07-ID-11

Committee: Infectious Diseases

Title: Revised National Surveillance Case Definition for Lyme disease

Statement of the problem:

Lyme disease is the most commonly reported vector-borne disease in the United States. In 1990, CSTE adopted the position statement that added Lyme disease to the list of nationally notifiable diseases (http://www.cste.org/ps/1990/1990-01.htm). The Lyme disease national surveillance case definition was last updated in 1996 (http://www.cste.org/ps/1996/1996-18.htm) (ftp://ftp.cdc.gov/pub/Publications/mmwr/rr/rr4610.pdf).

The purpose of this position statement is to revise the Lyme disease national surveillance case definition in accordance with the 2007 CSTE position statement template for placing diseases or conditions under national surveillance (http://www.cste.org/PositionStatement.asp). This revision permits states and territories to report confirmed and probable cases of Lyme disease to the National Notifiable Diseases Surveillance System (NNDSS), updates the laboratory evidence section to reflect current testing practices, and provides measures to assess the public health surveillance burden.

Statement of the desired action(s) to be taken:

Revise the national surveillance case definition for Lyme disease.

Goals of surveillance:

(1) Define the demographic, geographic, and seasonal distribution; (2) consistently monitor disease trends; (3) identify risk factors for transmission in areas where Lyme disease is newly emerging; and (4) evaluate the impact of prevention and control measures.

Methods for surveillance:

Case finding is conducted through standard clinician and laboratory reporting to local or state health agencies. Core surveillance data will be reported by state health agencies to the CDC NNDSS through the National Electronic Telecommunications System for Surveillance (NETSS) or the National Electronic Disease Surveillance System (NEDSS), as per state protocol.

Case definition:

This surveillance case definition was developed for national reporting of Lyme disease; it is not intended to be used in clinical diagnosis.

Clinical presentation:
A systemic, tickborne disease with protean manifestations, including dermatologic, rheumatologic, neurologic, and cardiac abnormalities. The best clinical marker for the disease is erythema migrans (EM), the initial skin lesion that occurs in 60%-80% of patients.

For purposes of surveillance, EM is defined as a skin lesion that typically begins as a red macule or papule and expands over a period of days to weeks to form a large round lesion, often with partial central clearing. A single primary lesion must reach greater than or equal to 5 cm in size across its largest diameter. Secondary lesions also may occur. Annular erythematous lesions occurring within several hours of a tick bite represent hypersensitivity reactions and do not qualify as EM. For most patients, the expanding EM lesion is accompanied by other acute symptoms, particularly fatigue, fever, headache, mildly stiff neck, arthralgia, or myalgia. These symptoms are

typically intermittent. The diagnosis of EM must be made by a physician. Laboratory confirmation is recommended for persons with no known exposure.

For purposes of surveillance, late manifestations include any of the following when an alternate explanation is not found:

- Musculoskeletal system. Recurrent, brief attacks (weeks or months) of objective joint swelling in one or a few joints, sometimes followed by chronic arthritis in one or a few joints. Manifestations not considered as criteria for diagnosis include chronic progressive arthritis not preceded by brief attacks and chronic symmetrical polyarthritis. Additionally, arthralgia, myalgia, or fibromyalgia syndromes alone are not criteria for musculoskeletal involvement.
- Nervous system. Any of the following, alone or in combination: lymphocytic meningitis; cranial neuritis, particularly facial palsy (may be bilateral); radiculoneuropathy; or, rarely, encephalomyelitis. Encephalomyelitis must be confirmed by demonstration of antibody production against *Borrelia burgdorferi* in the cerebrospinal fluid (CSF), evidenced by a higher titer of antibody in CSF than in serum. Headache, fatigue, paresthesia, or mildly stiff neck alone are not criteria for neurologic involvement.
- Cardiovascular system. Acute onset of high-grade (2nd-degree or 3rd-degree) atrioventricular conduction defects that resolve in days to weeks and are sometimes associated with myocarditis. Palpitations, bradycardia, bundle branch block, or myocarditis alone are not criteria for cardiovascular involvement.

Laboratory evidence

For the purposes of surveillance, the definition of a qualified laboratory assay is (1) a positive culture for *B. burgdorferi*, (2) two-tier testing interpreted using established criteria [1], or (3) single-tier IgG immunoblot seropositivity interpreted using established criteria [1-4].

Exposure

Exposure is defined as having been (less than or equal to 30 days before onset of EM) in wooded, brushy, or grassy areas (i.e., potential tick habitats) in a county in which Lyme disease is endemic. A history of tick bite is not required.

Disease endemic to county

A county in which Lyme disease is endemic is one in which at least two confirmed cases have been acquired in the county or in which established populations of a known tick vector are infected with *B. burgdorferi*.

Suggested codes for case ascertainment

To be developed (e.g. LOINC codes for finding suspected cases through automated review of laboratory electronic data).

