I. Statement of the Problem
According to the 2008 and 2011 national surveillance case definitions for Lyme disease, a physician-diagnosed erythema migrans rash in a person with exposure to potential tick habitats in a Lyme-endemic county is sufficient evidence for a confirmed case. The case definition defines endemic as a county in which at least two confirmed Lyme disease cases have been acquired or in which populations of a known tick vector are infected with *Borrelia burgdorferi*, the Lyme disease spirochete. It does not include a time period for these detections. In non-endemic counties, the patient must have clinically compatible symptoms and laboratory evidence of Lyme infection in order to meet the confirmed case definition.

In areas of the country where Lyme disease is absent, or present but rare, cases are more likely to get an erroneous diagnosis and be misclassified for surveillance purposes. The erythema migrans rash can be difficult to identify correctly for the untrained eye and the positive predictive value of the serologic tests used for Lyme disease is lower than in high incidence states.

In 2014, as in previous years, 14 states accounted for more than 96% of confirmed US Lyme disease cases. While the average annual U.S. incidence between 2005 and 2014 (CDC website) has ranged from 7.0/100,000 (in 2012) to 9.8/100,000 (in 2009), nearly all of the high incidence states have reported Lyme disease incidence that is orders of magnitude higher than the U.S. average for every year on record.

We propose to change the exposure criteria for the Lyme disease case definition to consider whether the exposure occurred in a state that consistently reports a high incidence of Lyme disease vs. in a state where Lyme disease is less frequently reported. In states with a high incidence rate of Lyme disease, an erythema migrans rash and exposure to potential tick habitats will be sufficient to confirm Lyme disease; in states where Lyme disease is less frequently reported, case confirmation will require both clinical and laboratory evidence of infection. In this position statement, we designate a state as high or low incidence based on a cutoff of 10 reported Lyme disease cases per 100,000 persons. This cutoff was chosen because it is a straightforward integer that has clearly and conservatively delineated, for greater than a decade, states having a high burden and high incidence of Lyme disease from states having low or no incidence.

We also propose that final Lyme disease case numbers be included in the MMWR annual surveillance reports and omitted from the weekly MMWR surveillance tables. Lyme disease cases are complex to classify and reporting of reliable case numbers is often delayed, making weekly case numbers of limited utility when comparing week to week or to that week in previous years. A large proportion of case numbers reported in the weekly MMWR tables are deleted after case review so these numbers do not accurately reflect Lyme disease trends and are inconsistent with final data.

II. Background and Justification
In the United States, Lyme disease is a tick-borne disease caused primarily by infection with *Borrelia burgdorferi sensu stricto*. More recently, a novel *B. burgdorferi sensu lato* genospecies (*candidatus B. mayonii*) has also been shown to cause Lyme disease among persons having a history of tick bite in the upper Midwestern US. However, more information is needed to define the burden of disease due to this new genospecies and its clinical range of illness (Pritt et al., 2016).

Historically, the majority (60–80%) of reported Lyme disease cases present with a characteristic rash, *erythema migrans*, typically accompanied by fever, headache and fatigue. Untreated Lyme disease can
progress to chronic arthritis, neurological disease and cardiac disease. Lyme disease cases have been identified in most states, but the highest risk areas are in the Northeastern and upper Midwestern United States. Ongoing surveillance is needed to monitor the demographic, geographic, and temporal patterns of disease, identify risk factors for transmission and evaluate prevention and control strategies. CSTE adopted a position statement in 1990 adding Lyme disease to the list of nationally notifiable diseases. The case definition was modified in 1996 and again in 2007.

