Kansas Department of Health and Environment
Division of Health

Hansen's disease (Leprosy) Investigation Guideline

Contents

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Revision History:

Date	Replaced	Comments
05/2018	12/2016	Updated format and web links. Updated Notification sections and Isolation with updated regulations.
01/2013	07/2010	New case definition; added notification section; updated web links. Updated fact sheet. Removed references to KS-EDSS in 02/2012.

Hansen's disease (Leprosy)
Disease Management and Investigation Guidelines

CASE DEFINITION – (CDC 2013)

Clinical Description for Public Health Surveillance:

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 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, *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 Case Classification:

Confirmatory:

- 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 illness that is laboratory confirmed.

LABORATORY ANALYSIS:

The state laboratory does not require isolates to be sent and does not provide testing for *M. leprae*. Skin biopsy is needed for definitive diagnosis, and PCR for *M. leprae* DNA may be needed in special circumstances

Indications for skin biopsy: Patient with a non-responsive skin lesion and is:

1) Immigrant from country with high incidence of leprosy
2) U.S. resident with history of foreign travel
3) Resident of Texas or Louisiana
4) Has history of multiple physician/specialist and/or emergency room visits

Biopsy: Obtain a full-thickness biopsy (important to see a bit of subcutaneous fat) from the most active margin. An elliptical or punch biopsy (4 mm) is sufficient.

Fixation and processing: Routine 10% neutral buffered formalin OR embedded in paraffin by a pathology laboratory

Testing can be performed at the National Hansen's Disease Programs (NHDP):

Clinical Laboratory
National Hansen's Disease Programs
1770 Physician Park Dr.
Baton Rouge, LA 70816
Tel 225-756-3733

- For additional information: www.hrsa.gov/hansensdisease/diagnosis/index.html.

EPIDEMIOLOGY

Leprosy occurs worldwide particularly in South and Southeast Asia, tropical Africa and some areas of Latin America. Between 150 and 200 new U.S. cases are reported annually. The largest numbers of U.S. cases are in California, Texas, Hawaii, Louisiana, Florida, New York and Puerto Rico within the persons migrating from endemic areas. Indigenous cases also occur in Texas, California, Louisiana, Hawaii and Puerto Rico.

DISEASE OVERVIEW

A. Agent:
Mycobacterium leprae, an acid-fast, gram-positive bacillus.

B. Clinical Description:
Leprosy is a chronic bacterial disease of the skin, peripheral nerves and/or the upper airway with a broad range of clinical manifestations. The skin involvement can be either nodular/papular or restricted to the level of skin. (Review the case definitions for characteristics of major forms of the disease.)

General features include:

- Hypopigmented or reddish skin lesion(s) with definite loss of sensation
- Enlargement and tenderness of the peripheral nerves with definite thickening and loss of sensation (hyperesthesis, anesthesia, paralysis, muscle wasting or trophic ulcers).

Clinical forms of leprosy reflect the cellular immune response to *M. leprae*; therefore, leprosy may be masked in patients with advanced HIV disease, and only seen after immune reconstitution while under retroviral treatment.

More than 25 percent of patients may have reactive episodes ("reactions") of varying severity during the course of disease. Some before treatment is started or after therapy is completed, but most during therapy, particularly during the first year. Erythema nodosum leprosum (ENL) manifests with fever and painful erythematous nodules, but peripheral neuritis, orchitis, lymphadenitis, iridocyclitis, nephritis, periostitis and arthralgias may occur. "Reversal reactions" are characterized by edema and erythema of pre-existing lesions. Neuritis and occasionally new lesions or fever may also occur. A rare Lucio's phenomenon can occur where multiple ulcers of varying size develop that are often difficult to heal.

Injuries are common in all patients with Hansen's disease who have significant degrees of sensory and motor loss.

C. Reservoirs:

Humans. Feral armadillos in Louisiana and Texas have been found naturally afflicted; however, transmission to humans is uncertain.

D. Mode(s) of Transmission:

Most commonly accepted theory is transmission by way of the respiratory tract, since large numbers of bacteria can be found in the nose of some untreated patients. The degree of susceptibility of the person, the extent of exposure and environmental conditions are among factors important in transmission.

