

## Cardiac Sarcoidosis

### What Is Cardiac Sarcoidosis?

Sarcoidosis is a poorly understood disease that commonly affects the lungs. It can also involve the lymph nodes, liver, spleen, eyes, skin, bones, salivary glands and heart. Cardiac sarcoidosis is estimated to be present in 10 to 25 percent of people with sarcoidosis.

Sarcoidosis is characterized by the presence of granulomas. These are ball-like collections of inflammatory cells that cluster around and react to a foreign substance. The inflammation associated with granulomas can damage virtually every part of the heart, including the electrical system, muscle, valves, arteries and surrounding tissue called the pericardium. It is important to remember, sarcoidosis of the heart can precede, follow, or occur as the same time as other non-cardiac forms of sarcoidosis. The complications from heart involvement of sarcoidosis may occur suddenly and without warning.



### What Causes Cardiac Sarcoidosis?

The exact cause of sarcoidosis is unknown. Granulomas appear to be the result of an immune system response to an unidentified trigger. Infectious agents as well as environmental exposures are thought to be possible precipitants for this immune response; however, no clear triggers or causes of sarcoidosis have been identified. Genetic mutations in white blood cell proteins (called human leukocyte antigens, or HLA) as well as chemicals that control inflammation (called cytokines) have also been linked to sarcoidosis.

### How Can Sarcoidosis Affect the Heart?

Cardiac sarcoidosis can take many forms, some benign and others serious. These include:

**Heart Rhythm Disorders:** A complete block of electrical movement through the heart is the most common form of sarcoidosis of the heart. Alternatively, fast heart rhythms such as atrial flutter, atrial fibrillation, supraventricular tachycardia and ventricular tachycardia can be present. Ventricular tachycardia occurs in almost 25 percent of people with cardiac sarcoidosis and is of particular

concern since it can lead to sudden cardiac death. These possible complications may occur suddenly and without warning.

**Heart Failure:** Heart failure is another complication of cardiac sarcoidosis. Sarcoidosis can cause the heart muscle to weaken and/or stiffen. This leads to fluid retention in the lungs, abdomen and lower extremities. In extreme cases, an aneurysm can form due to weakening of the heart wall. Granulomas can also infiltrate the heart valves, causing leaky valves, also resulting in heart failure.

**Coronary Disease:** Although rare, sarcoidosis of the heart can cause an inflammatory disorder of the heart arteries called vasculitis. In severe forms, vasculitis can lead to coronary artery blockages, chest pain, and ultimately heart attacks.

**Pericardial Disease:** Inflammation of the sac around the heart, called pericarditis, is another rare but important form of cardiac sarcoidosis.

## How Is Cardiac Sarcoidosis Diagnosed?

Diagnosing cardiac sarcoidosis can be very challenging. There are no widely accepted guidelines for either screening or diagnosing sarcoidosis of the heart. Moreover, the current available diagnostic tests are variable in their ability to detect cardiac sarcoidosis. Because of the devastating nature of cardiac sarcoidosis, most people with other forms of sarcoidosis are screened for sarcoidosis of the heart.

Initial cardiac evaluation may include an electrocardiogram (EKG), a signal-averaged EKG, an echocardiogram (ultrasound of the heart) and a Holter monitor (extended EKG). Additional imaging tests may include single positron emission computed tomography (SPECT), positron emission tomography (PET), and cardiac MRI. A positive heart biopsy confirms cardiac sarcoidosis, but may more often be negative or normal even when there is sarcoidosis in the heart, especially if heart function is normal.

## What Is the Treatment?

Controversy exists as to the best treatment for cardiac sarcoidosis. However, treatment is often directed at minimizing the inflammation seen with cardiac sarcoidosis and protecting against the life-threatening complications. Because of their anti-inflammatory properties, corticosteroids (cortisone, prednisone, and methylprednisolone) are the first-line therapy to treat the inflammation. When people cannot take steroids, or when combination therapy is needed, other medications are used. These include: methotrexate, azathioprine, mycophenolate and antimalarials.

Additional therapies for specific cardiac sarcoidosis related heart disorders may be necessary. For example, heart rhythm disorders such as complete heart block typically require placement of a permanent pacemaker, whereas ventricular tachycardia generally requires internal cardiac defibrillator (ICD) placement.

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