# Cardiac Sarcoidosis

## What is Cardiac Sarcoidosis?

Sarcoidosis is a poorly understood inflammatory disease that commonly affects the lungs. It may 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 percent of people with sarcoidosis. Sarcoidosis is characterized by the presence of granulomas. These are ball-like collections of white blood cells. Granulomas may be associated with inflammation. The inflammation associated with granulomas can damage every part of the heart, including the electrical system, muscle, valves, arteries and outer lining of the heart called the pericardium. It is important to remember that CS may precede, follow, or occur at the same time as other (non-cardiac) forms of sarcoidosis.

## What causes Cardiac Sarcoidosis?

The exact cause of sarcoidosis is unknown. Granulomas occur when the immune system overreacts to an unknown foreign substance or substances. Infectious agents as well as environmental exposures may stimulate an overactive 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 electricity that flows 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 may be present. Ventricular tachycardia occurs in almost 25 percent 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 may cause the heart muscle to weaken and/or stiffen. These abnormalities may lead to fluid retention in the lungs, abdomen, and lower extremities. In extreme cases the wall of the heart may weaken and bulge outward like a thinning or balding tire. Granulomas may also infiltrate the hearts valves, causing leaky valves also resulting in heart failure.

**Coronary Disease:** Although rare, CS may 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, holter and/or event monitor 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 fails to identify granulomas (proof of cardiac sarcoidosis) in the majority of people.

## What is the Treatment?

Controversy exists as to the best treatment for CS. However, treatment is often directed at suppressing the inflamed granulomas within the heart. Corticosteroids (cortisone, prednisone, and methylprednisolone) are the first-line therapy to treat the inflammation. When people cannot take corticosteroids, other medications may be used. These include: methotrexate and azathioprine. 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 treatment with antiarrhythmic medications, which regulate the electricity within the heart and/or placement of an internal cardiac defibrillator (ICD).

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Notes

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