

Hansen's Disease (Leprosy)

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Section 1

ABOUT THE DISEASE

A. Etiologic Agent

Hansen's disease (also called leprosy) is a chronic infectious disease caused by the bacterium *Mycobacterium leprae*.

B. Clinical Description

Hansen's disease (HD) is a chronic bacterial disease characterized by the involvement primarily of skin, as well as peripheral nerves and the mucosa of the upper airway. Clinical forms of HD 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, hypopigmented, and hypoesthetic or 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 infiltrate of the face, hands, and feet with lesions in a bilateral and symmetrical distribution that progress to thickening of the skin, possibly with reduced sensation.
- *Borderline (dimorphous)*: skin lesions characteristic of both the tuberculoid and lepromatous forms
- *Indeterminate*: early lesions, usually hypopigmented macules, without developed tuberculoid or lepromatous features but with definite identification of acid-fast bacilli in biopsies.

Borderline HD tends to shift toward the lepromatous form in the untreated patient and toward the tuberculoid form in the treated patient. Indeterminate leprosy is an early form that may develop into any of the other forms.

C. Vectors and Reservoirs

Humans are the only reservoir of proven significance for HD, however evidence suggests that leprosy in armadillos may be naturally transmitted to humans.

D. Modes of Transmission

The exact mechanism for the acquisition and transmission of HD is not known, however, it is not acquired from casual contact, such as shaking hands, sitting next to someone on a bus, or sitting together at a meal. HD is far less contagious than other infectious diseases. More than 95 percent of the human population has a natural immunity to the disease. Healthcare workers rarely contract HD. Most cases of HD respond to treatment and become non-infectious within a very short time. Household contact and prolonged close contact, however, may result in transmission. Large numbers of the organism are shed in the nasal discharge of untreated patients with lepromatous HD, and the bacilli may remain viable in dried nasal

secretions for up to seven days. Large numbers of bacilli are also shed in the skin lesions in the lepromatous form of HD.

E. Incubation Period

The bacteria that cause HD grow very slowly. Signs and symptoms may not appear for 2 – 20 years.

F. Period of Communicability or Infectious Period

Clinical and laboratory evidence suggests that infectiousness is lost in most instances within a day of beginning treatment with multi-drug therapy.

G. Epidemiology

As of 2015, a total of 13,950 HD cases were registered in the National Hansen’s Disease Registry in the United States since 1894, 178 of which were added in 2015. While 57% of the 2015 cases recorded a location other than the US as their place of birth, more than two-thirds of cases from Texas, Louisiana, Arkansas, Mississippi, and Florida were native-born US citizens with no residence history outside the US. Evidence suggests that zoonotic transmission from nine-banded armadillos is the principal source of infection perpetuating infection in these locales.

Based on estimates of life expectancy, some 9,140 of total cases in the US are potentially still living. The National Hansen’s Disease Registry is administered by the Health Resources and Services Administration, Healthcare Systems Bureau’s Division of National Hansen’s Disease Programs.

In 2014, the World Health Organization reported that only 213,899 new cases were registered worldwide, representing a greater than 60 percent decline in annual new case numbers since 2001.

H. Bioterrorist Potential

This pathogen is not considered to be of risk for use in bioterrorism.

Section 2

REPORTING CRITERIA AND LABORATORY TESTING

A. What to Report to the Massachusetts Department of Public Health (MDPH)

- Report any case with demonstration of acid fast bacilli in skin or dermal nerve from a biopsy of a skin lesion using FITE stain, without growth of mycobacteria on conventional media OR
- Identification of noncaseating granulomas with peripheral nerve involvement, without the growth of mycobacteria on conventional media (if done)

B. Laboratory Testing Services Available

The MDPH State Public Health Laboratory (MA SPHL) can detect *M. leprae* by AFB smear microscopy of skin biopsy specimens. Although it is not possible to grow *M. leprae* in either bacteriological or in cell culture, further testing of specimens using DNA amplification techniques may be coordinated between the MA SPHL and the Centers for Disease Control and Prevention (CDC).

For more information about testing, call the MA SPHL Mycobacteriology Laboratory at (617) 983-6381.

