to 25% to 40% of people with sarcoidosis. It can be difficult to diagnose. Heart involvement can occur without symptoms. It can also appear with heart rhythm abnormalities (too fast or too slow). These can cause heart palpitations and lightheadedness. Heart involvement can also lead to problems with the heart muscle's ability to pump blood or heart failure.

Brain and Nervous System

Granulomas can develop in the brain and the nerves and cause many symptoms. Symptoms may include loss of sensation, loss of muscle strength, headaches and dizziness. This affects about one in 100 people with sarcoidosis.

Salivary Gland

The salivary gland can be involved, and can be associated with swelling. People with salivary involvement may have trouble with a dry mouth.

Kidneys and Calcium

Kidney involvement is not common, although kidney damage can occur. In addition, calcium levels may be elevated in the urine, leading to kidney stones. High calcium levels in the blood can be more serious and cause constipation and generalized weakness.

How Is Sarcoidosis Diagnosed?

The first step in diagnosing sarcoidosis is a thorough evaluation. The granulomas in the organ affected with sarcoidosis are similar to those in other diseases such as tuberculosis or fungal infections, berylliosis and farmer's lung. Because of this, a careful evaluation is important to rule out other diagnoses that can look like sarcoidosis. 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: This can help rule out other diseases that may be similar to sarcoidosis. Chest X-ray: Doctors look at chest X-rays 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 – 4 is commonly used, with 4 having the highest amount of lung involvement.

Pulmonary Function Tests: These 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. Pulmonary function tests can show obstruction of airflow 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 bloodstream. 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 is done.

Tissue Biopsy: A microscopic exam of tissue samples from the lungs or other affected organs may also be needed to be sure of the diagnosis and to exclude other causes. A bronchoscopy can obtain this tissue. A bronchoscopy is a 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 other organs such as the skin, liver or enlarged lymph nodes.

Bronchoalveolar Lavage: When a bronchoscopy (a procedure that looks inside the lungs and airways) is done, a small part of lung can be washed (lavaged) with sterile saline solution. This lets the doctor see what cells of the immune system are in 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.

Eye Exam: A slit lamp exam by an eye doctor (ophthalmologist) is an important part of an eye exam to detect inflammation.

CT (Computerized Tomography) 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.

Calcium Levels in the Blood and Urine: Regulation of calcium may be disturbed in sarcoidosis. This results in too much calcium in the blood and/or urine. Exposure to the sun, calcium and vitamin D supplements and high dairy intake can stimulate this process. A 24-hour urine sample can measure the calcium level in the urine. A blood sample can be drawn for levels of calcium in the blood. High calcium levels in the blood can be seen with symptoms that may include fatigue, abdominal pain/constipation and mental fogginess. High calcium levels can also lead to kidney dysfunction.

PET (Positron Emission Tomography) Scan: A radioactive labeled sugar is given intravenously. The person is placed in a special scanning machine to see where the radioactive labeled sugar accumulated. Areas of active inflammation take up the radioactive labeled sugar and are detected with the scanning machine. This gives your doctor a better idea of which areas or organs might be involved with sarcoidosis.

Heart Testing: The doctor may order an electrocardiogram (EKG) to evaluate the electrical system of the heart. An echocardiogram may also be done to evaluate the structure and function of the heart. Sometimes, the doctor might order further tests for the heart if there is suspicion of heart involvement with sarcoidosis. These tests might include a Holter monitor and/or a heart MRI (magnetic resonance imaging).

What is the Treatment for Sarcoidosis?

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 goals of treatment are to:

- Maintain good organ function
- Lessen symptoms
- Prevent organ damage

Medication for Sarcoidosis

Several medicines are used to treat sarcoidosis.

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. Prednisone can decrease symptoms, improve lung function, reduce granuloma formation, and possibly lessen scarring of the lungs. Prednisone can be associated with a number of side effects. Because of this, your doctor will carefully monitor you. Prednisone is not the medication of choice for long-term management of sarcoidosis.

Methotrexate: For long-term management of sarcoidosis, steroid-sparing agents are often used. Methotrexate is an anti-inflammatory medicine. It is often used as a second-line medicine. It may be used with corticosteroids or after stopping corticosteroids.

Other Medicines: 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 can include:

- Cellcept® (mycophenolate)
- Humira® (adalimumab)
- Imuran® (azathioprine)
- Plaquenil® (hydroxychloroquine)
- Remicade® (infliximab)

Oxygen Therapy

Oxygen therapy may be an important part of a treatment plan for people with severe sarcoidosis. It can help reduce heart and lung long-term problems caused by low oxygen levels.

Pulmonary Rehabilitation

For people who develop chronic, progressive sarcoidosis, pulmonary rehabilitation also may be helpful. This includes exercise, healthy eating and education. Because treatment is so important, a person can improve the outcome of sarcoidosis by seeing 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.

Lifestyle Management

Calcium and Vitamin D The primary source of vitamin D in our bodies comes from our diet. In addition, vitamin D is produced in our skin when exposed to the sun. Vitamin D from our diet or skin goes through an initial activation step in the liver where an enzyme (25-hydroxylase) converts vitamin D into 25-hydroxy-vitamin D. The next step in activation occurs in the kidneys where another enzyme (1-alpha-hydroxylase) converts 25-hydroxy-vitamin D into 1,25 dihydroxy-vitamin D (1,25 vit D), which is the active form. The 1,25 vit D regulates calcium levels in the blood by enhancing calcium absorption from our intestines and by increasing bone resorption to maintain a normal calcium level in the blood.

In patients with sarcoidosis, the granulomas contain cells called macrophages. Macrophages can have the enzyme 1-alpha-hydroxylase that converts vitamin D to its final active product, which then works to increase calcium levels in the blood and/or urine. About 5 percent of sarcoidosis patients have elevated calcium levels in their blood, and about 15 percent have elevated calcium levels in their urine. Chronically elevated calcium levels can increase the risk of developing kidney stones, can potentially affect kidney function, and weaken bones.

Your doctor usually checks your blood calcium level by doing routine blood tests and your urine calcium levels by checking the calcium levels in a 24 hour collection of urine. Your doctor may perform other tests to make sure there is no other cause for your elevated calcium levels.

If the calcium level is elevated in the blood and/or urine, then initial conservative measures such as decreasing vitamin D and calcium intake from diet or supplements and avoiding excessive sun exposure can potentially reduce the calcium levels. If these measures do not control the calcium level, then your doctor might start a medication to help control the calcium levels.

At this time, we have no definitive studies that recommend any particular type of nutrition. A diet of fruits, vegetables with less processed foods and sugars, can help reduce inflammation. Exercise, as with many other diseases, can have significant beneficial effects. Getting good amounts of quality sleep is known to be important for the immune system function. Treating any underlying sleep disorder is also recommended.

Visit our website for more information about support groups, clinical trials and lifestyle information.