Echinococcus multilocularis: Information for health care providers

Introduction

Echinococcus tapeworms are the cause of echinococcal disease in humans. There are currently four known species that cause echinococcal infection. E. granulosus and E. multilocularis are the most common. The geographic distribution and animal host species vary by Echinococcus species. This summary specifically describes echinococcal infection with the species E. multilocularis (alveolar echinococcosis) and the possibility of the emergence of E. multilocularis among the Ontario population.

Life cycle

The life cycle for E. multilocularis requires two mammalian species. Adult tapeworms live within the intestine of the definitive host (usually dogs or wild canids such as fox or coyote). Eggs are passed in the feces of the definitive host into the environment where they can be ingested by an intermediate host or an incidental human. Unlike some other zoonotic parasites, Echinococcus eggs are immediately infective. These eggs are resistant and are able to survive in a moist environment including water at temperatures of 4 C and above for over a year.

Rodents are the intermediate host. Upon ingestion, the eggs hatch, releasing oncospheres that pass through the intestinal mucosa into the blood or lymphatics. They move to the liver or other organs where they become a metacestode (alveolar hydatid cyst).

Humans can be accidental hosts, becoming infected by ingesting eggs shed in the stool of the definitive host (i.e. a dog or wild canids such as fox or coyote).

In intermediate hosts and accidental hosts (such as humans), the alveolar hydatid cyst behaves like a tumour and undergoes exogenous budding (i.e. buds externally), and may spread elsewhere in the body. Definitive hosts acquire infection by ingestion of metacestodes, typically from ingestion of infected rodents. Thereafter, there is attachment to the intestinal mucosa and development into adult tapeworms.

As urban or suburban human populations encroach on areas with established fox and coyote populations, or ranges of those wild canids expand, risk of exposure for humans and domestic animals to wild canids (and wild canid feces) increase. If the life cycle of this parasite becomes established in an area, dogs that consume wild rodents may develop an intestinal infection and pass infective eggs into
the environment. Unusually, dogs may also develop metacestodes through consuming eggs in the stool of wild canids; however, the metacestode stage is of no direct zoonotic risk.

Transmission of this parasite from human to human does not occur as two different mammalian species are needed to complete the life cycle.

**Epidemiology in humans**

Human infection with *E. multilocularis* has been documented in central Europe, Russia, central Asian countries, China and Alaska. Until recently, *E. multilocularis* was not believed to be present in Ontario. However, recent documentation of cases in multiple dogs and wildlife in different areas of southwestern Ontario may indicate otherwise.

In endemic regions, humans with close contact to dogs that potentially consume infected rodents (such as farmers, hunters, rural and suburban dog owners) are at increased risk of alveolar echinococcosis. Canadian human incidence data are limited and often lack details about the *Echinococcus* species and likely region of exposure. However, to date, there has not been documented acquisition of human disease with *E. multilocularis* in Ontario.

**Emerging evidence in Ontario**

Prior to 2012, *E. multilocularis* had never been diagnosed in either wildlife or domestic animals in Ontario. However, between 2012 and 2016, multiple dogs across southern Ontario have been diagnosed as being infected with alveolar echinococcosis. These cases have been unrelated and due to the lack of travel, appear to have acquired the infection in southern Ontario. The parasite has also been identified in wild rodents in the province.

Since all Ontario dogs must have become infected by exposure to the eggs of *E. multilocularis*, this suggests wild canids are likely shedding *E. multilocularis* eggs into the environment in multiple areas of the province. Since canine cases of alveolar echinococcosis have previously only been reported in highly endemic areas, this also suggests that there are likely significant levels of infection in wild canids in multiple areas of southern Ontario. A recent study has identified shedding of *E. multilocularis* by wild canids in multiple areas of the province (personal communication, Dr. A. Peregrine, Ontario Veterinary College, University of Guelph).

**Clinical manifestations in humans**

Frequently, infections with *E. multilocularis* are asymptomatic for 5-15 years prior to the development of nonspecific symptoms. Since extra-hepatic disease is very rare (1 per cent), the presenting complaints are consistent with hepatic disease including malaise, weight loss, and right upper quadrant pain. Symptoms consistent with hepatic duct involvement include jaundice, cholangitis, portal hypertension and Budd-Chiari syndrome may all occur. The disease can be confused with hepatocellular carcinoma at initial presentation.

*E. multilocularis* can cause very severe disease in humans and if untreated the case fatality can be as high as 90 percent within 10 years of onset of clinical symptoms, and virtually 100 percent by 15 years.
With treatment the prognosis of this disease has improved significantly. Early diagnosis and treatment with albendazole improves life expectancy significantly.

**Diagnosis**

The diagnosis of alveolar hydatid in humans is confirmed serologically, and supported by compatible radiographic imaging (i.e., abdomen CT scan or MRI), and histopathology of biopsied tissue. Serologic testing for alveolar hydatid 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 PHO Laboratory parasitologist.

**Prevention**

Since exposure to eggs in canid feces is the route of exposure, good hygiene practices when handing canine feces, avoiding exposure to feces from wildlife and basic hygiene practices (especially handwashing after exposure to areas where canids may have defecated) are important preventive measures. Routine (monthly) treatment of dogs that may be exposed to infected rodents (e.g. dogs that are allowed to roam or that have access to rodent habitats in parks and gardens) with antiparasitics that are effective against *E. multilocularis* should be considered.

Investigation of human contacts of dogs with *E. multilocularis* infection is important to identify situations where prophylactic treatment of those contacts may be indicated. Concerns regarding human exposure apply to dogs with intestinal infections as well as dogs with alveolar echinococcosis, since the latter group may also have concurrent intestinal infections. Dogs are no longer considered infectious after 24-48h of appropriate treatment. However, defining the onset of infectivity is impossible because of the potential for long-term intestinal infection.

**Summary**

Although *E. multilocularis* has yet to be diagnosed in a human resident of Ontario, the diagnosis of alveolar hydatid in three domestic dogs in Ontario raises the concern that this pathogen is established within the wild canid population. The long incubation period for alveolar echinococcosis means that there could a significant delay from establishment of this parasite in the province and identification of canine infections to detection of human infections. Population level surveillance research projects are required within the Ontario wild canid population to gather baseline knowledge of the presence of this potentially zoonotic infection. Greater awareness of this pathogen is required for physicians who diagnosis or treat hepatic disease. While there is currently no evidence of disease in humans in Ontario, the presence of this pathogen raises the potential for serious zoonotic infection of humans as incidental hosts.

For an overview of this information, see the fact sheet, *Echinococcus multilocularis: Five things to know.*
Sources


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