E. Incubation Period:

Ranges from 9 months to 20 years, but usually 3 to 5 years.

F. Period of Communicability:

Clinical and laboratory evidence suggest infectiousness is lost usually within a day of beginning treatment with multidrug therapy.

G. Susceptibility and Resistance:

The high prevalence of antibodies specific for *M. leprae* among close contacts suggests that infection is frequent; however, clinical disease occurs in only a small proportion of these contacts. More than 95 percent of the human population has a natural immunity to the disease.

H. Treatment:

Refer to the National Hansen Disease Program's recommended treatment: www.hrsa.gov/hansensdisease/diagnosis/recommendedtreatment.html.

NOTIFICATION TO PUBLIC HEALTH AUTHORITIES

Cases of Hansen's disease (regardless of laboratory evidence) shall be reported within 24 hours, except if the reporting period ends on a weekend or state-approved holiday, the report shall be made by 5:00 p.m. on the next business day after the 24-hour period:

1. Health care providers and hospitals: report to the local public health jurisdiction or KDHE-BEPHI (see below)
2. Local public health jurisdiction: report to KDHE-BEPHI (see below)
3. Laboratories: report to KDHE-BEPHI (see below)

Kansas Department of Health and Environment (KDHE)
Bureau of Epidemiology and Public Health Informatics (BEPHI)
Phone: 1-877-427-7317
Fax: 1-877-427-7318

Further responsibilities of state and local health departments to the CDC:

As a nationally notifiable condition, confirmed Hansen's disease is ROUTINELY NOTIFIABLE to the Center of Disease Control and Prevention (CDC).

- **Local public health jurisdiction** will report information requested on the disease reporting forms as soon as possible.
- KDHE-BEPHI will file an electronic case report the next regularly scheduled electronic transmission.
  (KDHE-BEPHI files electronic reports weekly with CDC.)

INVESTIGATOR RESPONSIBILITIES

1) Report all confirmed, probable and suspect cases to the KDHE.
2) Use current case definition, to confirm diagnosis with the medical provider.
   - Collect all information requested in Step 1 of case investigation.
   - Ensure that case/proxy is aware of the diagnosis.
3) Conduct case investigation to identify potential source of infection.
4) Conduct contact investigation of close contacts.
5) Complete all information requested in the EpiTrax.
6) Use the disease fact sheets, as needed.

STANDARD CASE INVESTIGATION AND CONTROL METHODS

Case Investigation

1) Contact the medical provider who ordered testing of the case and obtain the following information. (This includes medical records for hospitalized patients.)
   Note: *If the physician, submitted samples to NHDP, a Hansen's disease Surveillance Form may already be completed or started – obtain a copy.*
   - Collect patient's demographics (address, birth date, gender, race/ethnicity, primary language, and phone number(s)). [Demographic]
   - Record patient's occupation [Epidemiologic]
   - Examine laboratory testing: record biopsy and skin smear results, dates.
   - Record onset date of symptoms and type of leprosy diagnosed. [Clinical]
   - Record diagnose date. [Clinical]
   - Record current treatment [Clinical]
   - Record hospitalizations: location and duration of stay [Clinical]
   - Record outcomes: survived or date of death [Clinical]
2) Interview the case to determine source and risk factors:
   - Record patient's residence in U.S. or other countries starting at present and going back 5 years from the onset date (include military service)
   - Identify any risk factors, including:
     - Immigrant from country with high incidence of leprosy
     - U.S. resident with history of foreign travel
     - Resident of Texas or Louisiana
     - History of touching an armadillo. [Investigation – Exposure]
3) Examining the epidemiological information:
   - Record where the infection was most likely imported from. (Indigenous or out-of-county, state, or U.S.) [Epidemiologic].
   - Determine if case had contact with a suspect Hansen's disease case.
   - For suspected outbreaks refer to Managing Special Situations section.

Contact Investigation

More than 95 percent of the human population has a natural immunity to the Hansen's disease. Most cases of Hansen's disease respond to treatment and become non-infectious within one day of treatment.

