DermNet NZ

Authoritative facts about the skin from the New Zealand Dermatological Society Incorporated.

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Sarcoidosis

What is sarcoidosis?

Sarcoidosis is a multisystem granulomatous disease. This means there are scattered collections of mixed inflammatory cells (granulomas) affecting many different parts of the body. Characteristically these are non-caseating epithelioid granulomas (a pathological description distinguishing sarcoidal granulomas from the caseating or cheese-like granulomas seen in tuberculosis).

Sarcoidosis usually starts in the lungs or lymph nodes in the chest. It is thought that inflammation of the alveoli (tiny sac like air spaces in lungs where carbon dioxide and oxygen are exchanged) is the start of the disease process in the lungs. This may either clear up on its own or lead to granuloma formation and fibrosis (scarring). Over 90% patients have some type of lung problem. Once considered a rare disease, sarcoidosis is now the most common of the fibrotic lung disorders.

Other commonly affected organs (i.e. outside the lungs) include the following:

- Skin 20-35%
- Eye 20-30%
- Liver 30-40%
- Heart 5-25%
- Nervous system 1-5%
- Musculoskeletal 2-38%

Who is at risk of sarcoidosis?

Sarcoidosis occurs worldwide, affecting persons of all races, age, and gender.

- Risk appears to be greater if you are of African-American, Scandinavian, German, Irish or Puerto Rican descent.
- Mainly affects people between 20-40 years of age.
- Risk is two times greater in black women than in black men

What causes sarcoidosis?

Not much is known about sarcoidosis and to date there is no known cause or causes. It is thought to be a disorder of the immune system where there is a malfunction in the body's natural defence mechanisms. Whether this abnormal immune response is a cause of sarcoidosis or an effect of the disease still needs to be answered.

Continued research is necessary to answer remaining questions such as:

- Is sarcoidosis triggered by a hypersensitivity response to one or many agents (bacteria, virus, fungi, chemical toxin)?
- In which body organ does sarcoidosis actually start?
- Do hereditary, environment, and lifestyle play a role in how the disease presents and progresses?
- How can sarcoidosis be prevented?

What are the signs and symptoms of sarcoidosis?

Sarcoidosis may not result in symptoms and the disease may come and go without the patient or doctor ever being aware of it. Symptoms can appear suddenly and then just as quickly resolve spontaneously. Sometimes, however they can continue over a lifetime.

Symptoms can be related to the specific organ affected, or they can be non-specific general symptoms, including:

- weight loss
- loss of apetite
- fatigue
- fever
- chills and night sweats.

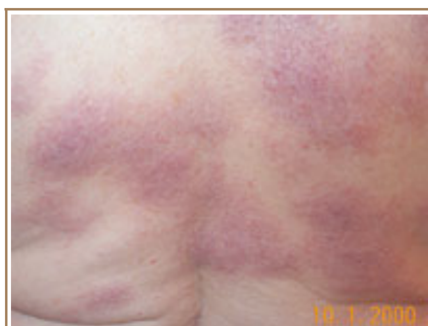
Sarcoidosis may involve one organ system or several.

<table>
<tr><th colspan="2">Symptoms and signs of specific organ involvement</th></tr>
<tr><th>Organ</th><th>Comments</th></tr>
<tr><td>Lungs</td><td>Respiratory symptoms include coughing (dry or with phlegm) and shortness of breath
Occasionally chest pain and a feeling of tightness in the chest
In some patients upper respiratory tract involvement produces hoarseness, nasal obstruction and recurrent or persistent sinusitis</td></tr>
<tr><td>Skin (cutaneous sarcoidosis)</td><td>About 1/3 of patients with cutaneous sarcoidosis have no involvement of other organs
Lesions may be non-specific (similar to several other skin conditions) or specific
Non-specific lesions include:
erythema nodosum: tender, 1-2cm bumps mostly on the shins; often accompanied by arthritis in the ankles, elbows, wrists and hands
nummular eczema
erythema multiforme
calcinosis cutis (deposition of calcium salts within the skin)
pruritus (itch)
Specific lesions show granulomas on histology (microscopic examination of a skin biopsy) and include:
lupus pernio: large bluish-red and dusky purple infiltrated nodules and plaque-like lesions on nose, cheeks, ears, fingers and toes
skin plaques: purple-red or brown, thickened, circular skin lesions
maculopapular eruptions
nodular lesions deeper in the skin
infiltration (thickening) of old scars (scar sarcoidosis)</td></tr>
<tr><td>Eye</td><td>Red or watery eyes
Any part of the eye can be involved:
granulomatous uveitis: most common, results in blurred vision, watery eyes and photophobia (dislike of light)
iris nodules
retinochoroiditis
conjunctivitis
lacrimal gland involvement</td></tr>
</table>

