Leprosy

Revision Dates

Case Definition	August 2011
Reporting Requirements	May 2018
Remainder of the Guideline (i.e., Etiology to References sections inclusive)	June 2005

Case Definition

Confirmed Case

Clinical illness[A] with laboratory confirmation of infection:

- Detection of *Mycobacterium leprae* nucleic acid (e.g., PCR) in an appropriate clinical specimen (e.g., tissue biopsy usually skin nodes, nasal scrapings)[B]

OR

- Positive Acid Fast stain with typical morphology for *M. leprae*

OR

- Histopathological report from skin or nerve biopsy compatible with leprosy.

Probable Case

Clinical illness[A] in a person who is epidemiologically linked to a confirmed case.

[A] Clinical illness is characterized by the involvement primarily of skin as well peripheral nerves and the mucosa of the upper airway. Clinical forms of Hansen's disease represent a spectrum reflecting the cellular immune response to *Mycobacterium leprae*. The following characteristics are typical of the major forms of the disease:
Tuberculoid: one or a few well-demarcated, hypo-pigmented, and anesthetic skin lesions, frequently with active, spreading edges and a clearing center; peripheral nerve swelling or thickening also may occur.
Lepromatous: a number of erythematous papules and nodules or an infiltration of the face, hands and feet with lesions in a bilateral and symmetrical distribution that progress to thickening of the skin.
Borderline (dimorphous): skin lesions characteristic of both the tuberculoid and lepromatous forms.
Indeterminate: early lesions, usually hypopigmented macules, without developed tuberculoid or lepromatous features.

[B] Refer to the Provincial Laboratory for Public Health (ProvLab) Guide to Services for current specimen collection and submission information.

Reporting Requirements

1. **Physicians, Health Practitioners and others**
   Physicians, health practitioners and others shall notify the Medical Officer of Health (MOH) (or designate) of the zone, of all confirmed and probable cases in the prescribed form by mail, fax or electronic transfer within 48 hours (two business days).

2. **Laboratories**
   All laboratories shall report all positive laboratory results by mail, fax or electronic transfer within 48 hours (two business days) to the:
   - Chief Medical Officer of Health (CMOH) (or designate), and
   - MOH (or designate) of the zone.

3. **Alberta Health Services and First Nations Inuit Health Branch**
   - The MOH (or designate) of the zone where the case currently resides shall forward the initial Notifiable Disease Report (NDR) of all confirmed and probable cases to the CMOH (or designate) within two weeks of notification and the final NDR (amendment) within four weeks of notification.
   - For out-of-province and out-of-country reports, the following information should be forwarded to the CMOH (or designate) by phone, fax or electronic transfer within 48 hours (two business days):
     - name,
     - date of birth,
     - out-of-province health care number,
     - out-of-province address and phone number,
     - positive laboratory report, and
     - other relevant clinical / epidemiological information.

Etiology (1)

Leprosy is caused by *Mycobacterium leprae*, an acid-fast bacillus. The organism multiplies slowly. It occurs in large numbers in the lesions of lepromatous leprosy and has a unique ability to enter nerves. *M. leprae* can remain potent even when dried out.

Clinical Presentation (2,3)

Leprosy, also known as Hansen's Disease, is a chronic disease predominantly involving the skin, peripheral nerves and/or the mucosa of the upper airway. The bacterium tends to invade cooler areas of the body such as the chin, cheekbones, earlobes, knees, and distal extremities. *M. leprae* is the only bacterium that invades the peripheral nerves. Almost all of the complications are a result of that invasion.

Manifestations depend on the infected person's immune response to the bacterium. The clinical syndromes of the disease represent a spectrum that reflects the cellular immune response to the bacteria.

Tuberculoid leprosy (paucibacillary form) features one or a few skin lesions. The lesions are typically raised with active spreading edges, a central clearing and are bilaterally asymmetrical. Peripheral nerve involvement tends to be present.

Numerous macules, papules, nodules, and plaques with bilaterally symmetrical distribution characterize lepromatous leprosy. There may be involvement of the nasal mucosa leading to crusting, difficulty breathing, and epistaxis. Ocular involvement leading to iritis and keratitis may also occur.

