Appendix 1:
Case Definitions and Disease-Specific Information

Disease: Lyme Disease

Effective: April 2023
Lyme Disease

☐ Communicable
☐ Virulent

Health Protection and Promotion Act (HPPA)¹
Ontario Regulation (O. Reg.) 135/18 (Designation of Diseases)²

Provincial Reporting Requirements

☒ Confirmed case
☒ Probable case

As per Requirement #3 of the “Reporting of Infectious Diseases” section of the Infectious Diseases Protocol, 2018 (or as current), the minimum data elements to be reported for each case are specified in the following:

- O. Reg. 569 (Reports) under the HPPA;
- The iPHIS User Guides published by Public Health Ontario (PHO); and
- Bulletins and directives issued by PHO.³⁴

Type of Surveillance

Case-by-case

Case Definition

Confirmed Case

- Clinician-confirmed erythema migrans (EM) greater than five cm in diameter with a history of residence in, or visit to, a Lyme disease endemic area or-risk area (see Comments section #1, #4 and #5);

OR
• Clinical evidence of Lyme disease (see Comments section #2) with laboratory confirmation by polymerase chain reaction (PCR) or culture (see Comments section #3);

OR

• Clinical evidence of Lyme disease with laboratory support by serological methods (see Comments section #3), and a history of residence in, or visit to, an endemic area or a risk area (see Comments section #4 and #5).

Probable Case

• Clinical evidence of Lyme disease with laboratory support by serological methods (see Comments section #3), with no history of residence in, or visit to an endemic area or a-risk area (see Comments section #4 and #5);

OR

• Clinician-confirmed erythema migrans (EM) greater than five cm in diameter with no history of residence in, or visit to an endemic area or-risk area (see Comments section #1, #4 and #5).

Outbreak Case Definition

Not applicable

Clinical Information

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 EM, the initial skin lesion that occurs in 70 to 80% of patients. Skin lesions may also be atypical, and not have a classic “bull’s-eye” appearance. 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 an authorised health practitioner. 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:
  
  o Nervous system: 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.

  o 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.

  o Cardiovascular system: Acute onset of high-grade (second or third 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.

**Clinical Presentation**

Lyme borreliosis is generally divided into three stages in which infected persons may experience any of the following symptoms:

• Early localized disease
o Erythema migrans (EM) or “bull’s eye” rash at the site of a recent tick bite, fever, malaise, headache, myalgia, neck stiffness, fatigue, lymphadenopathy and arthralgia;

- Early disseminated disease
  o Multiple EM in approximately 15% of people occurs several weeks after infective tick bite, cranial nerve palsies, lymphocytic meningitis, radiculitis, conjunctivitis, arthralgia, myalgia, headache, fatigue, carditis (heart block); and

- Late disease
  o May develop in people with early infection that was undetected or not adequately treated. Involves the heart, nervous system and joints; arrhythmias, heart block and sometimes myopericarditis; recurrent arthritis affecting large joints (i.e., knees); peripheral neuropathy; central nervous system manifestations – meningitis; encephalopathy (i.e., behaviour changes, sleep disturbance, headaches); ophthalmic conditions such as conjunctivitis, optic neuritis, keratitis, and uveitis; and fatigue.5,6

### Laboratory Evidence

### Laboratory Confirmation

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

- Isolation of *Borrelia burgdorferi* (*B. burgdorferi*) from an appropriate clinical specimen;
- Positive nucleic acid amplification test (NAAT) for *B. burgdorferi*; and
- Serological evidence using a two-tier enzyme-linked immuno-sorbent assay (ELISA) using CPHLN/CDC based interpretation criteria.

### Approved/Validated Tests

- Standard culture for *B. burgdorferi*;
• Commercial *B. burgdorferi* Immunoglobulin M (IgM) and Immunoglobulin G (IgG) tests (ELISA); and

• NAAT for *B. burgdorferi*.

**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 two to four weeks, however if patients are treated during this time, subsequent testing may be negative.

• Sera that are screened negative for antibodies using the first ELISA test should not be subjected to the second ELISA.

• When patients are treated very early in the course of illness, antibodies may not develop.

• If the patient visited Europe they should be tested for the European strains of Lyme disease. This can be done by entering ‘European Lyme disease’ under Test Description of the General Test Requisition Form and provide travel history including location of travel and dates.

Diagnosis of Lyme disease is primarily based on clinical symptoms and risk factors as assessed by authorised health practitioners.

For further information about human diagnostic testing, contact the Public Health Ontario Laboratories.

**Case Management**

In addition to the requirements set out in the Requirement #2 of the “Management of Infectious Diseases – Sporadic Cases” and “Investigation and Management of Infectious Diseases Outbreaks” sections of the *Infectious Diseases Protocol, 2018* (or as current), the board of health shall investigate cases to determine the source of infection. Refer to Provincial Reporting Requirements above for relevant data to be collected during case investigation. Additional disease specific information may include:
• Travel to endemic or blacklegged tick risk area and activities in the previous 30 days;
• Outdoor recreational activities and outdoor occupations;
• Symptoms and date of symptom onset and presence or history of EM-like rash; and
• Date of tick bite.3

Treatment is under the direction of the attending healthcare provider. Provide education about the infection and how it is acquired and ways to prevent Lyme disease.

