Hansen’s disease  
(Leprosy)  
Investigation Guideline

CONTENT:  
VERSION DATE:

Investigation Protocol:

• Investigation Guideline 01/2013
• Hansen’s Disease Surveillance Form 05/2010

Supporting Materials found in attachments:

• Fact Sheet 01/2013

Revision History:

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<td>01/2013</td>
<td>07/2010</td>
<td>New case definition; added notification section; updated web links. Updated fact sheet.</td>
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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 case 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:
- Immigrant from country with high incidence of leprosy
- U.S. resident with history of foreign travel
- Resident of Texas or Louisiana
- 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](http://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 (hyperesthesia, 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](http://www.hrsa.gov/hansensdisease/diagnosis/recommendedtreatment.html).
NOTIFICATION TO PUBLIC HEALTH AUTHORITIES

Hansen’s disease infections shall be designated as infectious or contagious in their nature, and cases or suspect cases shall be reported within seven days:

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)
4. KDHE-BEPHI will contact the local public health jurisdiction by phone within one hour of receiving any suspected Hansen’s disease report.

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 cases require a STANDARD report 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, completing the forms within 7 days of receiving a notification of a report.
- 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) Use current case definition, to confirm diagnosis with the medical provider.
2) Conduct case investigation to collect information on potential sources.
3) Conduct an initial contact investigation of close contacts.
4) Complete and report all information requested in EpiTrax.
5) As appropriate, use the notification letter(s) and the disease fact sheet.

STANDARD CASE INVESTIGATION AND CONTROL METHODS

Case Investigation

1) Contact the medical provider who reported or ordered testing of the case.

   Note: If the physician, submitted samples to NHDP, the Hansen’s disease Surveillance Form may already be completed or started – try to obtain a copy.

   - Obtain information from the provider or medical chart.
   - If patient hospitalized, obtain medical records, including admission notes, progress notes, lab report(s), and discharge summary.
   - Identify onset date of symptoms and type of leprosy that was diagnosed.
   - Identify the diagnosis date of disease.
   - Examine laboratory testing: record biopsy and skin smear results, dates.
   - Record current treatment.
• Record hospitalizations: location, dates
• Record outcomes: disabilities, survived or date of death
• Collect case’s demographic data and contact information (birth date, county, sex, race/ethnicity, occupation, address, phone number(s))

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

3) Investigate epi-links among cases (clusters, household, co-workers, etc).
• 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

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: www.hrsa.gov/hansensdisease. 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.

Those with Hansen's disease in the U.S. can receive needed medications at no cost through their own doctor or through a NHPD Ambulatory Care Clinic. Information on ambulatory clinics is on-line at www.hrsa.gov/hansensdisease/ambulatoryclinics.html.
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.

MANAGING SPECIAL SITUATIONS

A. Outbreak Investigation:
   1) 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.
   2) Notify KDHE immediately, 877-427-7317 of suspected outbreaks.
   3) Active case finding will be an important part of any investigation.
   4) Recommendations will be made based on the CDC guidance.

DATA MANAGEMENT AND REPORTING TO THE KDHE

Note: When the NHDP becomes aware of a new case, a surveillance form is sent to the provider to obtain the data needed to register the patient. If the information has already been reported there may be no need for the local health department to collect the information.

A. Organize and collect data.
B. Report data via EpiTrax
   • Especially data that collected during the investigation that helps to confirm or classify a case.

ADDITIONAL INFORMATION / REFERENCES

C. Case Definitions: CDC Division of Public Health Surveillance and Informatics, Available at: www.cdc.gov/nndss/
D. Kansas Regulations/Statutes Related to Infectious Disease: www.kdheks.gov/epi/regulations.htm
E. Additional Information (CDC): www.cdc.gov/health/default.htm
Supporting Materials are available under attachments:

CLICK HERE TO VIEW ATTACHMENTS

Then double click on the document to open.

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