

# MEDfacts

An Educational Health Series From National Jewish Health®



## 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 (CS) is estimated to be present in 10-25% of people with sarcoidosis. Sarcoidosis is characterized by the presence of granulomas. These are ball-like collections of white blood cells that cluster around and react to a foreign substance. The inflammation associated with granulomas can damage every part of the heart, including the electrical system, muscle, valves, arteries and surrounding tissue called the pericardium. It is important to remember CS can precede, follow, or occur as the same time as other non-cardiac forms of sarcoid.

### 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. 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 CS. Alternatively, fast heart rhythms such as atrial flutter, atrial fibrillation, supraventricular tachycardia, and ventricular tachycardia can be present. Ventricular tachycardia occurs in almost 25% of people with CS and is concerning since it can lead to sudden cardiac death.

**Heart Failure:** Heart failure is the second most common form of CS. Sarcoidosis can cause the heart muscle to weaken and/or

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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 hearts valves, causing leaky valves also resulting in heart failure.

**Coronary Disease:** Although rare, CS 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 sack around the heart, called pericarditis, is another rare but important form of CS.

### How is Cardiac Sarcoidosis Diagnosed?

Diagnosing CS can be very challenging. There are no widely accepted guidelines for either screening or diagnosing CS. Moreover, the current available diagnostic tests are variable in their ability to detect CS. Because of its devastating nature, most people with other forms of sarcoidosis are screened for CS. Initial cardiac evaluation may include an EKG, a signal averaged EKG, and an echocardiogram. Additional imaging tests may include single positron emission computed tomography (SPECT), positron emission tomography (PET), and cardiac MRI. A heart biopsy is the most definitive diagnostic test, but is accurate in only 63% cases.

### What is the Treatment?

Controversy exists as to the best treatment for CS. However, treatment is often directed at minimizing the inflammation seen with CS. 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 CS-related heart disorders may be necessary. For example, heart rhythm disorders such as complete heart block often require placement of a permanent pacemaker. Ventricular tachycardia often requires placement of an internal cardiac defibrillator (ICD).

### What do we do at National Jewish Health?

We provide comprehensive cardiology evaluation and consultation and non-invasive cardiac testing. We evaluate and treat heart problems such as coronary artery disease, high blood pressure, high cholesterol, heart valve problems and heart failure. In addition to traditional heart problems, we offer expertise in many other focus areas, including evaluation of patients with shortness of breath with exercise, sarcoid of the heart, diastolic dysfunction and secondary pulmonary hypertension.

### Why National Jewish Health?

At National Jewish Health, we treat the whole person, not just the disease. Our cardiology team works with healthcare providers from all areas of the medical center, including rehabilitation therapists, dietitians and clinical researchers.

Note: This information is provided to you as an educational service of **LUNG LINE® (1-800-222-LUNG)**. It is not meant to be a substitute for consulting with your own physician.