Section 3

REPORTING RESPONSIBILITIES AND CASE INVESTIGATION

A. Purpose of Surveillance and Reporting

- To identify infection and possible modes of acquisition

B. Laboratory and Health Care Provider Reporting Requirements

HD is reportable to the local board of health (LBOH). The MDPH requests that health care providers immediately report to the LBOH in the community where the case is diagnosed, all confirmed or suspect cases of HD, as defined by the reporting criteria in Section 2A above.

Laboratories performing examinations on any specimens derived from Massachusetts residents that yield evidence of HD shall report such evidence of infection directly to the MDPH within 24 hours.

C. Local Board of Health (LBOH) Reporting and Follow-Up Responsibilities

Reporting Requirements

MDPH regulations (*105 CMR 300.000*) stipulate that HD is reportable to the LBOH and that each LBOH must report any confirmed or suspect case of HD as defined by the reporting criteria in Section 2A. Cases should be reported to the MDPH Bureau of Infectious Disease and Laboratory Sciences (BIDLS), Office of Integrated Surveillance and Informatics Services (ISIS) via MAVEN. Refer to the List of Diseases Reportable to Local Boards of Health for information on prioritization and timeliness requirements of reporting and case investigation <http://www.mass.gov/eohhs/docs/dph/cdc/reporting/rprtbdiseases-lboh.pdf>

Case Investigation

It is the responsibility of the LBOH to complete all questions in each of the question packages by interviewing the case and others who may be able to provide information. Much of the information required can be obtained from the health care provider or from the medical record.

Calling the provider

If the case was hospitalized (i.e. reporting facility is a hospital), call infection control at the named hospital. A list of infection preventionists can be found in the help section of MAVEN. If the case was seen at a clinician's office, ask to speak to a nurse working with the ordering provider.

Calling the case or parent/guardian of the case

Before calling the case, review the disease fact sheet by clicking on the Help Button located in MAVEN and review all the information in this chapter. The call may take a few minutes, so in order to maximize the chance of getting the information needed, it might be good to note the potential length of the call with your contact, and offer the opportunity to call back when it is more convenient. Asking questions about how the case or child is feeling may get the case or parent talking. If you are unable to answer a question they have, don't hesitate to call the Division of Epidemiology and Immunization at 617-983-6800 for assistance, and call the case back with the answer later. People are often more than willing to talk about their illness, and they may be very happy to speak with someone who can answer their questions.

Using MAVEN

Administrative Question Package

Monitor your Online "LBOH Notification for Routine Disease" workflow in MAVEN for any new cases of HD. Once a new event appears in this workflow, open the Administrative Question Package (QP) and under the "Local Health and Investigation" section, answer the first question "**Step 1** - LBOH acknowledged" by selecting "Yes". The "LBOH acknowledged date" will then auto populate to the current day. Completing this first step will move the event out of this workflow and into your "Online LBOH notified but Case Report Forms (CRF) are pending" workflow. Note the date you started your investigation by answering "**Step 2** - Investigation started" as "Yes" and then note the date where shown. Record your name, agency, and phone numbers where shown in "**Step 3** - LBOH/Agency Investigator."

Demographic Question Package

Record all demographic and employment information. It is particularly important to complete the Race/Ethnicity and Occupation questions.

Clinical Question Package

Complete the "Diagnosis/Clinical Information" section, providing the diagnosis date, symptom information and date of symptom onset, hospitalizations, and other medical information

Risk Exposure/Control & Prevention Question Package

Accurately record the answers to all risk questions regarding travel. You will be able to list multiple places of travel. Ask questions about place of birth to determine if a person has resided or was born in a country endemic for HD. If a person traveled to Texas, Louisiana, Mississippi, Arkansas, or Florida, inquire about contact with armadillos and record it in the notes section. There is evidence that armadillos might be a source of exposure for the bacteria. As you enter data into MAVEN, additional questions will appear for you to answer regarding risk/exposure. Ask about household contacts and other contacts to determine the possible source of infection as well as whether others have been exposed. Record as much treatment information as possible.

Completing Your Investigation

1. If you were able to finish the case investigation and follow-up is complete, mark "**Step 4** - Case Report Form Completed" as "Yes" and then choose Local Board of Health (LBOH) -Ready for MDPH review for the Completed by variable.