Borderline leprosy has skin lesions similar to both tuberculoid and lepromatous lesions but with a raised center area, depending on treatment or no treatment.

Indeterminate leprosy is an early form of the disease. It is manifested by a hypopigmented macule with ill-defined borders and, if untreated, may progress to tuberculoid, borderline or lepromatous disease.

Serious consequences of leprosy may occur from immune reactions i.e., acute adverse episodes which are termed erythema nodosum leprosum (ELM) in lepromatous patients and reversal reactions in borderline leprosy. Other serious consequences may occur as a result of nerve impairment with resulting anesthesia that may lead to trauma, fractures, and bone resorption.

Diagnosis (1)

The diagnosis is usually made based on anesthesia of a skin lesion or in the distribution of a peripheral nerve, thickened nerves, and typical skin lesions, as well as skin smear results. The organism does not grow on bacteriologic media or cell cultures. It may be grown on mouse footpads. Skin scrapings/biopsy should be submitted to the PLPH and examined for acid-fast bacteria. There are two indices for the quantification of infection: the bacteriological index (BI: measure of bacterial load) and the morphological index (MI: measure of bacterial viability).

Epidemiology

Reservoir

Humans are the only significant reservoir. Naturally acquired disease has been identified in a mangabey monkey (Nigeria) and a chimpanzee (Sierra Leone). Feral armadillos in Texas and Louisiana have been found to be naturally afflicted, and there have been reports suggesting transmission of disease to humans.

Transmission

The mode of transmission remains unclear but it is not highly communicable. It has been suggested that person to person transmission occurs from aerosolized droplets and, less commonly, by direct contact. The primary site of inoculation is the nose. The number of bacilli in a sneeze is very high. Bacilli have been shown to remain viable for at least seven days in dried nasal secretions. In addition, cutaneous ulcers may also shed large numbers of bacilli.

Skin trauma may allow for a point of entry of organisms. As well, insect vectors (bed bugs and mosquitoes) may also play a role in transmission. Transmission is presumed to be transplacental when disease occurs in children less than one year of age.

Incubation Period

The incubation period is long ranging from one to several years but is typically three to five years. The incubation period of tuberculoid cases tends to be shorter than that for lepromatous cases. The long incubation period makes it difficult to determine where or when the disease was contracted.

Period of Communicability

The infectivity most likely ceases within three months of beginning treatment but may be sooner depending on the treatment provided.

Host Susceptibility (4)

Population studies indicate that over 90% of individuals can resist leprosy successfully. Some research suggests that possible genetic factors may determine if an individual will develop leprosy and the type. Children are more susceptible than adults, but the disease is rarely seen in children under the age of three. Pregnant women have decreased cell-mediated immunity and thus may have an increased risk at developing infection. Risk groups include close contacts of patients with active untreated disease and persons living in countries with highly endemic disease.

Occurrence

General (1,2,4-6)

Leprosy is one of the oldest known pathogens to afflict humans. It has been recognized since the ancient civilizations of China, Egypt, and India dating back to 600 B.C. *M. Leprae* was discovered in 1872 by G. A. Hansen. It was the first bacterium identified as causing human disease.

Global prevalence is decreasing. In 1977, the WHO estimated there to be 1.15 million cases of leprosy worldwide. The prevalence in 1994 was estimated to be 2.4 million cases and in 2000, there were approximately 739,000 cases. Currently, approximately half a million cases are detected worldwide each year. Rates of more than 5/1,000 have been estimated in the rural tropics and subtropics.

The infection can start at any age but most commonly it begins in individuals in their twenties and thirties. The severe form of leprosy (lepromatous) has a male to female ratio of 2:1. The milder form (tuberculoid) occurs equally in both sexes. Leprosy occurs in all races. Individuals with light colored skin and Asians have a tendency to acquire the lepromatous type and African blacks have a higher incidence of the tuberculoid form.