Those at greatest risk are the family of a person with untreated disease. Risk is based on genetic susceptibility and/or prolonged contact with the infected case. A spouse is the least at-risk family member. At greatest risk are children, brothers or sisters, or parents of an individual with untreated Hansen's disease.

Hansen's disease is not passed on from a mother to her unborn baby during pregnancy. Neither is it transmitted through sexual contact

- Collect names and contact information of current household members or family members with close household exposure to untreated cases.

Isolation, Work and Daycare Restrictions

Isolation is not necessary. No restrictions in employment or school attendance

Case Management

A Cases will be managed by attending medical provider. **Physician awareness is key to the early diagnosis and treatment that can prevent disability.**

Medical providers can access information on the U.S. Health Resources and Administration (HRSA) National Hansen's Disease Program (NHPD) website at: https://www.hrsa.gov/hansens-disease/index.html. In treating acute reactions in patients with a delayed diagnosis, physicians may seek prompt consultation with the National Hansen's Disease Program at 1-800-642-2477, weekdays 9 am to 5:30 pm ET.

Individuals living in the continental United States, Puerto Rico or the U.S. Territories may receive medical care for the diagnosis and treatment of Hansen's disease (leprosy)-related conditions at one of the 16 Federally-supported outpatient clinics in 10 States and Puerto Rico. https://www.hrsa.gov/hansens-disease/ambulatory-clinics.html

Hansen's disease medications can be provided to patients living in an area not served by an HD clinic through the National Hansen's Disease Program (NHDP). A private physician can order the HD medications (dapsone, rifampin, clofazamine) from the NHDP at no charge to the patient. The NHDP also provides consultant and biopsy processing services to the physician at no cost to the patient. Office visit and laboratory charges are not covered by the NHDP.

Contact Management

Household or other close family contacts (of untreated cases) are considered to be at risk of infection should have a thorough physical examination annually for five years. If questionable skin rash develops, they should notify their health care providers and have the skin rash biopsied to determine whether or not Hansen's disease is present.

Education

1) As needed the following resources can be givent to the patient:
   - KDHE Fact Sheet
   - HRSA Hansen's Disease Patient Information Sheet.

MANAGING SPECIAL SITUATIONS

A. Outbreak Investigation:

- Outbreak definition:
  - In outbreaks, cases are clustered in time and place among groups that share a common space.
  - For Hansen's disease, a single acute case is unusual, especially outside the states and territories of Texas, Louisiana, California, New York, Hawaii, Florida, and Puerto Rico. A complete investigation of risk factors and location of contacts is warranted.
- Notify KDHE immediately, 877-427-7317 of suspected outbreaks.
- Active case finding will be an important part of any investigation.
- Recommendations will be made based on the CDC guidance

DATA MANAGEMENT AND REPORTING TO THE KDHE

A. Accept the case assigned to the LHD and record the date the LHD investigation was started on the [Administrative] tab.

B. Organize and collect data, using appropriate data collection tools including:

- The Hansen's Disease (Leprosy) Surveillance Form can be used.
- Investigators can enter all required information directly into EpiTrax [Investigation], [Clinical], [Demographics], [Epidemiological] tabs.

C. Report data collected during the course of the investigation via EpiTrax**.**

- Verify that all data requested has been recorded on an appropriate EpiTrax [tab], or that actions are completed for a case lost to follow-up.
- Some data that cannot be reported on an EpiTrax [tab] may need to be recorded in [Notes] or scanned and attached to the record.
- Paper report forms do not need to be sent to KDHE after the information is recorded and/or attached in EpiTrax. The forms should be handled as directed by local administrative practices.

D. If a case is lost to follow-up, after the appropriate attempts to contact the case have been made:

- Indicate 'lost to follow-up' on the [Investigation] tab with the number of attempts to contact the case recorded.
- Record at least the information that was collected from the initial reporter.
- Record a reason for 'lost to follow-up' in [Notes].

E. After the steps listed under Case Investigation have been completed, record the "Date LHD investigation completed" field located on the [Administrative] tab.

- Record the date even if the local investigator's Case or Contact Management for the contact is not "Complete".

