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MEDFACTS

Cardiac Sarcoidosis

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Cardiac Sarcoidosis

What Is Cardiac Sarcoidosis?

Sarcoidosis is a poorly understood disease that most often affects the lungs. However, it also can involve almost any part of the body, including the heart. Sarcoidosis most often affects people between the age of 25 and 45. When sarcoidosis affects the heart, it is called cardiac sarcoidosis. Cardiac sarcoidosis is estimated to be present in 10 to 25 percent of people with sarcoidosis. Some estimates say that up to one in four patients with sarcoidosis of the lungs have cardiac sarcoidosis without any symptoms, meaning that the number of people with cardiac sarcoidosis may be much higher than estimated.

Sarcoidosis is an inflammatory disease, which 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 cause of sarcoidosis is unknown. Though it can be treated, there is no known cure. Severity of sarcoidosis varies from mild to severe and even life-threatening. When left unchecked, it can cause permanent scarring, or fibrosis, of organ tissue. This is also true of cardiac sarcoidosis.

Cardiac sarcoidosis is serious and requires treatment. Though cardiac sarcoidosis most often occurs in the heart muscle. 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 that 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. and may occur after there is no longer any inflammation from active sarcoidosis.

What Causes Cardiac Sarcoidosis?

The exact cause of sarcoidosis is unknown. We know that African American are three to four times more likely to develop sarcoidosis than Caucasians, and may have more severe disease. The granulomas of sarcoidosis appear to be the result of an immune system response to an unidentified trigger. Researchers are not certain as to whether that trigger could be an environmental exposure to a chemical or linked to an organism, such as a bacteria or virus, or if there is another unknown cause. Some chemicals in the environment, such as beryllium, aluminum and zirconium, can cause lung disease that is similar to sarcoidosis, but it is unproven as to whether these are a factor in sarcoidosis. The damage that is done to tissues and organs such as the heart is a result of the bodies immune system trying to destroy the “foreign” sarcoidosis.

Some studies also indicate that there is a genetic (inherited) factor in sarcoidosis. Genetic mutations in white blood cell proteins (called human leukocyte antigens, or HLA) as well as chemicals that control inflammation (called cytokines) have been linked to sarcoidosis, including cardiac sarcoidosis.

Some experts say that both genetic and environmental factors may play a role in cardiac sarcoidosis. A genetically susceptible person may be more likely to react to environmental triggers and develop the disease.
The percentage of people with sarcoidosis who have been diagnosed with cardiac sarcoidosis has increased dramatically in recent years. However, experts say that this is more likely an improvement in diagnostic techniques than an increase in disease prevalence.

Researchers are continuing to study this little-understood disease. Learning more about the cause of sarcoidosis will lead to improvements in diagnosis and treatment.

How Can Sarcoidosis Affect the Heart?

Cardiac sarcoidosis can have many symptoms and take many forms. Some are benign, and others are serious. Some of the most common symptoms include:

- Syncope (lightheadedness or fainting)
- Shortness of breath
- Coughing
- Chest tightness or chest pain
- Fast or irregular heartbeat

These and other symptoms are linked to the possible forms and complications of cardiac sarcoidosis, which include:

Heart Rhythm Disorders: A complete block of electrical movement through the heart (heart block) is one of 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. It 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 resulting in abnormal heart valve function.

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.

Pulmonary Hypertension

Pulmonary hypertension is high blood pressure in the lung arteries which can be a direct results of sarcoidosis affecting the lung arteries, or from lung problems such as sarcoidosis. Pulmonary hypertension may cause shortness of breath, decreased exercise capacity, chest pain and other symptoms.

How Is Cardiac Sarcoidosis Diagnosed?

Diagnosing cardiac sarcoidosis can be very challenging because the symptoms of cardiac sarcoidosis are similar to those of many other diseases. Since cardiac sarcoidosis is relatively rare, many times other diseases are suspected instead of cardiac sarcoidosis. There are no widely accepted guidelines for either screening or diagnosing sarcoidosis of the heart. Moreover, the currently 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. This is done because it is rare that cardiac sarcoidosis occurs without another form of sarcoidosis. About one in four people with another form of sarcoidosis will have cardiac sarcoidosis as well; some say that number is even higher.

Initial cardiac evaluation may include:

- Electrocardiogram (EKG). This test checks for an abnormal heart rhythm or abnormalities in the electrical system of the heart.
- Signal-averaged EKG. This is a special EKG that averages several hundred heartbeats to detect subtle abnormalities in the movement of electrical signals through the heart.
- Echocardiogram (ultrasound of the heart). The ultrasound shows provides assessment of the pumping and relaxing function of the heart, valve function and estimate of pressures in the heart. It does this by using sound waves.
- Holter monitor (extended EKG). A Holter monitor is a monitor that continually records the electrical signals of your heart for 24 hours or more.

Additional imaging tests may include:

- Single photon emission computed tomography (SPECT). This test can show how blood flows to tissues and organs in the body.
- Positron emission tomography (PET)/CT Scan. A PET scan can identify active inflammation in the heart muscle suggesting active cardiac sarcoidosis. A CT scan or CAT scan is a shortened name of computerized tomography. A CT scan takes pictures of the inside of the body. The pictures are more detailed than a typical x-ray.
- Cardiac MRI. A cardiac MRI is the best test to see evidence of any scar tissue in the heart which may lead to the cardiac complications from cardiac sarcoidosis. It also provides assessment of the size and pumping function of the heart.

A positive heart biopsy confirms cardiac sarcoidosis. However, it may more often be negative or normal even when there is sarcoidosis in the heart, especially if heart function is normal. For this reason, taking a biopsy is a less common test for detecting cardiac sarcoidosis.

What Is the Treatment?

There is no cure for cardiac sarcoidosis, but it can be treated. However, controversy exists as to the best treatment for cardiac sarcoidosis. 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. Corticosteroids are very good at reducing inflammation (swelling). The steroids (corticosteroids) used to treat cardiac sarcoidosis are not the same as anabolic steroids, used illegally by some athletes for bodybuilding. Corticosteroids do not affect the liver or cause sterility.

When people cannot take steroids, or when combination therapy is needed, other medications to suppress the immune system are used. These include: methotrexate, azathioprine, mycophenolate, antimalarials and others.

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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