

Echinococcus multilocularis

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Background

Echinococcus tapeworms are the cause of echinococcosis, a zoonotic parasitic infection in humans. There are currently four known species of tapeworm that cause echinococcal infection in humans; *E. multilocularis, E. granulosus, E. vogeli,* and *E. oligarthrus.*¹ Of these, *E. granulosus* and *E. multilocularis* are the most relevant from a public health perspective.¹ The geographic distribution and animal host species vary by *Echinococcus* species. This summary specifically describes echinococcal infection with the species *E. multilocularis* (alveolar echinococcosis), an emerging pathogen in Ontario.

This document provides information on *Echinococcus multilocularis* (*E. multilocularis*) for health care providers. This includes local public health unit investigators, medical officers of health, attending physicians, nurses, and other health care providers involved in delivery of patient care.

Key Messages

- Echinococcus multilocularis (E. multilocularis) is a small tapeworm that is normally found in canids (coyotes, foxes, dogs) and rodents.¹
- Ontario is now considered an endemic area for *E. multilocularis*, creating the potential for human exposure.²
- Infection of humans can occur following ingestion of *E. multilocularis* eggs, which hatch in the small intestine and enter the blood or lymphatic system.¹ The larvae then travel to the liver and other organs where they encyst.¹
- Alveolar echinococcosis is a slow progressing disease, but can be serious if untreated, and is often difficult to treat.³
- Persons with exposure to a domestic dog with confirmed shedding of *E. multilocularis* eggs should be investigated serologically.⁴ Prophylaxis should be considered in persons with a high likelihood of egg ingestion and with serologic evidence of infection.³
- Echinococcus multilocularis is a disease of public health significance in Ontario.⁴
- Regular (monthly) deworming of dogs that may have potential exposure to infected rodents can reduce the risk of human exposure.⁵ Praziquantel is preferred.⁵

Life Cycle

The life cycle for *E. multilocularis* requires both a definitive host and an intermediate host.^{1,6} Adult tapeworms live within the intestine of the definitive host (usually dogs or wild canids such as foxes, wolves or coyotes), whereas intermediate hosts harbour tapeworms in their larval stage.¹ Eggs are passed via the feces of the definitive host into the environment where they can be ingested by an intermediate host such as a rodent, rabbit, or other small mammal.^{1,6} Humans may become an accidental intermediate host if they ingest the eggs of the tapeworm.¹ This may occur through direct contact with feces of domestic dogs (including fecal contamination on an dog's mouth and fur), contact with other surfaces contaminated with feces, or ingestion of food or water that has been contaminated with the feces of an infected definitive host (wild or domestic).^{1,6,7} Humans cannot become infected by ingesting the tissues of an infected intermediate host (e.g., by eating infected tissue (meat) from a small mammal).⁷ Definitive hosts acquire infection by ingestion of cysts, typically from eating the viscera or tissues of an intermediate host.^{1,6} When ingested by a definitive host, the larvae in the cysts migrate to the small intestine where they mature into adult tapeworms, completing the life cycle of the parasite.^{1,6}

Unlike some other zoonotic parasites, *E. multilocularis* eggs are immediately infective. The eggs are tolerant to cold and freezing temperatures, and under optimal conditions, can survive for prolonged periods (over 12 months) in the environment.⁸

Upon ingestion by an intermediate host, the tapeworm eggs hatch, releasing larvae that pass through the intestinal mucosa into the blood or lymphatic system.¹ The larvae move to the liver or other target organs where they form cysts. In intermediate hosts and accidental hosts (such as humans), the alveolar hydatid cysts behave like malignant neoplasias, and may spread elsewhere in the body.¹

In humans, the period between initial ingestion of eggs and development of symptoms is prolonged, with most people infected with the parasite remaining asymptomatic for 5-15 years.⁴ This can make identification of the source of exposure challenging. Although a human may become an accidental intermediate host, the infection cannot be transmitted from person-to-person.¹

As urban or suburban human populations encroach on areas with established fox, wolf, and coyote populations, or as ranges of wild canids expand, the risk of exposure for humans and domestic animals to wild canid feces increases. In endemic regions, people who have close contact with dogs that may consume infected rodents, or who may have increased contact with soil contaminated with canid feces, (such as farmers, hunters, rural and suburban dog owners) are at increased risk of alveolar echinococcosis.