Lyme disease can be challenging to diagnose. Most clinical symptoms are non-specific, and laboratory testing of patients living in areas where Lyme disease is rare has a low positive predictive value (PPV). (Tugwell et al., 1997; Lantos et al., 2015). Human disease and entomologic surveillance efforts have detected low levels of *B burgdorferi* transmission in the southeastern United States and other “non-endemic” states (Forrester et al. 2015). According to the 2008 Lyme disease case definition, such states have an increasing number of “endemic counties” scattered among “non-endemic” counties. As a result, different case definitions are used across small geographic regions and data become difficult to compare. However, in states (or parts of states) where Lyme disease is rare, the detection of *B burgdorferi* or the diagnosis of two human cases will not necessarily improve the PPV of other Lyme disease tests done. Nor are health care providers in these communities necessarily more experienced in recognizing the erythema migrans rash. The proposed change to the case definition would require laboratory confirmation of Lyme disease in states where infections are rare. We expect that as a result, the specificity of Lyme disease case data will improve and that disease statistics will be easier to interpret both within and across states.

III. Statement of the desired action(s) to be taken

1. Utilize standard sources (e.g. reporting*) for case ascertainment for Lyme disease. Surveillance for Lyme disease should use the following recommended sources of data to the extent of coverage presented in Table III.

<table>
<thead>
<tr>
<th>Source of data for case ascertainment</th>
<th>Coverage</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Population-wide</td>
</tr>
<tr>
<td>Clinician reporting</td>
<td>X</td>
</tr>
<tr>
<td>Laboratory reporting</td>
<td>X</td>
</tr>
<tr>
<td>Reporting by other entities (e.g., hospitals, veterinarians, pharmacies, poison centers)</td>
<td>X</td>
</tr>
<tr>
<td>Death certificates</td>
<td>X</td>
</tr>
<tr>
<td>Hospital discharge or outpatient records</td>
<td>X</td>
</tr>
<tr>
<td>Extracts from electronic medical records</td>
<td>X</td>
</tr>
<tr>
<td>Telephone survey</td>
<td></td>
</tr>
<tr>
<td>School-based survey</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td></td>
</tr>
</tbody>
</table>

2. Utilize standardized criteria for case identification and classification (Sections VI and VII) for Lyme disease and add Lyme disease to the *Nationally Notifiable Condition List.*

- ☑ 2a. Immediately notifiable, extremely urgent (within 4 hours)
- ☑ 2b. Immediately notifiable, urgent (within 24 hours)
- ☑ 2c. Routinely notifiable

CSTE recommends that all States and Territories enact laws (statute or rule/regulation as appropriate) to make this disease or condition reportable in their jurisdiction. Jurisdictions (e.g. States and Territories) conducting surveillance (according to these methods) should submit case notifications** to CDC.
Expectations for Message Mapping Guide (MMG) development for a newly notifiable condition: NNDSS is transitioning to HL7-based messages for case notifications; the specifications for these messages are presented in MMGs. When CSTE recommends that a new condition be made nationally notifiable, CDC must obtain OMB PRA approval prior to accepting case notifications for the new condition. Under anticipated timelines, notification using the Generic V2 MMG would support transmission of the basic demographic and epidemiologic information common to all cases and could begin with the new MMWR year following the CSTE annual conference. Input from CDC programs and CSTE would prioritize development of a disease-specific MMG for the new condition among other conditions waiting for MMGs.

3. CDC should publish data on Lyme disease as appropriate in MMWR and other venues (see Section IX).

CSTE recommends that all jurisdictions (e.g. States or Territories) with legal authority to conduct public health surveillance follow the recommended methods as outlined above.

IV. Goals of Surveillance
To provide information on the temporal, geographic, and demographic occurrence of Lyme disease to facilitate its prevention and control.

V. Methods for Surveillance: Surveillance for Lyme disease should use the recommended sources of data and the extent of coverage listed in Table III.

Surveillance for Lyme disease should use the recommended sources of data and the extent of coverage listed in Table III.

VI. Criteria for case identification
A. Narrative: A description of suggested criteria for case ascertainment of a specific condition.

Report any illness to public health authorities that meets any of the following criteria:

1. Any person with erythema migrans.

2. Any person with laboratory evidence of Lyme disease. Laboratory evidence includes any of the following:
   a. A positive culture for *B. burgdorferi*
   b. Antibody to *B. burgdorferi* detected in serum by EIA or IFA
   c. A Western immunoblot test positive for *B. burgdorferi*-specific IgM or IgG
   d. Antibody to *B. burgdorferi* detected in CSF by EIA or IFA

3. A person whose healthcare record contains a diagnosis of Lyme disease.

4. A person whose death certificate lists Lyme disease as a cause of death or a significant condition contributing to death.

