Appendix A: Disease-Specific Chapters

Chapter: *Echinococcus multilocularis* infection

Effective: February 2019
Echinococcus multilocularis infection

☐ Communicable
☐ Virulent

Health Protection and Promotion Act:
O. Reg. 135/18 (Designation of Diseases)

1.0 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

2.0 Case Definition

2.1 Surveillance Case Definition

Refer to Appendix B for Case Definitions.

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.

3.0 Identification

3.1 Clinical Presentation

Proliferation of the larval stage of E. multilocularis produces a highly invasive, destructive disease called alveolar echinococcosis. Once clinical signs of alveolar echinococcosis develop, lesions are usually found in the liver. As 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.²

### 3.2 Diagnosis

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.

See Appendix B for diagnostic criteria relevant to the Case Definitions.

For further information about human diagnostic testing, contact the Public Health Ontario Laboratories or refer to the Public Health Ontario Laboratory Services webpage: http://www.publichealthontario.ca/en/ServicesAndTools/LaboratoryServices/Pages/default.aspx.

### 4.0 Epidemiology

#### 4.1 Occurrence

Distribution is limited to areas in the Northern Hemisphere: central Europe, Commonwealth of Independent States, northern Japan, China, India, Alaska, Canada and north-central USA.³

Historically, *E. multilocularis* was thought to occur in Canada in two zones: northern tundra regions equivalent to the range of arctic fox populations; and a central endemic zone including parts of Alberta, Saskatchewan and Manitoba, and extending southward into 13 contiguous states of the US.¹

Prior to 2009, *E. multilocularis* had never been diagnosed in a dog in Canada. However, in 2009, hepatic alveolar echinococcosis was diagnosed in a 3 year old dog that had never been outside British Columbia. In 2012, a second case was diagnosed in a 2 year old dog that resided in southern Ontario and a third case was diagnosed in a 4 year old dog that lived in Alberta and Manitoba. Between 2013 and 2018, five additional cases were diagnosed in southern Ontario. None of the 8 dogs were related, none had any travel history outside of Canada, and 6 of the dogs had lived their entire lives in provinces where *E. multilocularis* had never been diagnosed prior to the occurrence of these cases. Since then, alveolar echinococcosis has also been identified in two lemurs, who were allowed to roam outdoors in southern Ontario and were coming into contact with wildlife.

A research study conducted by the University of Guelph in 2015-2017 confirmed that *E. multilocularis* has become established in coyote and fox populations across southern Ontario, with potential hotspots of higher rates of infection in some health units.
Passive wildlife surveillance also identified *E. multilocularis* infection in a chipmunk from southern Ontario in 2016, further confirming that all stages of the tapeworm’s lifecycle are now present in the Ontario environment.

While alveolar echinococcosis in humans was not reportable prior to 2018, data from the Canadian Institute for Health Information indicate that there have been at least three human cases of alveolar echinococcosis diagnosed in Ontario since 2014.

Please refer to Public Health Ontario’s (PHO) Reportable Disease Trends in Ontario reporting tool and other reports for the most up-to-date information on infectious disease trends in Ontario.

http://www.publichealthontario.ca/en/DataAndAnalytics/Pages/DataReports.aspx

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

4.2 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

4.3 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

4.4 Incubation Period

Infection of humans with *E. multilocularis* is characterized by an initial asymptomatic incubation period of 5 to 15 years.1

4.5 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

4.6 Host Susceptibility and Resistance

Susceptibility is general. Due to the long incubation period, alveolar echinococcosis usually affects adults.2
5.0 Reporting Requirements

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:

- Ontario Regulation 569 (Reports) under the Health Protection and Promotion Act (HPPA);\(^4\)
- The iPHIS User Guides published by PHO; and
- Bulletins and directives issued by PHO.

6.0 Prevention and Control Measures

6.1 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.

6.2 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 at [www.publichealthontario.ca](http://www.publichealthontario.ca) to search for the most up-to-date information on Infection Prevention and Control.
6.3 Management of Cases

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 Section 5: 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.

6.4 Management of Contacts

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.

6.5 Management of Outbreaks

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.

7.0 References


8.0 Document History

Table 1: History of Revisions

<table>
<thead>
<tr>
<th>Revision Date</th>
<th>Document Section</th>
<th>Description of Revisions</th>
</tr>
</thead>
<tbody>
<tr>
<td>March 2018</td>
<td>Entire appendix</td>
<td>E. multilocularis was designated as a disease of public health significance effective May 1, 2018.</td>
</tr>
<tr>
<td>February 2019</td>
<td>General</td>
<td>Common text included in all Disease Specific chapters: Surveillance Case Definition, Outbreak Case Definition, Diagnosis, Reporting Requirements, Management of Cases, and Management of Outbreaks.</td>
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<tr>
<td>February 2019</td>
<td>4.1 Occurrence</td>
<td>First sentence added. “Distribution is limited to area in the Northern Hemisphere….”</td>
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