### Treatment

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#### Overview

Timely diagnosis and treatment with medication is important for a favourable prognosis.

Medications can reduce symptoms and improve the pressures in the blood vessels.

#### General medications for Sarcoidosis

Your doctor may prescribe medication to suppress the sarcoidosis:

- **Corticosteroids in tablet form**: usually prednisolone. These medications must be used for a number of years, or even permanently.
- **Other medicines that suppress the immune system**, for example: methotrexate, mycophenolate or azathioprine.

These drugs suppress the inflammation but do not medicate the heart itself.

#### Heart Sarcoidosis Treatment

You may be prescribed the following to help regulate heart rhythm:

- **Specific heart medications (tablets)**;
- **Implant a pacemaker or defibrillator (ICD)**.

You may be prescribed:

- **Diuretics**;
- **Endothelin antagonists**;
- **Prostacyclins**.

#### Transplant

Heart transplant may be considered. This is very rare.

Lung transplant may be considered. This is very rare.

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### Who We Are

SarcoidosisUK is the UK Sarcoidosis Charity. We’re here to provide support, information and to fund research. The vast majority of our funds go towards researching a cure. We run support groups across the UK – please contact us for information on your nearest group.

### How You Can Help:

**Donate** to fund research: www.sarcoidosisuk.org/donate

**Join** our registry: www.sarcoidosisuk.org/patient-register

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- Dr. L. Williams & Dr. S. Agarwal, Consultant Cardiologists, Dr. K. Tweed, Consultant Radiologist and Dr. M. Thillai, Consultant Chest Physician (all Cardiac Sarcoidosis Unit, Royal Papworth Hospital);
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Sarcoidosis and the Heart

The heart can be affected by sarcoidosis in two ways. Firstly, sarcoidosis can occur in the heart muscle itself (cardiac sarcoidosis). Secondly, the heart may be indirectly affected as a result of sarcoidosis in the lung (pulmonary hypertension). In both cases serious consequences can occur.

Cardiac Sarcoidosis

Cardiac sarcoidosis occurs when the heart muscle itself is affected. The accumulation of immune cells causes clumps of tissue (granulomas) which can occur in the heart in different places:

- The heart muscle of the left or right ventricles and interventricular septum;
- The papillary muscles (connected to the heart valves);
- The pericardium (thin sac lining the heart);
- And (less often) the left or right atrium.

Granulomas can also occur in the electrical conduction system of the heart (which normally helps regulate your heart beat). Cardiac sarcoidosis occurs in up to a third of all sarcoidosis patients, but only causes specific symptoms in around 5% of cases.

Symptoms:

Symptoms of cardiac sarcoidosis include:

- Irregular heartbeat (pounding or fluttering sensation, or a ‘skipping of beats’);
- Dizziness and/or fainting spells;
- Shortness of breath;
- Chest pain;
- Swelling of the legs (in later stages).

Pulmonary Hypertension

Pulmonary sarcoidosis (sarcoidosis of the lung) can also indirectly affect heart function. Abnormalities of the lung, and of the blood vessels between the lung and the heart, can cause an increase in the pressure within the blood vessels in the lungs. This pressure can then overload the right ventricle. This can occur in up to 15% of all patients with sarcoidosis.

Techniques to Understand your Condition

Most newly diagnosed sarcoidosis patients will have an ECG, holter monitor and echocardiogram. Suspected cardiac sarcoidosis patients may go on to have further diagnostic tests. Most of these are described below.

ECG (Electrocardiogram):

This provides information about the electrical conduction system of the heart, as well as the heart rate and heart rhythm.

Holter Monitor:

Holter monitors also record the heart rhythm. It is a removable band that can be worn for a longer period of time (usually 24 hours but sometimes longer). It is particularly useful to identify any evidence of problems in the conducting system of the heart (i.e. very fast or slow heart beats or any abnormal rhythms).

Echocardiogram:

This uses ultrasound waves to create images which show the pumping function of the muscular heart chambers and the function of the heart valves. It also allows measurement of the blood pressure within the heart vessels (to look for pulmonary hypertension).

MRI (Magnetic Resonance Imaging):

This scan can identify involvement of sarcoidosis in the heart by injecting a special contrast agent/dye to evaluate the soft tissue in the heart muscle. It also provides information about heart function.

Nuclear Scans:

Thallium scans and PET scans create images that can identify any active inflammation within the heart muscle and can help to guide treatments. For the cardiac PET scan it is important that a strict diet is kept to in the days before the scan (this will be explained to you beforehand).

Clinical Electrophysiology Studies:

A catheter is used to map the conduction system of the heart. This can uncover abnormalities which may predispose abnormally slow heart rates, as well as any abnormally fast or dangerous heart rhythms which can arise from areas of inflammation or scarring.

Right Heart Catheterisation:

This test uses a probe to measure the pressures in the heart and in the nearby blood vessels to determine pulmonary hypertension involvement.

Biopsy of the Heart

This is rare and only used when there is a specific reason to do so. Patients who have had a lung biopsy will usually not need any further biopsies.

Blood Tests

The following may be tested from blood samples:

- Angiotensin converting enzyme (ACE)
- Brain natriuretic peptide (BNP)
- Troponin