Sarcoidosis is a disease in which parts of the body develop granulomas, clumps of immune system cells responding to a perceived threat (Figure). These most commonly form in the lungs, in lymph nodes, and on the skin, but can appear in virtually any organ. Granulomas can interfere with normal organ function, causing symptoms that may be similar to those of other diseases. Cardiac sarcoidosis refers to the development of sarcoid granulomas in the heart muscle. This can affect the heart’s ability to pump blood to the rest of the body, with potentially life-threatening consequences.

What Are Signs and Symptoms of Cardiac Sarcoidosis?
Cardiac sarcoidosis can cause complete heart block, ventricular and atrial arrhythmias (heart rhythm abnormalities affecting the bottom and top parts of the heart), and congestive heart failure. Complete heart block may result in extremely slow heart rate, causing lightheadedness, loss of consciousness, or even death. Depending on where sarcoid granulomas are located within the heart, they can cause a variety of arrhythmias by disrupting the heart’s normal contraction. Arrhythmias may cause lightheadedness, palpitations (sensation of the heart beating fast or irregularly), fainting, or death. Congestive heart failure is the condition where the heart is unable to pump well enough to maintain the body’s full function. Its most common symptoms are fatigue, cough, shortness of breath, and swelling in the lower body (Table).

What Causes Sarcoidosis?
The cause of sarcoidosis is not well understood, but it probably results from an immune response after exposure to an as-yet unknown substance. Viruses, bacteria, and organic and inorganic particles may be involved in triggering the immune response. Why sarcoidosis affects the heart in some patients but not in others is unknown.

How Is Cardiac Sarcoidosis Diagnosed?
A diagnosis of cardiac sarcoidosis can be made in a few different ways. The first tests performed when cardiac sarcoidosis is suspected are an ECG and echocardiogram, which are helpful in assessing the heart’s electric system and its pumping function, respectively. These tests are readily available and do not use radiation. X-rays may be used to assess the lungs and lymph nodes, the organs where sarcoidosis is most commonly found. Definitive diagnosis of cardiac involvement is made by endomyocardial (heart) biopsy. Access to the circulatory system for the biopsy may be through the jugular vein in the neck or the femoral vein at the groin. A biopsy is inserted through a catheter and positioned inside the heart where a tiny sample of the muscle tissue is retrieved for analysis. If a granuloma is found, the presence of cardiac sarcoidosis is confirmed. Unfortunately, because the disease process is spotty, the biopsy is often negative even in patients who have cardiac sarcoidosis. Diagnosis is often achieved instead by cardiac imaging with MRI, computed tomography, or more recently a nuclear medicine technique called positron emission tomography. In patients with arrhythmias, use of a Holter monitor, a wearable device that records the heart’s rhythm over 1 to 2 days, allows assessment of the frequency and duration of arrhythmic episodes. An electrophysiology study may also be performed to both identify and treat arrhythmias at their source locations in the heart.
How Is Cardiac Sarcoidosis Treated?

Treatment of cardiac sarcoidosis includes fighting inflammation with immunosuppressant medication and managing functional consequences of the disease on the heart. Prednisone is commonly used along with other immunosuppressant drugs at doses high enough to be beneficial but low enough to minimize potential side effects. The treatment of heart weakening or heart failure often includes diuretics, angiotensin-converting enzyme inhibitors, and β-blockers. Antiarrhythmic drugs may also be used to treat arrhythmias. Pacemakers and implantable cardioverter defibrillators may be used to manage arrhythmias such as fast and slow heart rates, to abort cardiac arrest, and sometimes to treat heart failure. In rare advanced cases of the disease, heart transplant may be required.

What Does Cardiac Sarcoidosis Mean for Daily Life?

There are no general restrictions for patients with cardiac sarcoidosis, but individual circumstances may warrant lifestyle changes to better manage the disease. Because of the many different ways that sarcoidosis can affect the heart, doctors will make patient-specific recommendations for exercise and work, in addition to prescribing medications and other treatments. If prednisone or other immunosuppressant medications are used, patients may be vulnerable to infections and should seek medical attention if they develop symptoms. Some patients take prophylactic antibiotics to address this vulnerability. Patients with pacemakers and implantable cardioverter defibrillators should ask their physicians about any restrictions in activities. Avoidance of contact sports and strong electromagnetic fields, like those generated by arc welding, are commonly encouraged.

Conclusions

Cardiac sarcoidosis is a disease in which the immune system is activated, causing the formation of clumps of cells that disrupt the organization and function of the heart. This is a potentially dangerous condition which should be assessed for progression and severity and treated appropriately. Effective treatment can markedly improve outcomes and quality of life, though there is much room for further study to better understand the disease.

Disclosures

None.
Cardiac Sarcoidosis
William Froehlich, Frank M. Bogun and Thomas C. Crawford

Circulation. 2015;132:e137-e138
doi: 10.1161/CIRCULATIONAHA.114.013308

The online version of this article, along with updated information and services, is located on the
World Wide Web at:
http://circ.ahajournals.org/content/132/10/e137