F. Once the entire investigation is completed, the LHD investigator will click the "Complete" button on the [Administrative] tab. This will trigger an alert to the LHD Administrator so they can review the case before sending to the state.

- The LHD Administrator will then "Approve" or "Reject" the CMR.
- Once a case is "Approved" by the LHD Administrator, BEPHI staff will review and close the case after ensuring it is complete and that the case is assigned to the correct event, based on the reported symptoms reported.

ADDITIONAL INFORMATION / REFERENCES

A. Treatment / Differential Diagnosis: Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015.

B. Epidemiology, Investigation and Control: Heymann. D., ed., Control of Communicable Diseases Manual (CCDM), 20th Edition. Washington, DC, American Public Health Association, 2015.

C. Case Definitions: CDC Division of Public Health Surveillance and Informatics, Available at: wwwn.cdc.gov/nndss/

D. Additional Information: https://www.hrsa.gov/hansens-disease/index.html

Instructions for Completing the Hansen's Disease *(Leprosy)* Surveillance Form

The Hansen's Disease or Leprosy Surveillance Form *(LSF)* is the document used to report leprosy cases to the U.S. National Hansen's Disease Registry. These data are used for epidemiological, clinical, and basic research studies throughout the National Hansen's Disease Program *(NHDP),* and are the official source for information on leprosy cases in the U.S.

The information requested on the LSF is used by many clinicians and researchers, and collection of all information is highly desirable. However, the fields that are **boldfaced** on the form and in the instructions below are considered to be the minimal information needed to register a patient. Failure to provide this information will result in the form being returned which creates additional work and may cause delays in obtaining program services for the patient.

1. **Reporting State:** Use the abbreviation of the state from which the report is being sent. This is usually the state of the clinician's office and not necessarily the patient's resident state.
2. **Date of Report:** This is date of the initial LSF completion. If patient was previously reported and has relapsed, write the word "RELAPSE" next to the date.
3. Social Security Number: self-explanatory.
4. **Patient Name:** Self-explanatory.
5. **Present Address:** Please include the county and zip code which are used to geographically cluster patients.
6. **Place of Birth:** Include state and county, if born in the U.S., or the country, if foreign born.
7. **Date of Birth/Sex:** Self-explanatory.
8. **Race/Ethnicity:** This information should be voluntarily provided by the patient. If the patient refuses or indicates a race/ethnicity category not listed, check the "Not Specified" box.
9. **Date Entered the U.S.:** For patients who have immigrated to the U.S., provide the month and year of entry.
10. Date of Onset of Symptoms: This information is usually the patient's recollection of when classic leprosy symptoms *(rash, nodule formation, paresthesia, decreased peripheral sensation, etc.)* were first noticed.
11. **Date Leprosy First Diagnosed:** Provide the month and year a diagnosis was made. This usually coincides with a biopsy date if one was performed.
12. **Initial Diagnosis:** Was the patient diagnosed in the U.S. or outside the U.S.
13. **Type of Leprosy:** Classify the diagnosis based on one of the ICD-9-CM diagnosis codes. (NHDP Clinic physicians: Please circle specific classification, if possible)

    030.0 Lepromatous Leprosy *(macular, diffuse, infiltrated, nodular, neuritic – includes Ridley-Jopling [RJ], Lepromatous [LL] and Borderline lepromatous [BL]*): A form marked by erythematous macules, generalized papular and nodular lesions, and variously by upper respiratory infiltration, nodules on conjunctiva or sclera, and motor loss.
    030.1 Tuberculoid Leprosy *(macular, maculoanesthetic, major, minor, neuritic – includes RJTuberculoid [TT] and Borderline tuberculoid [BT]*): A form marked by usually one lesion with well-defined margins with scaly surface and local tender cutaneous or peripheral nerves.
    030.2 Indeterminate *(uncharacteristic, macular, neuritic)*: A form marked by one or more macular lesions, which may have slight erythema.
    030.3 Borderline *(dimorphous, infiltrated, neuritic – includes RJ Borderline [BB] or true mid disease only)*: A form marked by early nerve involvement and lesions of varying stages.
    030.8 Other Specified Leprosy: Use this code when the diagnosis is specified as a "leprosy" but is not listed above *(030.0-030.3)*.
    030.9 Leprosy, Inactive: Use this code when the diagnosis is identified as a "leprosy" but inactive.
14. Diagnosis of Disease: Enter INITIAL biopsy and skin smear dates and results.
15. **Residence *(Pre-diagnosis)*:** List all cities, counties, and states in the U.S. and all foreign countries a patient resided in BEFORE leprosy was diagnosed. This information is used to map all places where U.S. leprosy cases have resided.
16. Disability: Indicate any sensory abnormalities or deformities of the hands and feet or lagophthalmos of the eyes.
17. Current Household Contacts: Self-explanatory.
18. Current Treatment for Leprosy: Date treatment started and indicate all drugs used for initial treatment.
19. **Name and Address of Physician or Investigator:** Self-explanatory.

