

Echinococcosis

[*Echinococcus granulosus*] [*Echinococcus multilocularis*] [*Echinococcus oligarthrus*]
[*Echinococcus vogeli*]

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Causal Agents

Human echinococcosis (hydatidosis, or hydatid disease) is caused by the larval stages of cestodes (tapeworms) of the genus *Echinococcus*. *Echinococcus granulosus* (sensu lato) causes cystic echinococcosis and is the form most frequently encountered. Another species, *E. multilocularis*, causes alveolar echinococcosis, and is becoming increasingly more common. Two exclusively New World species, *E. vogeli* and *E. oligarthrus*, are associated with "Neotropical echinococcosis"; *E. vogeli* causes a polycystic form whereas *E. oligarthrus* causes the extremely rare unicystic form.

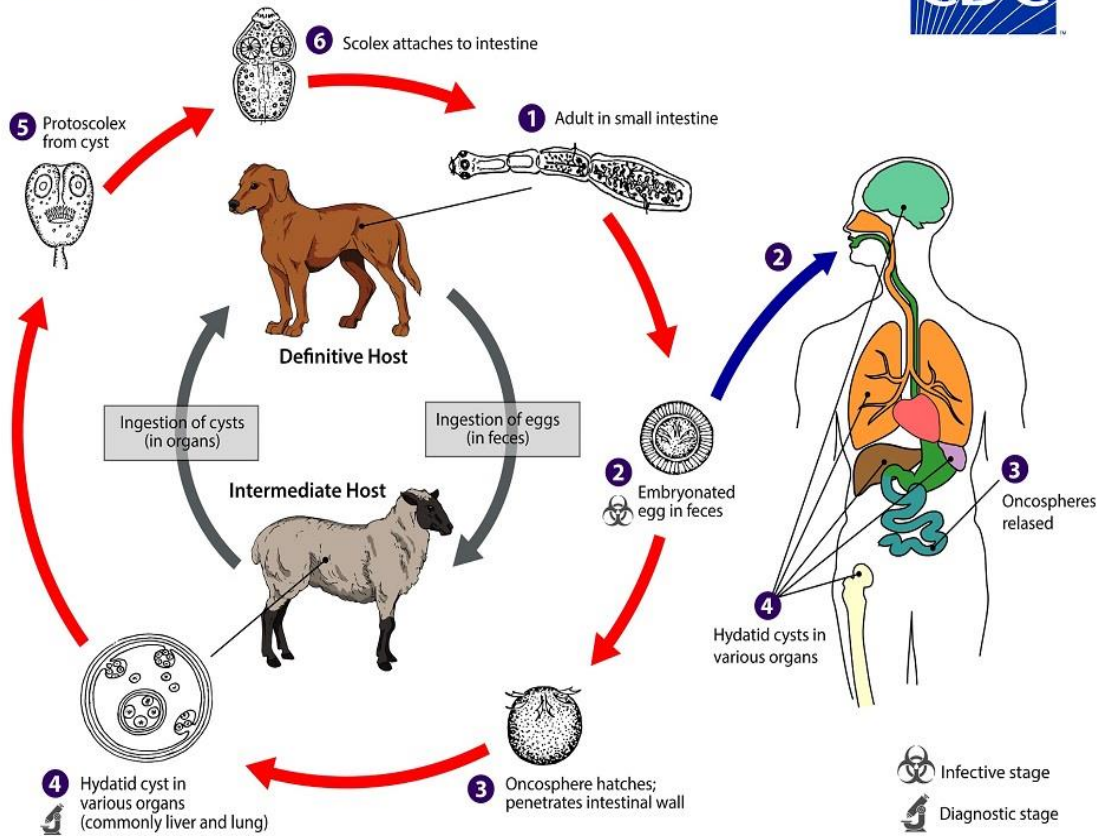
Many genotypes of *E. granulosus* have been identified that differ in their distribution, host range, and some morphological features; these are often grouped into separate species in modern literature. The known zoonotic genotypes within the *E. granulosus* sensu lato complex include the "classical" *E. granulosus* sensu stricto (G1–G3 genotypes), *E. ortleppi* (G5), and the *E. canadensis* group (usually considered G6, G7, G8, and G10). Research on the epidemiology and diversity of these genotypes is ongoing, and no consensus has been reached on appropriate nomenclature thus far.

Life Cycle

Cystic Echinococcosis (*Echinococcus granulosus sensu lato*)



