

Hansen's disease

Also known as Leprosy

What is leprosy?

Leprosy is a chronic bacterial infection caused by the organism *Mycobacterium leprae*. It primarily affects the skin and the nerves. It may also affect mucous membranes (such as the nose) as well as eyes and testicles. Long standing leprosy may cause limb deformities as well. The manifestation the disease takes largely depends on the person's immune response to the bacterium.

Leprosy is one of the oldest diseases known to mankind. It was often referred to in the Bible and was recognised in the ancient civilizations of China, Egypt and India. Throughout history, infected people have often been ostracised by their communities. Consequently they often hid their condition and avoided seeking treatment to avoid discrimination. In fact, the major form of treatment before effective drugs became available was forcible isolation and incarceration.

Leprosy is a worldwide condition. The majority of cases occur in Asia, South America and Africa but leprosy also occurs in the Pacific Islands, including Papua New Guinea and Australia.

What causes leprosy?

It is an air-borne infection spread from person to person. Although large numbers of people are exposed to the bacteria in endemic areas, only a small number go on to develop the disease. Usually long-term close contact is necessary for this to occur.

The disease has a very long incubation period from 1 year to 20 years or even longer. It is not highly infectious and genetic factors are thought to be the main risk factor. About 95% of the population may be naturally immune.

What does leprosy look like?

In a large majority of affected people the first sign of the disease is feeling a numbness which may precede any skin lesions by a number of years. Temperature is the first sensation lost, followed by light touch, pain and then deep pressure.

The first skin lesion is usually of indeterminate type, which causes one or more de-pigmented (pale) spots before spontaneous resolution or evolving into the borderline, tuberculoid or lepromatous types of leprosy.

Main forms of leprosy

Tuberculoid leprosy

- Classically appears as a single or several well-defined, pale or coppery-red, asymmetrical spots with well-defined borders

- Lesions become dry and hairless
- Loss of sensation in the patches is characteristic in tuberculoid leprosy. Tender thickened nerves with subsequent loss of function including sensory loss and weakness of muscles
- Spontaneous resolution may occur in a few years or it may progress to borderline or lepromatous forms of leprosy

Borderline leprosy

- Numerous red, irregular-shaped large plaques with raised edges
- Sensory loss
- Thickened nerves
- Disease may remain at this stage, improve or worsen

Lepromatous leprosy

- Numerous raised plaques, papules and nodules. Symmetrically enlarged nerve trunks and sensory impairment
- Symptoms include nasal stuffiness or bloody discharge from the nose
- If left untreated the following may occur:
 - Skin thickens over the forehead (“leonine facies”)
 - Eyebrows or eyelashes are lost, the nose becomes misshapen, the ear lobes thicken
 - Eye involvement including blindness
 - Skin on the legs thickens and may form ulcers
 - Testicles shrivel and breasts enlarge
 - Voice become hoarse due to involvement of the larynx
 - Slow scarring of the peripheral nerves resulting in nerve thickening and sensory loss
 - Fingers and toes may become deformed due to painless, repeated trauma
 - Swelling of legs or ankles

How is leprosy diagnosed?

Leprosy has very characteristic clinical features but the diagnosis must be confirmed because of the need for prolonged treatment. Tests include a skin biopsy, showing characteristic granulomata and involvement of the nerves. Special staining of the tissue may show acid fast bacilli, the number visible depending on the type of leprosy. In lepromatous leprosy, numerous

Mycobacterium leprae bacilli may be found on smears taken from skin slits made in the ear lobes. However, the smears may be negative in the tuberculoid or indeterminate forms of the disease.

How is leprosy treated?

Management of leprosy is aimed at treating the infection and minimising potential physical deformities. The choice of antibiotics depends on the type of leprosy being treated. The forms of leprosy with fewer bacilli (paucibacillary leprosy) often respond well to rifampicin and dapsone – for 6 months. The more severe lepromatous form of leprosy (multibacillary leprosy) requires the addition of clofazimine and treatment for 12 months or longer. If a certain drug cannot be given due to allergy or other side effects, ofloxacin or clarithromycin are considered as substitutes. Leprosy is not usually treated with a single drug alone, due to the risk of development of drug resistance.

Oral steroids and/or thalidomide are helpful in preventing nerve damage.

Surgery may be necessary to reconstruct a collapsed nose, decompression of a nerve abscess, or in managing limb deformities.

Patient education is paramount. Leprosy can be cured but it is essential to take the full course of medication. Affected people become non-infectious soon after commencement of treatment. As such, affected people should not be isolated. Physical, social and psychological rehabilitation is necessary for those whose disease has been neglected.

Reactions to leprosy treatment

These are known as lepra reactions and are due to changes in the immune system. There are two types of lepra reactions:

- Lepa type I reaction causes inflammation of pre-existing lesions and tender enlarged peripheral nerves. This is treated with oral corticosteroids and anti-inflammatories.
- Lepa type II reaction, also known as erythema nodosum leprosum, is an immune complex reaction. The red nodules can blister or ulcerate. It can be accompanied by fever, joint pains, nerve pains, eye disease and possible involvement of other organs. Treatment may include clofazimine, thalidomide, corticosteroids and colchicine.

It is important to understand that leprosy is not a rare disease with at least 200,000 cases worldwide annually, including Australia. With appropriate treatment, leprosy is eminently curable.