Emerging Evidence in Ontario

Echinococcus multilocularis is only found in the northern hemisphere, and human infections have been documented in continental Europe, Russia, China and Alaska.¹ Prior to 2012, *E. multilocularis* had never been diagnosed in either wildlife or domestic animals in Ontario.⁶ However, since 2012, multiple dogs and various wildlife species (including rodents) across southern Ontario have been diagnosed with alveolar echinococcosis or intestinal echinococcosis (canids only).^{6,9–11} A recent study examining the prevalence of *E. multilocularis* in wild canids (coyotes and foxes) in Ontario from 2015-2017 found that just under a quarter (23%; 95% confidence interval 20%-27%) of canids tested were positive for the parasite.⁶

Since canine cases of alveolar echinococcosis had previously only been reported in highly endemic areas, this suggests that there were likely previously unrecognized infections in wild canids in multiple areas of southern Ontario. Several human cases of infection have also been identified in Ontario.¹² Subsequently, *E. multilocularis* was declared a disease of public health significance in 2018, requiring the provincial reporting of all confirmed and probable human cases of infection, and Ontario is now considered an endemic area for *E. multilocularis*.^{2,4,13}

Diagnosis and Treatment of Human Infection

Humans infected with *E. multilocularis* are typically asymptomatic, displaying no outward signs of infection for the first 5-15 years after exposure, before developing non-specific symptoms such as abdominal pain, malaise and weight loss.^{1,4} Symptoms vary depending on the location and size of cysts, but may be confused with hepatocellular carcinoma or cirrhosis at initial presentation.⁴ If left untreated, the disease is invariably fatal.⁶

Diagnosis of alveolar echinococcosis is complicated and depends on whether an individual is in the early or later stages of infection.⁴ In Ontario, diagnosis of infection, and classification of an individual as a confirmed or probable case, is dependent on serology/histopathology.⁴ In early infection the disease may be diagnosed through serology (through identification of *E. multilocularis* antibodies in blood or serum).⁴ In the later stages of infection, when lesions are apparent in the liver or other organs, infection may be diagnosed through histopathology, where larval stages of the parasite are demonstrated through tissue biopsy.⁴

Currently, serological testing for *E. multilocularis* is not performed in Ontario or elsewhere in Canada.⁴ If alveolar echinococcosis is suspected in a patient, Public Health Ontario's laboratory can facilitate the submission of serum specimens to Switzerland for reference laboratory testing following consultation and approval by the microbiologist.⁴

With early diagnosis and treatment the prognosis of this disease has improved significantly.^{3,14} Treatment may involve surgical removal of parasitic lesions and long-term treatment with anti-parasitic or chemotherapeutic medications.^{3,14}

Disease Prevention

Since exposure to eggs in canid feces is the primary route of human exposure, important preventive measures include good hygiene practices (especially handwashing) when handling wild or domestic canid feces or after exposure to areas where wild or domestic canids may have defecated. Routine (monthly) deworming of dogs that may be exposed to infected rodents (e.g. dogs that are allowed to roam off-leash with access to rodent habitats in parks and gardens and that may hunt or scavenge small rodents) with praziquantel can reduce the risk of patent infection in domestic dogs, as well as within-household exposure to parasite eggs from infected pets.^{5,15}

Summary

Healthcare providers should be aware of the potential for domestic canine and human exposure to *Echinococcus multilocularis* in Ontario and should be aware of the symptoms associated with this disease, in the event that a patient presents with a compatible illness. Additionally, dog owners should be aware of actions that can be taken to minimize the risk of themselves or their pet being exposed to the parasite as a means of primary prevention.

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