Tidsskriftet Tidsskrift for Den norske legeforening

Leprosy is not just an infectious disease

PERSPECTIVES

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Published material on leprosy is often centred around the historical context of Armauer Hansen and the discovery of the leprosy bacillus, or on leprosy as an infectious disease, which remains a major health issue in 120 countries. However, leprosy is also a 'surgical disease' that can lead to severe health problems.

Foto: Bergen Museum / NTB

Leprosy is a chronic infectious disease that responds well to modern antibiotics (1, 2). However, despite such treatment, many patients still develop deformities in the face and extremities (3) due to the immune response to the leprosy bacteria. These leprosy reactions are one of the main causes of neuropathy and, in many cases, are triggered by the treatment itself. Considerable challenges therefore remain in treating the millions of patients worldwide affected by leprosy (4). The paradox is that successfully treating the *infectious disease* may transform it into a '*surgical disease*', causing significant disability for the patient. The frequency and severity of leprosy reactions vary, but in some areas, more than 50 % of patients are affected (4–6).

Increasing prevalence in Norway?

In Norway, a number of people with leprosy arrived as part of the large influx of boat refugees from Vietnam a generation ago. In the last few decades, there have only been a few dozen leprosy patients in Norway, but this could increase due to immigration from the Middle East, Asia, South America and North Africa, where millions are affected. It is therefore important for Norway to maintain its expertise in this disease.

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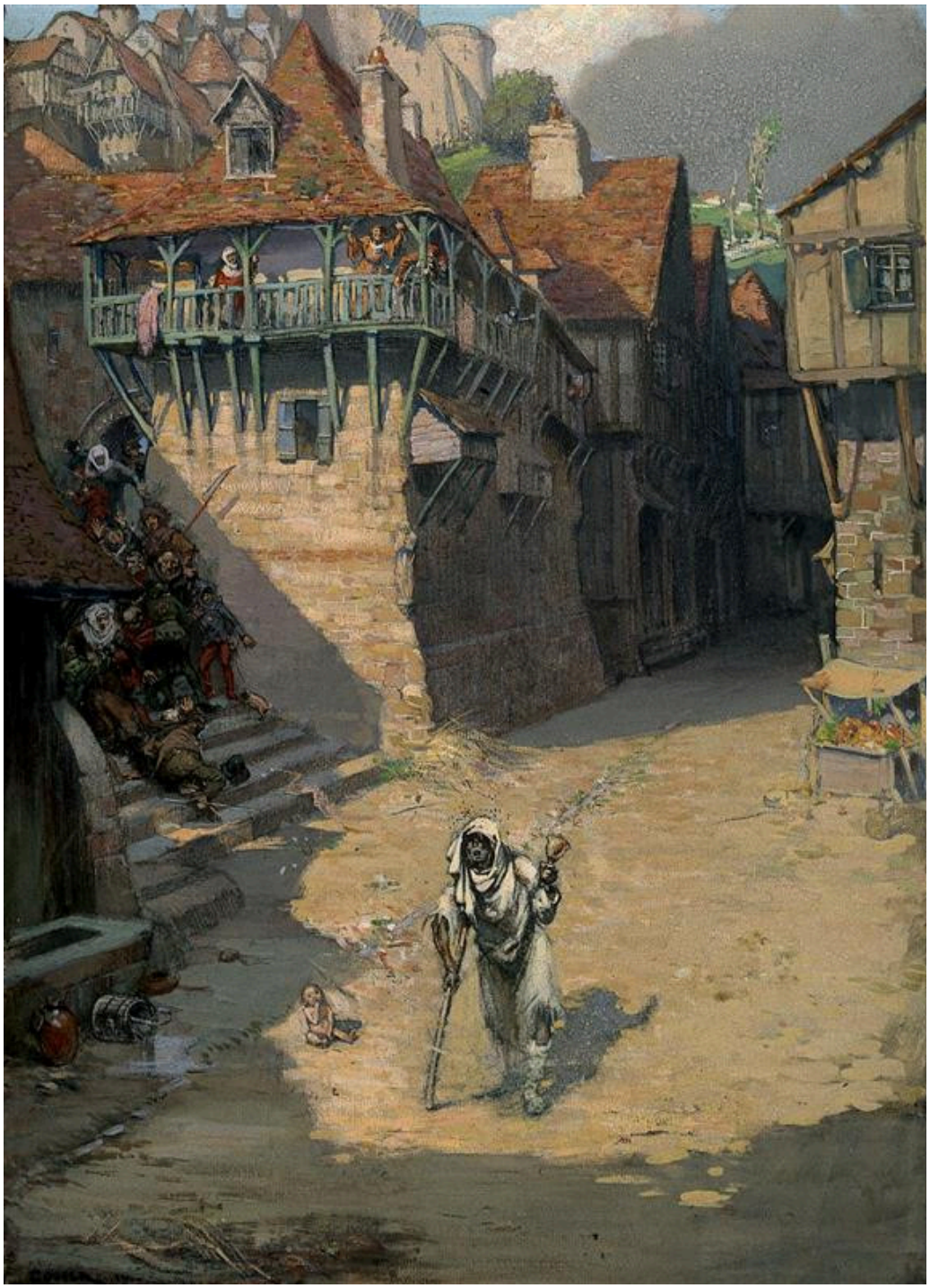
Mycobacterium leprae is an obligate intracellular bacterium with a particular affinity for mast cells in the skin and mucous membranes, and especially Schwann cells in peripheral nerves. The latter cells form myelin sheaths that

