

## **Sarcoidosis Fact Sheet**

### **1. What Is Sarcoidosis?**

Sarcoidosis involves inflammation that produces tiny lumps of cells in various organs in your body. The lumps are called granulomas because they look like grains of sugar or sand. They are very small and can be seen only with a microscope. These tiny granulomas can grow and clump together, making many large and small groups of lumps. If many granulomas form in an organ, they can affect how the organ works. This can cause symptoms of sarcoidosis.

Sarcoidosis can occur in almost any part of your body, although it usually affects some organs more than others. It usually starts in one of two places:

- Lungs
- Lymph nodes, especially the lymph nodes in your chest cavity

Sarcoidosis also often affects your skin, eyes and liver.

It can also affect your spleen, brain, nerves, and heart.

Sarcoidosis usually occurs in more than one organ at a time.

The course of the disease varies greatly among people.

- In many people, sarcoidosis is mild. The inflammation that causes the granulomas may get better on its own. The granulomas may stop growing or shrink. Symptoms may go away within a few years.
- In some people, the inflammation remains but doesn't get worse. You may also have symptoms or flare-ups and need treatment every now and then.
- In other people, sarcoidosis slowly gets worse over the years and can cause permanent organ damage. Although treatment can help, sarcoidosis may leave scar tissue in the lungs, skin, eyes, or other organs. The scar tissue can affect how the organs work. Treatment usually does not affect scar tissue.

Changes in sarcoidosis usually occur slowly (e.g., over months). Sarcoidosis does not usually cause sudden illness. However, some symptoms may occur suddenly. They include:

- Disturbed heart rhythms
- Arthritis in the ankles
- Eye symptoms

In very rare cases in which vital organs are affected, sarcoidosis can be fatal.

Sarcoidosis is not a form of cancer. There is no known way to prevent the disease.

Sarcoidosis was once thought to be an uncommon condition. It's now known to affect tens of thousands of people throughout the U.S. Because many people who have sarcoidosis have no symptoms, it's hard to know how many people have the condition.

Sarcoidosis was identified in the late 1860s. Since then, scientists have developed better tests to diagnose it and made advances in treating it.

## **2. What Causes Sarcoidosis?**

The cause of sarcoidosis is not known. And, there may be more than one thing that causes it.

Scientists think that sarcoidosis develops when your immune system responds to something in the environment (e.g., bacteria, viruses, dust, chemicals) or perhaps to your own body tissue (autoimmunity).

Normally, your immune system defends your body against things that it sees as foreign and harmful. It does this by sending special cells to the organs that are being affected by these things. These cells release chemicals that produce inflammation around the foreign substance or substances to isolate and destroy them.

In sarcoidosis, this inflammation remains and leads to the development of granulomas or lumps.

Scientists have not yet identified the specific substance or substances that trigger the immune system response in the first place. They also think that sarcoidosis develops only if you have inherited a certain combination of genes.

You can't catch sarcoidosis from someone who has it.

More research is needed to discover what causes sarcoidosis.

## **3. Who Gets It?**

Sarcoidosis affects people of all ages and races worldwide.

It occurs mostly in:

- Adults between the ages of 20 and 40
- African Americans (especially women)

- People of Asian, German, Irish, Puerto Rican and Scandinavian origin

In the United States, sarcoidosis affects African Americans somewhat more often and more severely than Caucasians.

Studies have shown that sarcoidosis is more likely to affect certain organs in certain populations. For example,

- Sarcoidosis of the heart and eye appears to be more common in Japan.
- Painful skin lumps on the legs occur more often in people from Northern Europe.

People who are more likely to get sarcoidosis include:

- Health care workers
- Nonsmokers
- Elementary and secondary school teachers
- People exposed to agricultural dust, insecticides, pesticides or mold
- Firefighters

Brothers and sisters, parents and children of people who have sarcoidosis are more likely than others to have sarcoidosis.

#### **4. What Are the Signs and Symptoms?**

Many people who have sarcoidosis have no symptoms. Often, the condition is discovered by accident only because a person has a chest x ray for another reason, such as a pre-employment x ray.

Some people have very few symptoms, but others have many.

Symptoms usually depend on which organs the disease affects.

##### ***Lung Symptoms***

- Shortness of breath
- A dry cough that doesn't bring up phlegm or mucus
- Wheezing
- Pain in the middle of your chest that gets worse when you breathe deeply or cough (rare).

### *Lymph Node Symptoms*

- Enlarged and sometimes tender lymph nodes—most often those in your neck and chest but sometimes those under your chin, in your arm pits, or in your groin.

