LUNG FOUNDATION AUSTRALIA

"When you can't breathe... nothing else matters"®

Sarcoidosis

What is sarcoidosis?

Sarcoidosis is the name given to a condition in which small nodules or "sarcoid granulomas" appear in the body's tissues. The cause of sarcoidosis is currently unknown.

Sarcoid granulomas are seen on tissue biopsy under the microscope and consist of groups of immune cells which are normally part of the body's defence system. Sarcoidosis is a form of inflammation that is not cancerous and it usually gets better by itself over time. In chronic or progressive disease, treatment with prednisolone or cortisone may be required.

Sarcoidosis can involve any organ in the body, but in 90% of cases, it involves the lungs or lymph nodes next to the lungs. Lymph nodes elsewhere in the body such as the neck or the armpits may be involved and in some individuals, sarcoidosis can involve other organs or tissues within the body including the salivary glands, liver, eyes, skin and heart.

What are the symptoms?

Many people with sarcoidosis feel perfectly well and are surprised when it shows itself on a chest x-ray and a diagnosis is made. Other people have a cough or fever with chest pain or shortness of breath while others feel simply, "flat" or unwell. Active sarcoidosis can cause fever, facial swelling, arthritis (particularly of the large joints) and painful red lumps on the front of the legs (erythema nodosum). Sarcoidosis can also affect the brain (headaches, weakness, numbness, or blurred vision), the heart (palpitations, dizziness or chest pain), the skin (with brown or red lumps, or rash) and it can cause inflammation of one or more joints of the body or the eyes. Very occasionally, it can lead to high levels of calcium in the blood, causing thirst and kidney damage, or simply increase the calcium in the urine and thereby pre-dispose the patient to kidney stones.

What causes sarcoidosis?

In spite of extensive worldwide research, no convincing cause has yet been identified. Our current thinking is that it is either due to an infective agent or an allergy combined with susceptible genes causing this unusual type of inflammation to develop. Some genes appear to increase the risk or sarcoidosis whilst others may be protective. Continued research hopes to identify these factors more clearly.

Occasionally, sarcoidosis runs in families. It appears to be more common in cooler climates and affects around 20-40 people per 100,000 of the population. It is most common in the 20-40 age groups but can occur in much younger or older individuals. We do know, however, that it is not a cancer or any other sort of malignant tumour. The incidence of the disease does differ in some races, being more common and severe in black people such as African-Americans than in Caucasians. It is unusual in those of Asian descent or in Aboriginal Australians.

How is sarcoidosis diagnosed?

Sarcoidosis can sometimes be diagnosed solely on the basis of the symptoms, x-rays and blood tests but often, a biopsy of an involved organ tissue is required. If a biopsy is needed, your doctor will advise what the simplest and fastest form of biopsy is in your particular case. The biopsy specimen is taken and presented to a pathologist who examines it under the microscope. That procedure may take several days. Lung biopsies are often performed via a fibre-optic bronchoscope or via a small operation on either side of the neck or through the ribs on one side.

Sarcoidosis can sometimes be difficult to diagnose. If the diagnosis is suspected when a chest x-ray is done for other reasons such as a chest infection, it is important to differentiate it from other conditions which can cause lymph gland enlargement and similar shadows on chest x-rays. Your doctor will usually arrange the relevant blood tests, urine tests, breathing test, electrocardiogram (ECG), echocardiogram (ECHO), chest CT scan, neurological and eye examinations. Other special tests may be required and your doctor will advise you as to which test you will need. A specialist opinion is usually indicated, depending on which organ system is involved.

What is the treatment?

In Australia, most cases of sarcoidosis usually get better over 1-3 years and may not require any specific treatment at all. Once sarcoidosis appears to be improving, it is rare for it to suddenly get worse, except when therapy is abruptly removed. Very few people require longer term treatment.

Occasionally treatment is required because of serious or progressive disease, or involvement of the eyes, heart or kidneys. Prednisolone or cortisone is the usual therapy and is often required for 6 to 12 months, sometimes longer. Initially the prednisolone dose may need to be high, but the dose can often be reduced to a much smaller level once control of the process is achieved. Sudden stoppage of prednisolone should not occur as it can be harmful and lead to rapid worsening of symptoms. Rarely, other forms of drug therapy may be needed as well, such as for sarcoid changes in the skin. Pregnancy is not affected by sarcoidosis and females with sarcoid who become pregnant usually get a little better. Very occasionally, the disease can get a little worse after the baby is born.

Be careful of internet information

Sarcoidosis can be a very severe disease in people of African descent. Reports on the internet often focus on these severe cases and give a distorted view of the condition. If you are worried about what you have read or heard, you need to discuss these concerns with your doctor.

Do I need to alter my lifestyle?

If you are a smoker, stopping smoking is advisable. Most people with sarcoidosis lead a normal active life. However, occasionally, too much sunshine can be harmful and lead to high calcium levels in the blood and urine. This can lead to kidney problems but your local doctor can check this situation for you if you are concerned.

Where to from here?

This information should reassure you that the vast majority of people with sarcoidosis recover and the condition rarely returns. Many patients do not require any active treatment other than observation. As the condition can be so varied in its distribution throughout the body, your particular case will need to be carefully assessed and monitored by your local doctor and/or specialist until you have recovered. Please feel free to ask them any further questions you may have.

Please consult your family doctor or specialist respiratory physician if you have further questions relating to the information contained in this leaflet.

TD0716V3SARC