Patient education: Sarcoidosis (Beyond the Basics)

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INTRODUCTION

Sarcoidosis is a disorder that causes tiny nodules (granulomas) of inflamed tissue to develop in the body's organs. These nodules can join together, forming larger nodules that interfere with normal body functions such as breathing. Sarcoidosis almost always involves the lungs, but it can also affect the skin, eyes, nose, muscles, heart, liver, spleen, bowel, kidney, testes, nerves, lymph nodes, joints, and brain. Granulomas in the lungs can cause narrowing of the airways and also inflammation and scarring (fibrosis) of lung tissue.

SARCOIDOSIS CAUSES

The cause of sarcoidosis is not known. One theory suggests that it develops when a genetically susceptible person is exposed to specific environmental agents. Although the specific agents are unknown, it is likely that there is more than one trigger around the world.

Noninfectious chemicals in the work environment, including beryllium, aluminum, and zirconium, can cause lung disease that has features similar to sarcoidosis.

SARCOIDOSIS RISK FACTORS

Sarcoidosis occurs throughout the world, affecting both sexes and all races and ages. In a small percentage of cases, more than one family member is affected. Within the United States, Black people are two to three times more likely to develop sarcoidosis and may have

more severe disease than White people. Sarcoidosis is slightly more common in females compared with males and rarely occurs in children.

SARCOIDOSIS SIGNS AND SYMPTOMS

Sarcoidosis frequently causes mild symptoms and resolves on its own. The most common symptoms of lung involvement are cough or shortness of breath. Affected individuals may also experience fatigue, chest pain, weakness, fever, and weight loss.

Other organs in the body can also be affected. The signs and symptoms in these organs depend upon the site and extent of organ involvement (eg, involvement of the heart can cause palpitations, dizziness, chest pain and/or heart failure). The following organ systems are commonly involved.

Skin — Skin lesions of different types can occur on the face, neck, arms, legs, or trunk. These lesions include subtle, painless rashes, red, red-brown, purple, or nonpigmented bumps that may be painful, and scarring lesions. People with more severe disease involving the internal organs often have more severe skin lesions.

Eyes — Sarcoid of the eyes can cause inflammation of different eye structures, including the iris, retina, or cornea. Symptoms include eye pain or redness, intolerance of bright lights, dry eyes, blurred vision, floaters, and swelling around the eye. Glaucoma, cataracts, and blindness are late complications of untreated sarcoidosis. Because some sarcoidosis-related eye problems do not cause symptoms, it is important that all patients with sarcoidosis have at least one eye evaluation that includes an examination while the eyes are dilated. New changes in vision should be evaluated promptly in any patient with sarcoidosis.

Kidney — Abnormalities in the way the body handles calcium can occur and, if untreated, it may rarely lead to kidney failure. Small nodules (granulomas) may also develop in the kidney, leading to abnormal kidney function. Patients with sarcoidosis should have kidney function testing (usually with blood and/or urine tests) as part of their initial evaluation and follow up testing.

Heart — Nodules may develop in the heart, interfering with its electrical conduction system. This can result in abnormal heart rhythms, palpitations, dizziness, fainting, and even death. An electrocardiogram (also referred to as an ECG or EKG) can generally detect abnormalities in the heart's electrical conduction. It may also cause weakening of the heart muscle and heart failure. Damage and scarring of the lung and lung blood vessels (called pulmonary hypertension) rarely makes it more difficult for the heart to pump blood through the lungs. This condition can lead to failure of the heart's right ventricle.

Nervous system — Neurologic involvement affects approximately 5 percent of patients with sarcoidosis and may be the first sign of the condition. Potential symptoms include headache, confusion, localized weakness or paralysis, seizures, and fatigue. Patients with sarcoid of the pituitary may have abnormal menstrual cycles, excessive thirst or frequent urination. Sarcoid meningitis (inflammation of the membranes covering the base of the brain) can cause loss of taste or smell, blurred vision, and/or facial weakness or paralysis. The disease may also affect the nerves in the arms and legs, resulting in muscle weakness, numbness or tingling, and pain.

Musculoskeletal system — Ten to 15 percent of people have sarcoid arthritis (with joint pain and swelling), changes in bone structure, or muscle discomfort and pain.

Reproductive system — Sarcoidosis can affect the male reproductive system, particularly the testes, and may cause male infertility. The disease rarely affects the female reproductive system. Sarcoidosis does not increase the risk of complications during pregnancy; however, the disease may worsen after the birth of the child. Therefore, follow-up evaluation of sarcoidosis is recommended for women with sarcoidosis within six months after delivery.