2. If you have made several attempts to obtain case information but have been unsuccessful (e.g., the case or health care provider does not return your calls or respond to a letter, or the case refuses to divulge information or is too ill to be interviewed), please complete “**Step 4 - Case Report Form Completed**” as “No” and then choose a primary reason why the case investigation was not completed from the choices provided in the primary reason answer variable list.
3. If you are not online for MAVEN you may submit a paper case report form. After completing the form, attach laboratory report(s) and fax or mail (in an envelope marked “Confidential”) to ISIS. The confidential fax number is (617) 983-6813. Call ISIS at (617) 983-6801 to obtain a copy of the case report form and to confirm receipt of your fax.

The mailing address is:

MDPH, Office of Integrated Surveillance and Informatics Services (ISIS)
305 South Street, 5th Floor
Jamaica Plain, MA 02130
Fax: (617) 983-6813

Section 4

CONTROLLING FURTHER SPREAD

A. Isolation and Quarantine Requirements (105 CMR 300.200)

Minimum Period of Isolation of Patient

No restrictions if under continuous medical care.

Minimum Period of Quarantine of Contacts

No restrictions.

B. Protection of Contacts of a Case

Health education should stress that patients under continuous medical treatment are not infectious, however, examination and follow up of close contacts is recommended. Household contacts of people with HD should have a thorough physical examination annually for five years. If they develop a questionable skin rash, they should notify their healthcare providers and have the skin rash biopsied to determine whether or not HD is present

C. Managing Special Situations

Response to Community Perceptions

Community and individual perceptions about HD may reflect inaccurate concerns about communicability and about the health implications for those diagnosed. These concerns may not be valid with regard to the nature of the disease, treatment, and prevention methods. It is important to convey to all concerned

parties, the low communicability of this disease and the availability of effective treatment and prevention regimens.

D. Preventive Measures

Education of the case should stress the availability and efficacy of therapy. Additionally, education of the case's household contacts should include modes of transmission and referral to a health care provider for follow-up. People with Hansen's disease in the U.S. can receive Hansen's disease medications at no cost through their own doctor or through the National Hansen's Disease Program Ambulatory Care Clinic closest to them. For further information: National Hansen's Disease Programs toll-free, weekdays 9 a.m. to 5:30 p.m. at 1-800-642-2477.

It is important to convey to the case and to the contacts the very low communicability of this disease and the availability of effective treatment and prevention regimens.

ADDITIONAL INFORMATION

The following is the formal CDC surveillance case definition for HD. It is provided for your information only and should not affect the investigation or reporting of a case that fulfills the criteria in Section 2A of this chapter. (The CDC and the MDPH use the CDC case definitions to maintain uniform standards for national reporting.) For reporting a case to the MDPH, always use the criteria outlined in Section 2A.

Note: The most up-to-date CDC case definitions are available on the CDC website at www.cdc.gov/epo/dphsi/casedef/case_definitions.htm.

Clinical Description

A chronic bacterial disease characterized by the involvement primarily of skin as well as peripheral nerves and the mucosa of the upper airway. Clinical forms of HD represent a spectrum reflecting the cellular immune response to *Mycobacterium leprae*. The following characteristics are typical of the major forms of the disease, *though these classifications are assigned after a case has been laboratory confirmed*.

- *Tuberculoid*: one or a few well-demarcated, hypopigmented, and hypoesthetic or 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, possibly with reduced sensation.
- *Borderline (dimorphous)*: skin lesions characteristic of both the tuberculoid and lepromatous forms
- *Indeterminate*: early lesions, usually hypopigmented macules, without developed tuberculoid or lepromatous features but with definite identification of acid-fast bacilli in Fite stained sections

Laboratory Criteria for Diagnosis

- Demonstration of acid fast bacilli in skin or dermal nerve from a biopsy of a skin lesion using Fite stain, without growth of mycobacteria on conventional media (if done) **OR**
- Identification of noncaseating granulomas with peripheral nerve involvement, without growth of mycobacteria on conventional media (if done)

Case Classification

Confirmed

A clinically compatible case that is laboratory-confirmed.

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