

Appendix B: Provincial Case Definitions for Reportable Diseases

Disease: Lyme Disease

Lyme Disease

1.0 Provincial Reporting

Confirmed and probable cases of disease

2.0 Type of Surveillance

Case-by-case

3.0 Case Classification

3.1 Confirmed case

- Erythema migrans (EM)¹ with laboratory confirmation by polymerase chain reaction (PCR)² or culture³
OR
- EM with laboratory support by serological methods², and a history of residence in, or visit to, an endemic area⁴
OR
- Objective symptoms of disseminated Lyme disease⁵ with laboratory confirmation by PCR or culture
OR
- Objective symptoms of disseminated Lyme disease with laboratory support by serological methods, and a history of residence in, or visit to, an endemic area

3.2 Probable case

- EM with laboratory support by serological methods but with no history of residence in, or visit to, an endemic area
OR
- Objective symptoms of disseminated Lyme disease with laboratory support by serological methods, but with no history of residence in, or visit to an endemic area
OR
- EM without laboratory confirmation, but with history of residence in, or visit to, an endemic area

4.0 Laboratory Evidence

4.1 Laboratory Confirmation

Any of the following will constitute a confirmed case of Lyme disease:

- Isolation of *B. burgdorferi* from an appropriate clinical specimen
- Positive nucleic acid amplification test (NAT) for *B. burgdorferi*
- Serological evidence using the two-tier enzyme-linked immuno-sorbent assay (ELISA) and Western Blot criteria
(Serological evidence alone is not confirmatory: positive predictive value is greater provided that the patient has EM or objective symptoms of disseminated Lyme disease, and has had contact with a region endemic for Lyme disease.)

4.2 Approved/Validated Tests

- Standard culture for *B. burgdorferi*

- Commercial *B. burgdorferi* Immunoglobulin M (IgM) and Immunoglobulin G (IgG) tests (ELISA and Western Blot)
- NAT for *B. burgdorferi*

4.3 Indications and Limitations

- Only serum samples are acceptable for serology
- Initial negative serological tests in patients with skin lesions suggestive of EM should have testing repeated after four weeks
- Sera that are screened negative for antibodies using an EIA should not be subjected to Western blot testing
- EIA tests presently in use lack the specificity necessary to base a diagnosis of Lyme disease on an unconfirmed result
- The possibility of false-positive Western blot results should not be ignored
- When patients are treated very early in the course of illness, antibodies may not develop

5.0 Clinical Evidence

- A systemic, tick-borne 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. Secondary lesions may also occur.
- 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 exposure⁶
- For purposes of surveillance, late manifestations include any of the following when an alternate explanation is not found:
 - Nervous system: Any of the following, 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.
 - 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.
 - 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.

6.0 ICD Code(s)

ICD 10 Code A69.2

7.0 Comments

¹ Erythema migrans is a pathognomonic sign of Lyme disease. It 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 round or oval expanding erythematous area. Some lesions are homogeneously erythematous, whereas others have prominent central clearing or a distinctive target-like appearance. A single primary lesion must reach ≥ 5 cm in size across its largest diameter. On the lower extremities, the lesion may be partially purpuric. EM represents a response to the bacterium as it spreads intradermally from the site of the infecting tick bite. It appears 1-2 weeks (range 3-30 days) after infection and persists for up to 8 weeks, by which time the bacterium leaves the skin and disseminates haematogenously. An erythematous skin lesion that presents while a tick vector is still attached or which has developed within 48 hours of detachment is most likely a tick bite hypersensitivity reaction (i.e., a non-infectious process), rather than erythema migrans. Tick bite hypersensitivity reactions are usually < 5 cm in largest diameter, sometimes have an urticarial appearance, and typically begin to disappear within 24–48 hours. Signs of acute or chronic inflammation are not prominent. There is usually little pain, itching, swelling, scaling, exudation or crusting, erosion or ulceration, except that some inflammation associated with the tick bite itself may be present at the very centre of the lesion.

² PCR and serological methods on cerebrospinal fluid (CSF) are investigational only. The role of PCR (or more appropriately NAT) testing should be limited to CSF or tissue samples as there is limited data to support its use on blood and/or urine samples.

³ Culturing for *B. burgdorferi* is a low-yield procedure and is not encouraged; if performed, it should be done only on biopsies from EM lesions and synovial or spinal fluid.

⁴ An endemic area is defined here as a census subdivision in which a reproducing population of *Ixodes scapularis* or *Ixodes pacificus* tick vectors is known to occur, which has been demonstrated by molecular methods to support transmission of *B. burgdorferi* at that site.

⁵ Symptoms of disseminated Lyme disease are those objective symptoms as described in the 2006 clinical practice guidelines of the Infectious Diseases Society of America. Other symptoms that are, or have been suggested to be associated with Lyme disease (including those of so-called 'chronic' Lyme disease and post Lyme disease syndromes) are considered too non-specific to define cases for surveillance purposes, whether or not they may be caused by *B. burgdorferi* infection.

⁶ Because available serological screening tests have limitations to their specificity, screening of patients with non-specific subjective symptoms is strongly discouraged. Patients should be made aware that antibody testing is subject to false-positive results, and that a positive test in the absence of objective findings and credible exposure histories usually represent false-positive results.

8.0 References

- Canadian Public Health Laboratory Network. The laboratory diagnosis of Lyme borreliosis: Guidelines from the Canadian Public Health Laboratory Network. *Can J Infect Dis Med Microbiol.* 2007 ;18(2):145-8.
- Ministry of Health and Long-Term Care, Public Health Division. iPHIS manual. Toronto, ON: Queen's Printer for Ontario; 2005.

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- Wormser GP, Dattwyler RJ, Shapiro ED, Halperin JJ, Steere AC, Klemperer MS, et al. The clinical assessment, treatment, and prevention of Lyme disease, human granulocytic anaplasmosis, and babesiosis: clinical practice guidelines by the Infectious Diseases Society of America. Clin Infect Dis. 2006 ;43(9):1089-134. Erratum in: Clin Infect Dis. 2007 ;45(7):941.

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