

SARCOIDOSIS of the skin

What are the aims of this leaflet?

This leaflet has been written to help you understand more about sarcoidosis of the skin. It will tell you what it is, what causes it, how it is diagnosed, how it can be treated and where to find out more about it.

What is sarcoidosis?

Sarcoidosis is a relatively uncommon disease that can affect just one part of the body, or different parts at the same time. Most commonly it affects the skin, lungs, lymph nodes, or eyes. Less frequently it can affect the joints, the nervous system, the liver and the kidneys. Sarcoid causes a type of inflammation known as a 'granuloma', which consists of a cluster of cells that can stop an organ from working properly. For example, granulomas in the lungs can cause breathing problems.

What causes sarcoidosis?

The cause of sarcoidosis is not fully understood. It is thought to be due to changes in the immune system. These changes may be partly inherited, but exposure to certain infections or environmental factors may also play a role.

Sarcoid itself is not infectious or malignant. It cannot be transmitted from person to person. It can affect people of all ages and races but is seen more commonly in women than men and in afro-caribbeans more than caucasians. The disease most commonly begins between the ages of 20 and 40.

What are the symptoms of sarcoidosis?

Sarcoidosis can present with several different symptoms, or in its early stages it may have none at all. Symptoms can be quite non-specific, like feeling tired, losing weight or aches and pains in the joints. Involvement of specific organs cause symptoms at that site. If the sarcoid is affecting the lungs this can cause a dry cough or shortness of breath; if it affects the eyes they can become red, sore and dry and the eyesight may become blurred; joints may become stiff and painful; lymph nodes ('glands') may become swollen.

When sarcoid affects the skin, it can have several different appearances.

What does sarcoid of the skin look like?

- Papules and plaques. This is the most common way for sarcoid to affect the skin. The papules are small raised bumps that are often purplish red or brown in colour. They can occur anywhere on the body but are commonly seen on the face. Plaques are a similar colour but are larger and tend to occur anywhere on the body.
- Erythema nodosum. These are fleshy tender bumps that occur on the front of the shins. There are many different diseases that can cause erythema nodosum and

sarcoid is just one of them. As the painful red bumps fade a bruise like area is left behind. Some people experience fever, sore joints and feel unwell when the lesions first appear. 1 in 4 people who have sarcoidosis will have this skin condition.

- Lupus pernio. Dusky purplish swollen areas that appear on the nose, cheeks, ears or fingers. This type of skin presentation is associated with more aggressive disease that may affect other parts of the body. It can be harder to treat and can cause scarring.

There are rarer types of sarcoid in the skin that includes scar sarcoidosis, where an existing scar or tattoo becomes red, lumpy and firmer to touch. Sarcoid can also affect the scalp, nails or mouth and occasionally causes areas of ulceration, or breakdown, in the skin.

How is sarcoidosis in the skin diagnosed?

In order to diagnose sarcoid in the skin a small sample (biopsy) is taken from the affected skin and then looked at under the microscope.

Other tests may then be performed to see if the sarcoid is affecting other parts of the body.

A blood test may be taken to check the level of calcium and angiotensin-converting-enzyme (ACE) in the blood. Both of these can be elevated in sarcoidosis.

As sarcoidosis commonly affects the lungs or the lymph nodes in the chest a chest x-ray will be performed. Pulmonary function tests may also be requested. Even though you may not have a cough or shortness of breath, it is important to check whether the lungs are involved.

The other tests performed will depend on the symptoms you are experiencing but often include a detailed eye examination.

Can sarcoidosis be cured?

At present there is no known cure for sarcoidosis. The aim of treatment is to suppress the disease activity. However in 6 out of 10 people affected by sarcoidosis the disease will completely resolve within 2 to 5 years.

How can sarcoidosis be treated?

Not all patients with sarcoidosis require treatment. The disease can remain stable, or 'burn out' by itself without requiring any treatment. If the sarcoidosis is causing symptoms or distress and therefore affecting your quality of life, it is recommended that it is treated. Even if treatment is not started you will be reviewed by your specialist doctor on a regular basis, often every six months.

For sarcoid that is only affecting the skin, the most common form of treatment is the application of potent steroid creams or ointments to the affected lesions. Sometimes a small injection of steroid is given directly into the skin lesion to help it resolve.

When the skin disease is not responding to topical treatments or the sarcoid is affecting other parts of the body and causing symptoms, oral steroid tablets are used to control the disease. These can work very well, and are normally given over several months, but there are side effects associated with long-term treatment with steroids. These include thinning of the bones, weight gain, diabetes, cataracts and high blood pressure.

Therefore another tablet treatment may be added in or used instead. These include:

- Hydroxychloroquine (see Hydroxychloroquine PIL)
- Methotrexate (see Methotrexate PIL)
- Mycophenelate Mofetil (MMF) (see MMF PIL)
- Thalidomide
- Tetracyclines e.g. Lymecycline, Doxycycline

There are also newer injectable treatments e.g. Infliximab and Adalimumab, that may be used in severe progressive disease, or disease that has failed to respond to other treatments. These target a very specific part of the immune system to try to stop the formation of granulomas.

Where can I get more information about sarcoidosis?

The Sarcoidosis Charity

020 7237 5912

www.sila.org.uk/

Sarcoidosis Association

www.sa-uk.org

www.uptodate.com/contents/sarcoidosis-beyond-the-basics