Other organs — Enlargement of lymph nodes, especially those in the chest, occurs frequently. The liver or spleen can also be affected. Involvement of the spleen may lead to anemia and other blood abnormalities. Sarcoid of the nose and sinuses can cause nasal obstruction, nasal crusting, and loss of sense of smell. Nasal bleeding and nasal polyps are less common.

SARCOIDOSIS DIAGNOSIS

There is no single test that confirms a diagnosis of sarcoidosis, so the diagnosis is based upon multiple factors, including symptoms, physical examination, abnormalities on chest X-ray (or computed tomography [CT] scan), and microscopic examination of one or more specimens from involved tissues or organs. In addition, the diagnostic process often involves tests that help to rule out other conditions, including tuberculosis and fungal infection, which share some features with sarcoidosis. (See "Patient education: Tuberculosis (Beyond the Basics)".)

A biopsy involves removing a small sample of tissue from an affected tissue or organ. This test is usually recommended to identify a granuloma. Samples can be obtained from lung tissue with a procedure called bronchoscopy. A biopsy may also be done on an affected lymph node, skin nodule, salivary gland, or the tear gland near the eye. (See "Patient education: Flexible bronchoscopy (Beyond the Basics)".)

Once the diagnosis of sarcoidosis is confirmed, other tests may be needed to determine the extent and severity of the disease. This may include imaging tests such as an MRI and/or PET scan to determine if there is evidence of the disease in the eyes, heart, or other organs.

SARCOIDOSIS TREATMENT

Because the cause of sarcoidosis is not known, there is no specific treatment. Some medications are effective in suppressing symptoms, and these are discussed below. Fortunately, many individuals with sarcoidosis do **not** require treatment since the nodules (granulomas) often resolve without treatment and leave behind few, if any, signs of inflammation or other complications.

There are many questions about the best time to start treatment for sarcoidosis and how long it should be continued. Treatment is usually recommended in patients with worsening lung problems, especially shortness of breath and cough or lung function (as determined by pulmonary function testing). Sarcoidosis affecting the eyes, heart, or kidneys is treated even when symptoms are mild because of the potentially serious risk of complications when these systems are involved. Other reasons for treatment include difficulty functioning due to fever, weakness, fatigue, joint pain, nervous system changes, disfiguring skin disease, or disease affecting the upper airway.

Current treatment is focused on improving symptoms, suppressing inflammation, reducing the impact of the granulomas, and preventing the development of lung fibrosis.

Glucocorticoids — Glucocorticoids (also known as steroids), most commonly prednisone, are particularly effective in reducing inflammation and are typically the first line treatment. In patients with mild disease, such as skin lesions, eye inflammation, or cough, topical glucocorticoid therapy with creams, eye-drops, or inhalers may be sufficient to control the disease.

When necessary, oral glucocorticoids are generally taken for 6 to 12 months. A relatively higher dose is usually recommended at first (usually 20 mg/day), followed by a slow taper to the lowest effective dose. Relapses may occur after glucocorticoid treatment has ended, although relapses usually respond to restarting treatment with glucocorticoids. Patients who improve and remain stable for more than one year after glucocorticoid treatment have a low risk of relapse.

Symptoms of sarcoidosis, especially cough and shortness of breath, generally improve with glucocorticoid therapy. However, there are potentially serious side effects of long-term glucocorticoids, and the benefits must be weighed against the risks. These include increased appetite, weight gain, acne, fluid retention, trembling, mood swings, and difficulty sleeping.

If glucocorticoids are taken for long periods of time, particularly if high doses are used, there is an increased risk of developing diabetes, thinning of the skin, easy bruising, a "cushingoid" appearance (widening of the face and a hump in the back), thinning of the bones, body hair growth, cataracts, high blood pressure, stomach ulcers, avascular necrosis (a serious joint problem), and infections. Because of the risk of these side effects, most patients are tapered off of glucocorticoids as soon as possible.

Researchers continue to examine the role of glucocorticoids in the treatment of sarcoidosis. The biggest question is what effect these drugs have on the long-term course of the disease.

Other therapies — Other therapies may be recommended for people who cannot tolerate glucocorticoids, do not respond to glucocorticoids, or want to avoid/reduce glucocorticoid side effects.