surround nerve fibres in the peripheral nervous system. Furthermore, the bacteria tend to multiply at low temperatures, which is why peripheral nerves located relatively close to the skin (and thus cooler) are usually affected. As a result, the damage tends to follow specific patterns. Permanent nerve damage can be partial or complete (6).

The World Health Organisation (WHO) defines a leprosy patient as someone who has the disease and has not completed antibacterial treatment. After completing multidrug therapy with dapsone, rifampicin and clofazimine, the person is no longer considered a leprosy patient, even if they are permanently disabled because of the disease. In clinical practice, the traditional classification of leprosy is still used, with lepromatous (nodular) leprosy at one end of the spectrum and tuberculoid leprosy at the other. Treatment duration according to WHO guidelines depends on whether the condition is classified as paucibacillary (PB)/tuberculoid leprosy or multibacillary (MB)/lepromatous leprosy. Six months of drug therapy is recommended for PB leprosy, and twelve months for MB leprosy (2, 4).

Immune-mediated neuropathy in the form of neuritis can occur acutely as part of a reaction, or develop more gradually. In the acute phase, it is generally treated with steroids (20 weeks of oral prednisolone) to potentially save nerve fibres from permanent damage. The dose is adjusted according to the clinical response (4).



Richard Tennant Cooper (1885–1957). Painted to order from Henry S. Wellcome, ca. 1912. Wellcome Collection / CC BY 4.0

Clinical presentation of 'surgical leprosy'

A patient's face can be affected where various facial nerves are relatively close to the surface of the skin. The result can be an inability to close the eyelid (lagophthalmos), often in combination with sensory loss in the cornea. Leprosy can also damage mucous membranes and cartilage in the nose as well as hair follicles in the eyebrows (6).

In the upper limbs, the ulnar nerve at the elbow joint is typically affected. The result is claw hand, due to paralysis of the small hand muscles and sensory loss in the little finger and half of the ring finger. The median nerve can be affected at the wrist. The result is loss of thumb opposition and sensory loss in the three radial fingers. The radial nerve has mostly a 'deep and warm' location and is very rarely affected. An exception is distal sensory branches in the forearm and hand, but these have little clinical significance (6, 7).

In the lower extremities, the fibular nerve is often affected where it lies superficially over the proximal fibula. The result is foot drop, which is an inability to dorsiflex the foot and toes. Involvement of the tibial nerve occurs where it is relatively superficial, behind the medial malleolus. The clinical outcome is paralysis of the intrinsic foot muscles and claw toe deformity. There is also significant sensory loss in the foot. Autonomic function impairments result in dry, cracked skin, which predisposes to ulceration and infections. Deep infections with extensive necrosis of soft tissue and bone are common in leprosy patients in developing countries (7). So-called Charcot foot (inflammation of the foot and ankle joints) is a dreaded condition that can lead to significant deformities.

Leprosy surgery

Leprosy surgery is often referred to as an unknown or little-known specialty. Published literature on the subject is sparse compared to the many articles on Norway's historical achievements, the epidemiological aspects of leprosy, the infectious disease itself and, not least, its fascinating immunology.