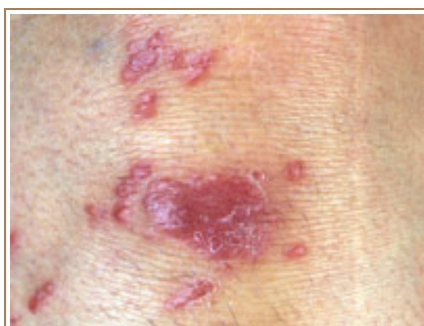
	◦ optic nerves ◦ proptosis (protruding eyeball) • Uncommonly, cataracts, glaucoma, and blindness can result
Liver	• Up to 1/3 have hepatomegaly (enlarged liver) or changes in their liver enzyme levels • Liver disease resulting in serious symptoms is rare • May result in fever, malaise and fatigue
Heart	• Sarcoidosis of heart muscle (myocardium) is much more common in the Japanese than in other races and is the leading cause of death from sarcoidosis in Japan • Chest pain, palpitations and rarely sudden death • Symptoms and signs associated with congestive heart failure, pericarditis or papillary muscle dysfunction (valvular heart disease). These include shortness of breath, ankle swelling, irregular heart beat and chest pain.
Nervous system	• Granulomas can appear in the brain, spinal cord, and facial and optic nerves • May result in headache, confusion and malaise • Facial paralysis
Musculoskeletal	• Arthritis (inflammation of the joints), periarthritis (inflammation of surrounding tissues) or arthralgia (painful joints) may occur • Arthritis is most commonly acute (coming on suddenly), resulting in swelling of the lower legs and tenderness of ankles, knees and fingers • Chronic (long term) sarcoid arthritis is rare
Calcium metabolism	• Raised serum calcium levels in 2-63% of sarcoidosis patients due to overproduction of vitamin D by sarcoid granulomas. It may not require treatment. • Kidney stones (nephrolithiasis) may result from the abnormal calcium metabolism

Cutaneous sarcoidosis

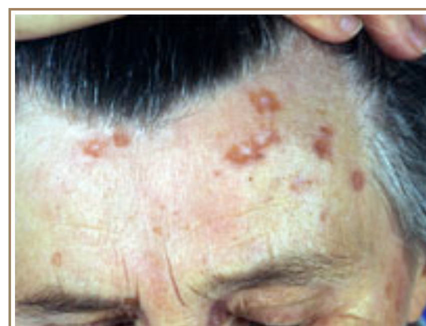
Sarcoidosis



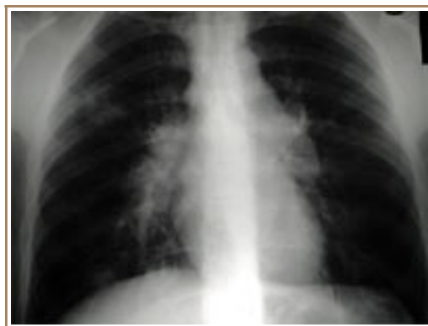
Sarcoid plaques

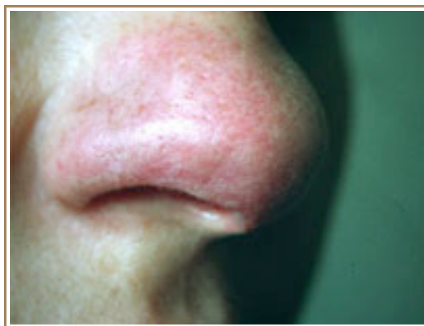


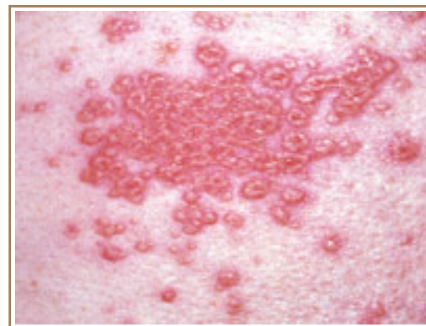
Sarcoid on knee arising in scars



Sarcoid granulomas on forehead


Enlarged lymph nodes on chest X-ray


Lupus pernio


Sarcoid papules

How is sarcoidosis diagnosed?