- **Methotrexate:** This drug reduces inflammation and suppresses the immune system and may allow for a lower dose of glucocorticoids to be used.
- **Azathioprine**, **leflunomide**, and **mycophenolate mofetil**: These drugs are sometimes used in conjunction with glucocorticoids if the condition is worsening despite treatment. They are often used instead of methotrexate as there are no definitive data to indicate which steroid-sparing option is best. These drugs work by suppressing the immune system.
- Cyclophosphamide: Due to its severe toxic effects, including the risk of cancer, cyclophosphamide is reserved for patients with the most severe disease.
- **Antimalarial medications**: Drugs such as **hydroxychloroquine** or **chloroquine** have been used to treat sarcoidosis affecting the skin, joints, and lungs or resulting in calcium abnormalities.
- **Nonsteroidal anti-inflammatory agents** (NSAIDs, including ibuprofen): NSAIDs may help reduce inflammation and relieve joint pain, swelling, and fever, although they are not recommended for the treatment of sarcoidosis affecting the lungs.
- **Tumor necrosis factor (TNF) antagonists: TNF antagonists** are medications that were originally designed for treatment of rheumatoid arthritis. They work by interfering with the production of certain proteins involved in inflammation. Drugs in this class include infliximab (Remicade) and adalimumab (Humira). Tumor necrosis factor antagonists are particularly useful for skin, neurologic, and ocular sarcoidosis, but have also been effective for refractory disease of the lungs, heart, and other organs. They are widely considered as third-line agents.

Anti-TNF medications may be used alone or in combination with methotrexate, azathioprine, and/or glucocorticoids. All anti-TNF treatments must be injected.

- A **cardiac pacemaker** or **intracardiac defibrillator** may be needed in patients with cardiac sarcoidosis.

Investigational agents — Information about clinical trials of investigational agents for sarcoidosis is available from the National Institutes of Health clinical trials registry.

SARCOIDOSIS LONG-TERM OUTCOME

In many patients, sarcoidosis resolves on its own or does not progress. In other patients, sarcoidosis may progress over many years and involve many organs. However, the overall death rate directly from sarcoidosis is less than 5 percent. Death most commonly results from progressive lung scarring, sometimes complicated by right heart failure or bleeding from the lungs, but can also result from sarcoidosis involving the heart.

WHERE TO GET MORE INFORMATION

Your healthcare provider is the best source of information for questions and concerns related to your medical problem.

This article will be updated as needed on our web site (www.uptodate.com/patients). Related topics for patients, as well as selected articles written for healthcare professionals, are also available. Some of the most relevant are listed below.

Patient level information — UpToDate offers two types of patient education materials.

The Basics — The Basics patient education pieces answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials.

Patient education: Sarcoidosis (The Basics)
Patient education: Erythema nodosum (The Basics)
Patient education: Interstitial lung disease (The Basics)
Patient education: Endobronchial ultrasound (The Basics)

Beyond the Basics — Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are best for patients who want in-depth information and are comfortable with some medical jargon.

Patient education: Tuberculosis (Beyond the Basics)
Patient education: Flexible bronchoscopy (Beyond the Basics)
Patient education: Disease-modifying antirheumatic drugs (DMARDs) in rheumatoid arthritis (Beyond the Basics)

Professional level information — Professional level articles are designed to keep doctors and other health professionals up-to-date on the latest medical findings. These articles are thorough, long, and complex, and they contain multiple references to the research on which they are based. Professional level articles are best for people who are comfortable with a lot of medical terminology and who want to read the same materials their doctors are reading.

Clinical manifestations and diagnosis of cardiac sarcoidosis
Clinical manifestations and diagnosis of sarcoidosis
Gastrointestinal, hepatic, pancreatic, and peritoneal sarcoidosis
Evaluation of the adult patient with hepatic granuloma
High resolution computed tomography of the lungs
Hypercalcemia in granulomatous diseases
Neurologic sarcoidosis
Pathology and pathogenesis of sarcoidosis
Kidney disease in sarcoidosis
Sarcoid arthritis
Sarcoid myopathy
Treatment of pulmonary sarcoidosis refractory to initial therapy
Treatment of pulmonary sarcoidosis: Initial approach

The following organizations also provide reliable health information.

- National Library of Medicine

(https://medlineplus.gov/sarcoidosis.html)

- National Heart, Lung, and Blood Institute

(https://www.nhlbi.nih.gov/health/sarcoidosis)

- American Lung Association

(https://www.lung.org/lung-health-diseases/lung-disease-lookup/sarcoidosis)

- Foundation for Sarcoidosis Research

(www.stopsarcoidosis.org)

- The American Thoracic Society

(https://www.thoracic.org/patients/patient-resources/resources/what-is-sarcoidosis.pdf)

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