In summary, leprosy surgery is generally aimed at addressing complications resulting from peripheral neuropathy. Nerve damage leads to the loss of autonomic functions and protective sensation, as well as muscle paresis. A range of surgical procedures has been developed for the face to restore a more normal appearance.

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Neuropathy caused by leprosy significantly impacts hand function. A numb hand is 'blind'. The grip function can also be impaired to varying degrees. Various surgical techniques can be used to improve grip function (7).

Neuropathic foot problems are also common. Foot drop can be corrected with tendon transfer surgery (8). Several other procedures can also be performed to address chronic ulcers and deformities. Many of the leprosy surgery techniques have also been adopted for other types of neurological deficits (7, 8). Tendon transfer is one of the most common reconstructive procedures performed on leprosy patients. This essentially involves relocating tendons (and their muscle

power) with intact function to replace muscles with loss of function. As the 'motor' (donor muscle), muscles with 'less critical function' are used to restore more vital function (9).

Large, chronic heel ulcers, where the soft tissue of the heel pad is virtually lost, were previously an indication for leg amputation. However, new surgical methods have been developed to reduce the number of amputations. Using an island flap technique, healthy tissue from the instep of the sole can be transferred to the heel pad (10, 11).

The big pioneers

Most authors who write about leprosy cannot avoid mentioning the Bergen consultants Daniel Cornelius Danielssen and Gerhard Armauer Hansen, as well as the Norwegian leprosy registry (1, 12, 13). References to the pioneers in leprosy surgery are, however, scarcer.

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Sterling Bunnell (1882–1957) was considered the 'founder of hand surgery'. During World War II, he was tasked with establishing ten regional hand surgery units for the many tens of thousands of American soldiers with hand injuries. About one-third of the combat-related injuries involved the hands. Bunnell's voluminous textbook from 1944, *Surgery of the Hand*, laid the foundation for hand surgery as a surgical specialty.

Professor Paul Brand (1914–2003) is a towering figure in leprosy surgery. He was an exceptionally gifted individual, as a craftsman, surgeon and researcher, and above all, he had deep compassion for every patient. As a missionary child in India, he was sent back to England at the age of nine to attend school. He trained as a carpenter and engineer before pursuing medical studies and qualifying as a specialist in orthopaedic surgery. He later returned to India, where he dedicated most of his life to leprosy surgery and was a true pioneer in the field.

Brand was the first to attempt reconstructive surgery for leprosy. He first had to demonstrate that the patients had normal healing ability and that the surgical wounds healed properly, which some of those in authority doubted. This was an absolute prerequisite for any surgical intervention. Over time, he developed techniques such as tendon transfers to correct claw hand deformities and the lack of thumb opposition. His hand surgery textbook *Clinical Mechanics of the Hand* (1985) was groundbreaking work that was appreciated by hand surgeons worldwide. Brand also worked extensively on designing

appropriate footwear and finding suitable materials for numb and deformed feet. He also developed tendon transfer techniques to correct patients' lack of ankle dorsiflexion. These techniques are now used worldwide for other neurological disorders and for injuries from foot drop (8). In 1966, Brand was headhunted to Louisiana, where he was appointed head of the United States' only leprosy hospital, The National Leprosarium, in Carville. He also took up a professorship at the University of Washington (14).

The Danish plastic surgeon Johannes Andersen (1922–2005) was one of Paul Brand's trainees, and he continued Brand's work with service in countries such as India, Nepal, Tanzania, Kenya and Ethiopia. He defended his thesis on leprosy in Denmark (15). At the ALERT Hospital (All Africa Leprosy Rehabilitation and Training Centre) in Addis Ababa, he was succeeded by one of the authors of this article (Paul Egil Gravem).

Discrimination and social exclusion

Leprosy is not, however, only an infectious, immunological and surgical problem. Patients in many places are subjected to discrimination and social exclusion (6). For several hundred years, special leprosy hospitals and isolated leper colonies reinforced the stigma of 'unclean' patients.

The Bible's characterisation of lepers has also been blamed for the discrimination these patients have faced. In earlier translations of the Bible, the word *leper* was used around 20 times, particularly in the Old Testament. However, most of these passages referred to diseases that were clearly not leprosy. Fortunately, this translation error was corrected in more recent translations of the Bible (16).

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Publisert: 14 February 2025. Tidsskr Nor Legeforen. DOI: 10.4045/tidsskr.24.0642
Received 4.12.2024, first revision submitted 12.12.2024, accepted 14.12.2024.