HANSEN'S DISEASE *(LEPROSY)* SURVEILLANCE FORM

NATIONAL HANSEN'S DISEASE PROGRAMS
1770 PHYSICIANS PARK DRIVE
BATON ROUGE, LA 70816
1-800-642-2477

1 Reporting State ☐☐

2 Date of Report Mo. ☐☐ Day ☐☐ Yr. ☐☐

3 Social Security Number *(optional)* _____ - _____ - _____

4 Patient Name: *(Last)* *(First)* *(Middle)*

5 Present Address: Street _____ City _____
County _____ State _____ Zip _____

6 Place of Birth:
State _____ County _____
Country _____

7 Date of Birth: Mo. ☐☐ Day ☐☐ Yr. ☐☐

Sex: ☐ Male ☐ Female

8 Race/Ethnicity:
☐ White, Not Hispanic ☐ White, Hispanic ☐ American Indian, Alaska Native ☐ Indian, Middle Easterner
☐ Black, Not Hispanic ☐ Black, Hispanic ☐ Asian, Pacific Islander ☐ Not Specified

9 Date Entered U.S.: Mo. ☐☐ Yr. ☐☐

10 Date of Onset of Symptoms: Mo. ☐☐ Yr. ☐☐

11 Date Leprosy First Diagnosed: Mo. ☐☐ Yr. ☐☐

12 Initial Diagnosis in: ☐ U.S. ☐ Outside U.S.

13 Type of Leprosy: ***(ICD-9-CM Code)*** **(NHDP Clinic physicians: Please circle specific classification, if possible)**
☐ Lepromatous *(030.0 - LL, BL)* ☐ Indeterminate *(030.2 - IN)* ☐ Other Specified Leprosy *(030.8)*
☐ Tuberculoid *(030.1 - TT, BT)* ☐ Borderline *(030.3 - BB)* ☐ Leprosy, Unspecified *(030.9)*

14 Diagnosis of Disease:
Was biopsy performed in U.S. ☐ Yes ☐ No
Date ____ / ____ / ____
Result _____

Skin Smear ☐ Yes ☐ No
Date ____ / ____ / ____

BI: Positive ____ Negative ____

15 List all places in the U.S.A. and all foreign countries a PATIENT resided *(Including Military Service)* BEFORE leprosy was diagnosed.

TOWN	COUNTY	STATE	COUNTRY	INCLUSIVE DATES From Mo./Yr.	INCLUSIVE DATES To Mo./Yr.

16 Disability:

	Hands Yes	Hands No	Feet Yes	Feet No	Eye
Sensory Loss	☐	☐	☐	☐	Lagophthalmos?
Deformity	☐	☐	☐	☐	Yes ☐ No ☐

17 Current Household Contacts:

Name/Relationship

1 _____
2 _____
3 _____
4 _____
5 _____

18 Current Treatment for Leprosy: ***(check all that apply)***

Date Treatment Started: ____ / ____ Mo. Yr.

☐ Dapsone
☐ Rifampin
☐ Clofazimine
☐ Other *(list)* _____

19 Name and Address of Physician: _____

Investigator:

This form may be FAXED to NHDP at (225) 756-3706 **DHNDP - 5/2010**
A copy of this form should be sent to your local or state health department.