Appendix 1: 
Case Definitions and Disease-Specific Information

Disease: *Echinococcus multilocularis* infection

Effective: May 2022
**Echinococcus multilocularis** infection

☒ Communicable  ☐ Virulent

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

**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.

**Type of Surveillance**

Case-by-case

**Case Definitions**

**Confirmed Case**

Laboratory confirmation of infection with clinically compatible signs and symptoms, based on:

- Demonstration of antibodies to *Echinococcus multilocularis* (*E. multilocularis*) in blood or serum sample
  
  OR
  
  - Demonstration of larval stages of *E. multilocularis* in histopathology samples from tissue biopsies
Probable Case

Laboratory confirmation in the absence of clinical signs of alveolar echinococcosis, based on:

- Demonstration of antibodies to *E. multilocularis* in blood or serum sample

Outbreak Case Definition

The outbreak case definition varies with the outbreak under investigation. Please refer to the *Infectious Diseases Protocol, 2018* (or as current) for guidance in developing an outbreak case definition as needed.

The outbreak case definitions are established to reflect the disease and circumstances of the outbreak under investigation. The outbreak case definitions should be developed for each individual outbreak based on its characteristics, reviewed during the course of the outbreak, and modified if necessary, to ensure that the majority of cases are captured by the definition. The case definitions should be created in consideration of the outbreak definitions.

Outbreak cases may be classified by levels of probability (*i.e.*, confirmed and/or probable).

The occurrence of two or more cases linked in time and place to a common exposure is suggestive of an outbreak.

Clinical Information

Clinical Evidence

Infection of humans with *E. multilocularis* is characterized by an initial asymptomatic incubation period of 5 to 15 years.

Proliferation of the larval stage of *E. multilocularis* produces a highly invasive, destructive disease called alveolar echinococcosis. Once clinical signs develop, lesions are usually found in the liver; because the growth of these lesions is not restricted by a thick laminated cyst wall, they expand at the periphery to produce solid, tumour-like masses. Metastases can result in secondary cysts and larval growth in other organs. Clinical manifestations depend on the size and location of cysts, but are often confused with hepatic carcinoma and cirrhosis.
Laboratory Evidence

Laboratory Confirmation
The following will constitute a confirmed case of *E. multilocularis* infection (in the presence of clinically compatible signs and symptoms):

- Demonstration of antibodies to *E. multilocularis* in blood or serum sample (See Approved/Validated Tests)

OR

- Demonstration of larval stages of *E. multilocularis* in histopathology samples from tissue biopsies

Approved/Validated Tests
Serologic testing for antibodies to *E. multilocularis* is performed at the Institute of Parasitology, University of Berne, Switzerland, using a combination of the following assays:

- Em2-antigen enzyme-linked immunosorbent assay (ELISA)
- II/3-10-antigen ELISA
- Em2Plus-antigen ELISA

Additional confirmatory techniques include direct immunofluorescence and/or *Echinococcus* polymerase chain reaction (PCR) of tissue biopsies.

Indications and Limitations
Diagnosis is complex and based on serodiagnosis for early stages of infection, and histopathology for later stages of infection, when lesions have become apparent in the liver or other organs.

Serologic testing for *E. multilocularis* is not performed in Canada. Rather, serum specimens with a requisition for "alveolar hydatid" or "*E. multilocularis*" are sent for reference laboratory testing in Switzerland, after approval of the request by the Public Health Ontario Laboratory.

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.

The following disease-specific information may also be collected:

- Date of symptom onset;
- History of travel to endemic areas;
- Occupation; and
- History of exposure to potential definitive hosts, including foxes, coyotes, wolves, dogs and cats, and/or their feces.

Treatment is under the direction of the attending health care provider. All patients require treatment.

Provide cases with information about the infection and how it is transmitted, as listed above.

Contact Management

None, except if exposed to same source. Contacts exposed to the same source should be assessed for serological testing to monitor for antibodies to *E. multilocularis*. For confirmed cases of alveolar echinococcosis, close family members and associates should be examined for suspicious cysts or tumours using ultrasound, X-ray or other imaging modalities.

Outbreak Management

Please see the *Infectious Diseases Protocol, 2018* (or as current) for the public health management of outbreaks or clusters in order to identify the source of illness, manage the outbreak and limit secondary spread.
Prevention and Control Measures

Personal Prevention Measures

Education of both pet owners and individuals in high risk occupations (veterinary staff, wildlife workers, hunters and trappers, etc.) about the lifecycle of the parasite and risks of exposure to *E. multilocularis* eggs is an important preventive measure.

Hunters and trappers handling foxes, coyotes or other wild canids should wear plastic gloves when handling these animals or their carcasses.

Wild fruits and vegetables should not be collected from the ground or eaten. All wild-picked foods should be washed carefully or cooked before being eaten.

All dogs and cats having access to wild rodents in areas known to be endemic for *E. multilocularis* should be dewormed monthly with praziquantel to reduce the risk of exposure to parasite eggs in household environments. Pet owners should prevent dogs and cats from eating rodents.

Regular, frequent hand hygiene after handling pets and their feces, and before handling food, can reduce the risk of transmission to humans. Areas inhabited by dogs and cats with known *E. multilocularis* infections should be decontaminated to prevent risk of exposure to parasitic eggs on surfaces such as pet beds, floors, carpets and car interiors.

Infection Prevention and Control Strategies

Biological samples containing living larval stages of *E. multilocularis* could be infective to humans if accidentally injected into a person. Therefore, precautions should be taken with regard to correct handling and disposal of needles, scalpel blades and glassware.¹

Routine practices are recommended for hospitalized cases.

Refer to PHO’s website to search for the most up-to-date information on Infection Prevention and Control (IPAC).
Disease Characteristics

**Aetiologic Agent** - *Echinococcus multilocularis* (*E. multilocularis*) infection in humans is caused by the ingestion of eggs of the *E. multilocularis* tapeworm. Once ingested, the eggs develop into the larval form, which grows as multiple, small budding cysts.\(^1\,^2\)

**Modes of Transmission** - Ingestion of eggs passed in the feces of foxes, coyotes, dogs or cats that have fed on infected rodents. Fecally soiled dog hair and other environmental fomites also serve as vehicles of infection.\(^2\)

**Incubation Period** – Infection of humans with *E. multilocularis* is characterized by an initial asymptomatic incubation period of 5 to 15 years.\(^1\)

**Period of Communicability** - *E. multilocularis* eggs are highly resistant, and may remain infective for approximately one year in a suitable, moist environment at lower temperatures.\(^1\) There is no person to person transmission.\(^3\)

**Reservoir** - Adult tapeworm found in foxes, wolves, coyotes, dogs and cats; intermediate hosts are voles, shrews, lemmings and mice; commonly maintained in nature in fox-rodent cycle.\(^3\)

*E. multilocularis* eggs are shed into the environment primarily by foxes, raccoon dogs, and coyotes with gastrointestinal infections with adult tapeworms. Dogs and cats that become infected from hunting wild rodents (such as mice, rats, voles, chipmunks, etc.) can also be sources of human infection.\(^2\)

**Host Susceptibility and Resistance** - Susceptibility is general. Due to the long incubation period, alveolar echinococcosis usually affects adults.\(^2\)

Please refer to [PHO’s Reportable Disease Trends in Ontario reporting tool](https://www.pho.ca/cn/en/disease-trends) 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.
References


Case Definition Sources


### Document History

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<td>April 2022</td>
<td>Entire Document</td>
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<td>April 2022</td>
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