The WHO has listed 91 countries as endemic including temperate, tropical, and subtropical climates. The chief endemic areas are South and Southeast Asia including the Philippines, Indonesia, Papua New Guinea, some Pacific Islands, India, Bangladesh, Burma, and tropical Africa and some areas of Latin America. Six countries accounted for 90% of the prevalence worldwide in early 2002. These countries include Brazil, India, Madagascar, Mozambique, Myanmar (Burma), and Nepal. Approximately 70% are cases registered in India, Myanmar and Nepal. In the United States, the majority of cases are imported occurring in immigrants and refugees from endemic areas, however, in Texas, California, Louisiana, and Hawaii indigenous cases continue to occur.

Canada (3, 7)
Leprosy is rare in Canada, however, increasing rates of immigration from countries where leprosy is endemic have led to the recognition of leprosy in Canada (North America). Transmission of the disease within Canada has never been reported. From 1989 to 1998, 114 cases of leprosy were reported. The number of cases reported annually ranged from 21 cases in 1991 to three cases in 1998. The prevalence is estimated at 0.6/100,000 population.

Alberta (8)
Between 1998 and 2000, five cases of leprosy were reported in Alberta. All cases were acquired outside Canada.

Key Investigation

Single Case/Household Cluster

- History of immigration from an endemic area.
- Past history of leprosy.
- Prolonged exposure to a family member or other contact with leprosy.
- Travel to an area of the world where leprosy is endemic.

Control

Management of a Case

- No public heath intervention is required, as the risk of communicability is low especially after initiation of treatment.
- Persons with leprosy require medical follow up by an infectious diseases specialist.
- Routine practices.
- Handwashing is the most effective measure to prevent transmission.
- AHW maintains a leprosy registry for the purpose of public health follow-up of persons receiving treatment.
- The MOH (or designate) is sent a letter every two years to request updates on the case.
    - The MOH (or designate) is responsible for ensuring that appropriate specimens (skin scraping/biopsy) are submitted.
    - When an individual has cleared the disease, moved, lost to follow-up or deceased they are removed from the active registry and placed on the inactive registry.

Treatment of a Case

- Leprosy is a curable disease. Early treatment averts disability.(6)
- Generally treatment is individualized, however, the standard regimen is as follows.
  - For tuberculoid leprosy, rifampin, and dapsone (DDS) are taken for six months.
  - For lepramatous leprosy, multiple drug therapy consisting of daily rifampin, DDS, and clofazimine is continued until skin smears are negative for two years. Antibiotic therapy must be continued for a long period of time because the bacteria are difficult to eradicate.(G Taylor, personal communication, April 15, 2003)
- Patients receiving treatment should be monitored for drug side effects, for leprosy reactions, and for the development of trophic ulcers. Consultation with an infectious diseases physician is recommended.
- Medication is provided free of charge from AHW. Contact the CMOH (or designate). (T Mersereau, personal communication, June 2003)

Management of Contacts

- The initial examination of contacts and periodic examination of household and other close contacts for skin lesions is recommended annually for up to five years after the last contact with an infectious case.

Preventive Measures

- Early diagnosis and treatment is the best preventive measure.
- Health education.

References

(1) Harrop E. *Leprosy*. Emedicine 2002. http://www.emedicine.com/derm/topic223.htm

(2) *Leprosy*. The Merck Manual Second Home Edition Online. Infections. 2004. http://www.merck.com/mmhe/sec17/ch104/ch194a.html

(3) Boggild A, Keystone J, Kain K. *Leprosy: A primer for Canadian physicians*. CMAJ 2004;170(1):71-78.

(4) *Leprosy*. MedlinePlus Medical Encyclopedia. January 2003. http://www.nlm.nih.gov/medlineplus/ency/article/001347.htm

(5) *Leprosy: the disease*. World Health Organization. 2004 http://www.who.int/lep/disease/frmain.htm

(6) *Leprosy*. World Health Organization. Fact Sheet No. 101. January 2003. http://www.who.int/mediacentre/factsheets/fs101/en/print.html

(7) Public Health Agency of Canada. *Notifiable Diseases On-Line - Leprosy*. November 2000. http://dsol-smed.phac-aspc.gc.ca/dsol-smed/ndis/disease2/lepr_e.html

(8) Alberta Health & Wellness, Disease Control and Prevention. *Notifiable Diseases – Alberta*. Communicable Disease Reporting System Mid Year Population. March 2003.