There is no single or specific diagnostic test for sarcoidosis. The following tests may be performed when sarcoidosis is suspected.

Test	Comments
Chest x-ray	• Changes in appearance of lungs, heart and lymph nodes can be first indication of sarcoidosis
Lung function	• Tests to see how well the lungs are doing their job of exchanging CO2 and O2 with the blood • Granulomas and fibrosis of lung tissue reduces lung capacity and disturbs the normal flow of gases
Blood	• Mild anaemia occurs due to granulomatous bone marrow involvement, or chronic disease state • Increase in serum calcium levels and abnormal liver function tests often accompany sarcoidosis
Biopsy	• Microscopic examination of specimens of lung tissue or other tissue of organs involved can show where granulomas have formed in the body and confirm diagnosis. Multiple biopsies may be necessary.
Slit-lamp eye examination	• Examines the inside of the eye • Used to detect silent damage such as asymptomatic uveitis
Other tests	• CT scanning, echocardiography, gallium scanning • ECG • Kveim test: an injection of sarcoidal spleen, biopsied to determine whether granulomas have formed (this test is no longer available in New Zealand) • Liver and kidney function tests • 24-hour urinary calcium excretion • Angiotensin converting enzyme

These tests will not only be used to help confirm diagnosis but can also help the doctor to monitor the progress of the disease over time and determine whether the condition is improving or deteriorating.

What is the treatment for sarcoidosis?

For most patients with sarcoidosis no treatment is required. Symptoms are usually not disabling and tend to disappear spontaneously. In mild-to-moderate cases, because the disease can resolve on its own spontaneously, a 3 months observation period is recommended before commencing any treatment. When treatment is necessary the aim is to keep the lungs and other affected organs functioning and to relieve symptoms.

In most patients initial treatment is with corticosteroids; these are used to treat inflammation and granuloma formation. Prednisone is the most often prescribed corticosteroid. This may need to be continued for several years as the disease can often relapse once treatment stops.

If prednisone fails to improve symptoms, other immune-modifying agents such as methotrexate, azathioprine, hydroxychloroquine or tetracyclines may be used.

Eye and skin lesions may be managed with topical corticosteroids (eye drops, creams or injections). Laser surgery has been used in treating disfiguring skin plaques and lupus pernio.

Success in individual cases has been reported with allopurinol, isotretinoin, leflunomide, pentoxifylline, infliximab and thalidomide.

It is often difficult to know when to start treatment, what drugs and dose to prescribe, and how long to treat for, as the disease can often resolve without any tratment. The decision depends on the organ systems involved and how far the inflammation or granuloma formation has progressed.

Monitoring the disease is essential as treatment can then be modified accordingly.

How serious is sarcoidosis and what is the prognosis?

In general, sarcoidosis appears briefly and resolves without relapse in most cases. 20-30% of patients are left with some permanent lung damage and 10-15% develop chronic sarcoidosis that may last for many years.

In 5-10% of cases the disease can become fatal if either granulomas or fibrosis seriously affects vital organs such as the lungs, heart, nervous system, liver or kidneys. End-stage lung disease may need lung transplantation.

Cutaneous sarcoidosis usually has a prolonged course. Papules and nodules tend to resolve over months or years, whilst plaques may be more resistant. Lupus pernio is often present in patients with chronic fibrotic sarcoidosis and is associated with involvement of the upper respiratory tract, advanced lung fibrosis, bone cysts and eye disease.

With correct diagnosis and proper management, most patients with sarcoidosis continue to lead a normal life.

Related information

On DermNet NZ:

- Erythema nodosum
- Erythema multiforme
- Nummular eczema

Other websites:

- Sarcoidosis - emedicine dermatology, the online textbook
- Sarcoidosis: U.S. Department of Health and Human Services, Public Health Service, National Institutes of Health, National Heart, Lung, and Blood Institute
- Sarcoidosis: A Primary Care Review — American Academy of Family Physicians
- The Foundation for Sarcoidosis Research

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a dermatologist for advice.

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