Detailed definitions for case classification

Confirmed: a) a case of EM with a known exposure (as defined above), or b) a case of EM with laboratory evidence of infection (as defined above) and without a known exposure or c) a case with at least one late manifestation that has laboratory evidence of infection.

Probable: any other case of physician-diagnosed Lyme disease that has laboratory evidence of infection (as defined above).

Suspected: a) a case of EM where there is no known exposure (as defined above) and no laboratory evidence of infection (as defined above), or b) a case with laboratory evidence of infection but no clinical information available (e.g. a laboratory report).

Lyme disease reports will not be considered cases if the medical provider specifically states this is not a case of Lyme disease, or the only symptom listed is "tick bite" or "insect bite."

Period of Surveillance:

Ongoing. This revision of the surveillance case definition will be effective January 1, 2008.

Data sharing/release and print criteria:

States and territories will send CDC case data for all confirmed and probable cases. CDC will release only fully de-identified case data to the general public. Other releases require signed data sharing agreements using a format pre-approved by the state/territorial health agency.

Final printed counts published in the weekly MMWR by CDC will distinguish between confirmed and probable cases. Publication criteria will exclude suspected cases from final printed counts published by CDC. Provisional case report data will not be used until verification procedures are complete.

Background and Justification:

Since 1991, the number of reported Lyme disease cases has nearly doubled increasing to over 20,000 reported cases in 2005 (5). The increase in reported cases is likely the result of multiple factors, including a true increase in disease incidence and surveillance artifact due to the addition of laboratory surveillance methods. The increase in the number of reported cases has created a burden on both the local and state health departments due to the overall volume of cases reported and the follow up of clinical manifestations required for laboratory reported cases to determine case confirmation status.

To address the surveillance burden and create more sustainable Lyme disease surveillance systems, some states have modified the components of their systems (e.g. by dropping laboratory reporting) or modified the number of laboratory cases that are followed up (e.g. sampling a proportion of reported cases for follow up to limit the burden but to still generate an estimate of overall confirmed cases). While these strategies have somewhat decreased the burden of Lyme disease surveillance for the health departments, these changes have also contributed to a decrease in the number of reported cases nationally. There is concern that the reduction in reported cases will give the erroneous impression that the incidence of Lyme disease is decreasing.

The process that begins with the receipt of a positive laboratory report by a public health department and ends with the determination of surveillance case status is often very labor intensive. The revised surveillance case definition permits local and state health departments the flexibility to classify Lyme disease reports as confirmed, probable, or suspect cases and permits the reporting of confirmed and probable case totals at the national level. The revised surveillance case definition alone will not decrease the Lyme disease surveillance burden, but will give public health officials a more complete measure of the surveillance burden that can be used to guide the allocation of scarce public health surveillance resources.

References:

1. Centers for Disease Control and Prevention. Recommendations for test performance and interpretation from the Second National Conference on Serologic Diagnosis of Lyme Disease. MMWR MMWR Morb Mortal Wkly Rep 1995; 44:590–1.
2. Dressler F, Whalen JA, Reinhardt BN, Steere AC. Western blotting in the serodiagnosis of Lyme disease. J Infect Dis 1993; 167:392–400.
3. Engstrom SM, Shoop E, Johnson RC. Immunoblot interpretation criteria for serodiagnosis of early Lyme disease. J Clin Microbiol 1995; 33:419–27.

4. Centers for Disease Control and Prevention. Notice to readers: caution regarding testing for Lyme disease. MMWR Morb Mortal Wkly Rep 2005; 54:125–6.
5. Centers for Disease Control and Prevention. Lyme Disease — United States, 2003–2005. MMWR Morb Mortal Wkly Rep 2007; 56:573–6.

Coordination:

Agencies for Response:
Julie L. Gerberding, MD
Director
Centers for Disease Control and Prevention
1600 Clifton Road
Atlanta, GA 30333
404-639-7000
jyg2@cdc.gov

Agencies for Information:
(1) Scott J. Becker, MS
Executive Director
Association of Public Health Laboratories
8515 Georgia Avenue, Suite 700
Silver Spring, MD 20910
Telephone: 240-485-2745 ext 201
Fax: 240-485-2700:
sbecker@aphl.org

(2) Paul E. Jarris, MD, MBA
Executive Director
Association of State and Territorial Health Officials
2231 Crystal Drive Suite 450
Arlington, VA 22202
Phone: (202) 371-9090
Fax: (571) 527-3189
pjarris@astho.org

Submitting Author:
Matthew L. Cartter, MD, MPH
Epidemiology Program Coordinator
Connecticut Department of Public Health
410 Capitol Ave.
MS# 11EPI
P.O. Box 340308
Hartford, CT 06134-0308
Phone: (860) 509-7994
Fax: (860) 509-7910
matt.cartter@ct.gov