*Other recommended reporting procedures*
• All cases of Lyme disease should be reported.
• Reporting should be on-going and routine.
• Frequency of reporting should follow the state health department’s routine schedule.

B. Table of criteria to determine whether a case should be reported to public health authorities

Table VI-B. Table of criteria to determine whether a case should be reported to public health authorities.

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Reporting Disease or Condition Subtype</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical Evidence</td>
<td></td>
</tr>
<tr>
<td>Erythema migrans</td>
<td>S</td>
</tr>
<tr>
<td>Healthcare record contains a diagnosis of Lyme disease</td>
<td>S</td>
</tr>
<tr>
<td>Death certificate lists Lyme disease as a cause of death or a significant condition contributing to death</td>
<td>S</td>
</tr>
<tr>
<td>Laboratory Evidence</td>
<td></td>
</tr>
<tr>
<td>Culture positive for <em>B. burgdorferi</em></td>
<td>S</td>
</tr>
<tr>
<td>Antibody positive for <em>B. burgdorferi</em> by EIA or IFA in serum or CSF</td>
<td>S</td>
</tr>
<tr>
<td>Western immunoblot positive for <em>B. burgdorferi</em>-specific IgM</td>
<td>S</td>
</tr>
<tr>
<td>Western immunoblot positive for <em>B. burgdorferi</em>-specific IgG</td>
<td>S</td>
</tr>
</tbody>
</table>

Notes:
S = This criterion alone is Sufficient to report a case.
N = All “N” criteria in the same column are Necessary to report a case.
O = At least one of these “O” (One or more) criteria in each category (e.g., clinical evidence and laboratory evidence) in the same column—in conjunction with all “N” criteria in the same column—is required to report a case.
* A requisition or order for any of the “S” laboratory tests is sufficient to meet the reporting criteria.

C. Disease-specific data elements

VII. Case Definition for Case Classification

A. Narrative: Description of criteria to determine how a case should be classified.

Clinical Criteria

Clinical presentation

A systemic, tick-borne disease with protean manifestations, including dermatologic, rheumatologic, neurologic, and cardiac abnormalities. The most common 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 signs that cannot be explained by any other etiology, alone or in combination: lymphocytic meningitis; cranial neuritis, particularly facial palsy (may be bilateral); radiculoneuropathy; or, rarely, encephalomyelitis. 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 Criteria**

For the purposes of surveillance, laboratory evidence includes:

1. A positive culture for *B. burgdorferi*, or
2. A positive two-tier test. (This is defined as a positive or equivocal enzyme immunoassay (EIA) or immunofluorescent assay (IFA) followed by a positive IgM\(^1\) or IgG\(^2\) western immunoblot (WB) for Lyme disease) or
3. A positive single-tier IgG\(^2\) WB test for Lyme disease\(^3\).

\(^1\) IgM WB is considered positive when at least two of the following three bands are present: 24 kDa (OspC)*, 39 kDa (BmpA), and 41 kDa (Fla). Disregard IgM results for specimens collected >30 days after symptom onset.

\(^2\) IgG WB is considered positive when at least five of the following 10 bands are present: 18 kDa, 24 kDa (OspC)*, 28 kDa, 30 kDa, 39 kDa (BmpA), 41 kDa (Fla), 45 kDa, 58 kDa (not GroEL), 66 kDa, and 93 kDa.

\(^3\)While a single IgG WB is adequate for surveillance purposes, a two-tier test is still recommended for patient diagnosis.

*Depending upon the assay, OspC could be indicated by a band of 21, 22, 23, 24 or 25 kDa.