**Contact Management**

Not applicable

**Outbreak Management**

Not applicable

**Prevention and Control Measures**

**Personal Prevention Measures**

Provide public education and advice on preventive measures including:

Education about the mode of tick transmission and the means for personal protection such as:

• Wearing closed shoes and light-coloured, long sleeve shirts and long pants, tucking pants into socks, and using DEET or icaridin insect repellents. The concentration of DEET should be no greater than 30% for adults and no greater than 10% for children;
• Avoiding tick-infested areas when possible; and
• Removing ticks from domestic animals.5,7
Infection Prevention and Control Strategies

The board of health shall develop and utilize a local vector-borne management strategy in order to mitigate risk. This strategy shall include measures such as:

- Local risk assessments; and
- Public education and source reduction when and where applicable.

Additional information on Ontario’s Lyme disease management strategies is available at www.ontario.ca/page/lyme-disease.

Disease Characteristics

Aetiologic Agent - Lyme disease is a tick-borne zoonotic disease caused by the bacterium, *Borrelia burgdorferi* (*B. burgdorferi*), a spirochete first identified in North America in 1982.5,6

Modes of Transmission - Mode of transmission is tick-borne and infection does not occur until an infected tick has been attached for at least 24-36 hours.5,8 This is the amount of time required for the bacteria to migrate from the tick’s gut to its salivary glands where the bacteria are injected into the host.8 *Borrelia* can potentially be transmitted through blood transfusions, although no such cases have been reported.5

Incubation Period – For EM rash, from three to 30 days after tick exposure with a mean of seven to 10 days. Early stages of the illness may not be apparent and the person may present with later manifestations.5

Period of Communicability - There is no evidence of person-to-person spread.5

Reservoir - Deer and small mammals such as rodents serve as important hosts to the tick vector, *Ixodes scapularis*, the primary *B. burgdorferi* vector in eastern Canada and Ontario. This tick is commonly known as a deer tick or blacklegged tick.5 Ixodid ticks become infected as larvae or nymphs when they feed on an infected reservoir host, and they remain infected for life.5
**Host Susceptibility and Resistance** - General susceptibility, with increased risk to those that live in or travel to Lyme disease endemic areas. Reinfection has occurred in those treated with antibiotics.⁵

Public Health Ontario (PHO) produces an annual Lyme Disease Estimated Risk Areas map that identifies areas in Ontario where they estimate you are more likely to find blacklegged ticks ([Lyme Disease](#)). The risk increases where there are established or endemic populations of ticks, however it is possible to contract Lyme disease anywhere in Ontario.⁸⁹

Please refer to [PHO’s Reportable Disease Trends in Ontario reporting tool](#) for the most up-to-date information on infectious disease trends in Ontario.

For additional national and international epidemiological information, please refer to the Public Health Agency of Canada and the World Health Organization.

**Comments**

- EM 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 greater than or equal to five 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 one to two weeks (range three to 30 days) after infection and persists for up to eight 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 EM. Tick bite hypersensitivity reactions are usually less than five cm in largest diameter, sometimes have an urticarial appearance, and typically begin to disappear within 24 to 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.

Clinical evidence of Lyme disease are those symptoms described in on the Public Health Agency of Canada’s Lyme disease webpage. Other symptoms that are or have been suggested to be associated with Lyme disease are considered too non-specific to define cases for surveillance purposes, whether or not they may be caused by *B. burgdorferi* infection.

- As of April 1, 2023 a modified two tier testing (MTTT) replaced the standard two tier testing (STTT), i.e., 2 ELISA (using different *B. burgdorferi* antigen targets) replaced the ELISA followed by the Immunoblot. This new algorithm will capture more positivity (literature shows ranges 15-25%) during the acute stage of Lyme infection which can be missed by the Immunoblot.

- PCR and serological methods on cerebrospinal fluid (CSF) are investigational only. PCR (or more appropriately NAAT) testing should be limited to CSF, joint fluid, or tissue samples as there is limited data to support its use on blood and/or urine samples. 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.

- An endemic travel area is defined here as a country in which reproducing populations of the *Ixodes* tick vectors are known to occur. Lyme disease is endemic in Europe with higher incidence in Central and Eastern Europe. While infectious *Ixodes* ticks occur in Asia, the risk of human infection is low. In the US, Lyme disease is mainly found in the mid-Atlantic, northeastern, and Midwestern states; there are also some reported cases in Washington, Oregon and northern California.

- A risk area is defined here as a location where one blacklegged tick was found during both spring and fall drag sampling of the same year, at the same location (see PHO Lyme Disease Estimated Risk Area Map).
References


Case Definition Sources


Document History

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<td>New template. Appendix A and B merged. No material content changes.</td>
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