### *Skin Symptoms*

- Various types of bumps, ulcers, or, rarely, flat areas of discolored skin, that appear mostly near your nose, eyes, back, arms, legs, and scalp. They usually itch but aren't painful. They usually last a long time.
- Painful bumps that usually appear on your ankles and shins and can be warm, tender, red or purple-to-red in color, and slightly raised. You may have fever and swollen ankles and joint pain along with the bumps. The bumps often are an early sign of sarcoidosis, but they occur in other diseases too. The bumps usually go away in weeks to months, even without treatment.
- Disfiguring skin sores that may affect your nose, nasal passages, cheeks, ears, eyelids, and fingers. The sores tend to be ongoing and can return after treatment is over.

### *Eye Symptoms*

- Burning, itching, tearing, pain
- Red eye
- Sensitivity to light
- Dryness
- Floaters (i.e., seeing black spots)
- Blurred vision
- Reduced color vision
- Reduced visual clearness
- Blindness (in rare cases)

## **5. How Is Sarcoidosis Diagnosed?**

Your doctor will find out if you have sarcoidosis by taking a detailed medical history and conducting a physical exam and several diagnostic tests. The purpose is to:

- Identify the presence of granulomas in any of your organs
- Rule out other causes of your symptoms
- Determine the amount of damage to any of your affected organs
- Determine whether you need treatment.

Your doctor may not need to find every one of your organs affected by sarcoidosis, only those that cause symptoms. Often the organs affected by the condition continue to function well and don't need to be treated.

## 6. How Is Sarcoidosis Treated?

The goals of treatment are to:

- Improve how the organs affected by sarcoidosis work
- Relieve symptoms
- Shrink the granulomas

Your treatment depends on:

- What symptoms you have
- How severe your symptoms are
- Whether any of your vital organs (e.g., your lungs, eyes, heart, or brain) are affected
- How the organ is affected.

Some organs must be treated, regardless of your symptoms. Others may not need to be treated. Usually, if you don't have symptoms, you don't need treatment, and you probably will recover in time.

### *Drugs*

The main treatment for sarcoidosis is prednisone. Prednisone is a corticosteroid, or anti-inflammatory drug. Sometimes it is used with other drugs. Sometimes other corticosteroids are used.

### *Other Drugs Used To Treat Sarcoidosis*

Other drugs are sometimes used to treat sarcoidosis. Your doctor may prescribe one of them if:

- Your condition gets worse while you are taking prednisone
- You can't stand the side effects of prednisone.

Most of these other drugs are immune system suppressants.

The other drugs used to treat sarcoidosis include:

- Hydroxychloroquine (Plaquenil)

- Methotrexate
- Azathioprine (Imuran)
- Cyclophosphamide (Cytosan)

### ***What Does the Future Hold?***

Scientists worldwide are trying to learn more about sarcoidosis and how to improve its diagnosis and treatment. Some recent studies have led to possible new treatments, which, in turn, are being studied. Current research includes studies of:

- The agent or agents that cause sarcoidosis
- Why sarcoidosis seems to act differently in people of different races
- Why sarcoidosis appears in some families
- How genes, passed from one generation to another, may make some people more likely than others to develop sarcoidosis
- How cells act and communicate with each other to cause sarcoidosis symptoms.

## **7. What May Help Make It Easier to Live with Sarcoidosis?**

You should take steps to stay healthy. This includes:

- Don't smoke.
- Avoid substances like dusts and chemicals that can harm your lungs.
- Try to follow a healthy eating plan.
- Be as active as you can but don't strain yourself.

Joining a patient support group may help you adjust to living with sarcoidosis. Talking to others who have the same symptoms can help you see how they have coped with them.

Your regular doctor may be able to diagnose and treat your sarcoidosis, but diagnosis and treatment by a doctor who specializes in sarcoidosis is recommended. If you prefer to use your regular doctor, you should see a doctor who specializes in the organs that are affected by your sarcoidosis at least once. For example, see an ophthalmologist if your eyes are affected or a pulmonologist if you have sarcoidosis in your lungs. These specialists will work with your regular doctor to help make a diagnosis, develop a treatment plan and schedule periodic exams and lab tests.

### *Follow-Up Care*

Regular follow-up care is important, even if you aren't taking medication for your sarcoidosis. New symptoms can occur at any time, and your condition can get worse slowly, without your noticing.

Follow-up exams usually include:

- A review of your symptoms
- A physical exam
- A chest x ray and CT scan
- Breathing tests
- An eye exam
- Blood tests
- An electrocardiogram (EKG).

How often you have your examinations and tests depends on:

- How severe your symptoms are
- Which organs were affected at diagnosis
- What treatment you are using
- Any complications that may develop during treatment.

You will probably need routine follow-up care for several years. Whether you see your regular doctor or a sarcoidosis specialist for this depends on your symptom during the first year of follow-up.

This information was adapted from the National Heart Lung and Blood Institute. For more information about sarcoidosis, visit [http://www.nhlbi.nih.gov/health/dci/Diseases/sarc/sar\\_what.html](http://www.nhlbi.nih.gov/health/dci/Diseases/sarc/sar_what.html).

The National Heart Lung and Blood Institute is part of the National Institutes of Health and the U.S. Department of Health and Human Services.