**Epidemiologic Criteria**

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) of Lyme disease vectors. Since infected ticks are not uniformly distributed, a detailed travel history to verify whether exposure occurred in a high or low incidence state is needed. An exposure in a high-incidence state is defined as exposure in a state with an average Lyme disease incidence of at least 10 confirmed cases/100,000 for the previous three reporting years. A low-incidence state is defined as a state with a disease incidence of <10 confirmed cases/100,000. (http://www.cdc.gov/lyme/stats/tables.html) A history of tick bite is not required.
Case Classification

**Confirmed:**
- A case of EM with exposure in a high incidence state (as defined above), OR
- A case of EM with laboratory evidence of infection and a known exposure in a low incidence state, OR
- Any 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 case of EM where there is no known exposure (as defined above) and no laboratory evidence of infection (as defined above), or
- A case with 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."

**Criteria to distinguish a new case of this disease or condition from reports or notifications which should not be enumerated as a new case for surveillance**
Case not previously reported to public health authorities.
### B. Classification Tables

#### Table VII-B. Criteria for defining a case of Lyme disease.

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Suspect</th>
<th>Probable</th>
<th>Confirmed</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Clinical Evidence</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Physician diagnosed Lyme disease</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>Medical provider does not specifically state this is not a case of Lyme disease</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>Erythema migrans</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>Arthritis with objective joint swelling</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td>Chronic arthritis</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td>Aseptic meningitis</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td>Cranial neuritis</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td>Facial palsy</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td>Radiculoneuropathy</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td>Encephalomyelitis</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td>AV conduction defects</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td>Myocarditis</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td>No Clinical Information is available</td>
<td>N</td>
<td>N</td>
<td></td>
</tr>
<tr>
<td><strong>Laboratory evidence</strong></td>
<td>S</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Culture positive for <em>B. burgdorferi</em></td>
<td>S</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Serum antibody positive for <em>B. burgdorferi</em> by EIA or IFA</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>Western immunoblot positive for <em>B. burgdorferi</em>-specific IgM (onset ≤30 days)</td>
<td>O</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>Western immunoblot positive for <em>B. burgdorferi</em>-specific IgG</td>
<td>O</td>
<td>S</td>
<td>O</td>
</tr>
<tr>
<td><strong>Epidemiologic evidence</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Possible exposure to infected tick vector</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>High Incidence State</td>
<td>N</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Criteria to distinguish a new case:</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Case not previously reported to public health authorities</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
</tbody>
</table>

**Notes:**
- S = This criterion alone is Sufficient to classify a case.
- N = All “N” criteria in the same column are Necessary to classify a case. A number following an “N” indicates that this criterion is only required for a specific disease/condition subtype (see below). If the absence of a criterion (i.e., criterion NOT present) is required for the case to meet the classification criteria, list the Absence of criterion as a Necessary component.
- O = At least one of these “O” (One or more) criteria in each category (e.g., clinical evidence and laboratory evidence) in the same column—in conjunction with all “N” criteria in the same column—is required to classify a case. (These “O” criteria are alternatives, which means that a single column will have either no O criteria or multiple O criteria; no column should have only one O.) A number following an “O” indicates that this criterion is only required for a specific disease/condition subtype.

### VIII. Period of Surveillance

Surveillance should be on-going
IX. Data sharing/release and print criteria

- Notification to CDC of confirmed and probable cases is recommended for cases of Lyme disease.
- Finalized data are published annually in the Summary of Notifiable Diseases. Longer articles describing and interpreting national trends are published in the MMWR on an ad-hoc basis (approximately once every other year). Summary data are also made available through the CDC Lyme disease website.
- State-specific compiled data will continue to be published in the annual MMWR.
- No specific plans for re-release. However, CDC may re-release finalized data on ad hoc basis for research or public health activities in accordance with the Data Release Guidelines for the National Notifiable Diseases Surveillance System.

X. Revision History

<table>
<thead>
<tr>
<th>Position Statement ID</th>
<th>Section of Document</th>
<th>Revision Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>10-ID-06</td>
<td>Transfer to new position statement format</td>
<td>Added tables needed for electronic disease reporting.</td>
</tr>
<tr>
<td>07-ID-11</td>
<td>Updated case definition</td>
<td>Updated laboratory testing criteria and added endemicity definition</td>
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<tr>
<td>1990</td>
<td></td>
<td>ADDED Lyme disease to the NNC list</td>
</tr>
</tbody>
</table>

XI. References


XII. Coordination

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