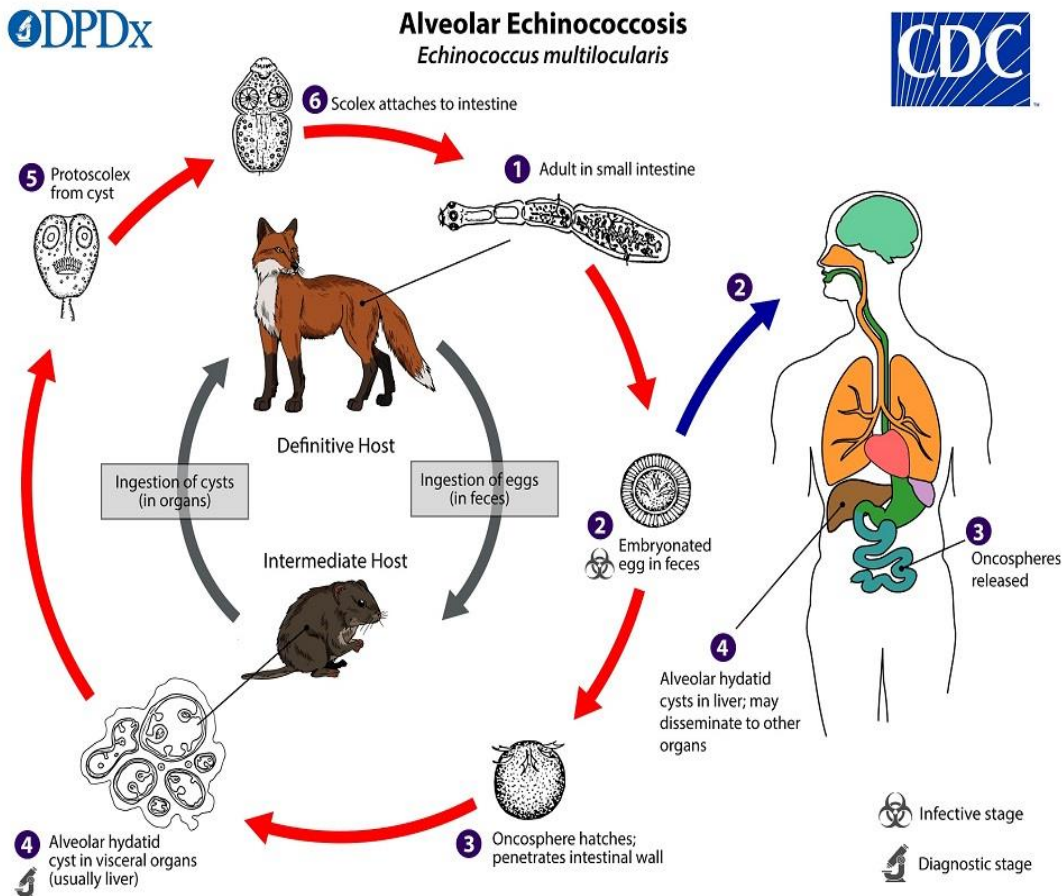
Cystic Echinococcosis *Echinococcus granulosus sensu lato*



The adult *Echinococcus granulosus* (sensu lato) (2—7 mm long) ¹ resides in the small intestine of the definitive host. Gravid proglottids release eggs ² that are passed in the feces, and are immediately infectious. After ingestion by a suitable intermediate host, eggs hatch in the small intestine and release six-hooked oncospheres ³ that penetrate the intestinal wall and migrate through the circulatory system into various organs, especially the liver and lungs. In these organs, the oncosphere develops into a thick-walled hydatid cyst ⁴ that enlarges gradually, producing protoscolices and daughter cysts that fill the cyst interior. The definitive host becomes infected by ingesting the cyst-containing organs of the infected intermediate host. After ingestion, the protoscolices ⁵ evaginate, attach to the intestinal mucosa ⁶, and develop into adult stages ¹ in 32 to 80 days.

Humans are aberrant intermediate hosts, and become infected by ingesting eggs ². Oncospheres are released in the intestine ³, and hydatid cysts develop in a variety of organs ⁴. If cysts rupture, the liberated protoscolices may create secondary cysts in other sites within the body (secondary echinococcosis).

Alveolar Echinococcosis (*Echinococcus multilocularis*)



The adult *Echinococcus multilocularis* (1.2—4.5 mm long) **1** resides in the small intestine of the definitive host. Gravid proglottids release eggs **2** that are passed in the feces, and are immediately infectious. After ingestion by a suitable intermediate host, eggs hatch in the small intestine and releases a six-hooked oncosphere **3** that penetrates the intestinal wall and migrates through the circulatory system into various organs (primarily the liver for *E. multilocularis*). The oncosphere develops into a multi-chambered ("multilocular"), thin-walled (alveolar) hydatid cyst **4** that proliferates by successive outward budding. Numerous protoscolices develop within these cysts. The definitive host becomes infected by ingesting the cyst-containing organs of the infected intermediate host. After ingestion, the protoscolices **5** evaginate, attach to the intestinal mucosa **6**, and develop into adult stages **1** in 32 to 80 days.

Humans are aberrant intermediate hosts, and become infected by ingesting eggs **2**. Oncospheres **3** are released in the intestine and cysts develop within in the liver **4**. Metastasis or dissemination to other organs (e.g., lungs, brain, heart,

bone) may occur if protoscolices are released from cysts, sometimes called "secondary echinococcosis."

Neotropical Echinococcosis (*Echinococcus vogeli*, *E. oligarthrus*)

The Neotropical agents follow the same life cycle although with differences in hosts, morphology, and cyst structure. Adults of *E. vogeli* reach up to 5.6 mm long, and *E. oligarthrus* up to 2.9 mm. Cysts are generally similar to those found in cystic echinococcosis but are multi-chambered.

Hosts

Echinococcus granulosus definitive hosts are wild and domestic canids. Natural intermediate hosts depend on genotype. Intermediate hosts for zoonotic species/genotypes are usually ungulates, including sheep and goats (*E. granulosus* sensu stricto), cattle ("*E. ortleppi*"/G5), camels ("*E. canadensis*"/G6), and cervids ("*E. canadensis*"/G8, G10).

For *E. multilocularis*, foxes, particularly red foxes (*Vulpes vulpes*), are the primary definitive host species. Other canids including domestic dogs, wolves, and raccoon dogs (*Nyctereutes procyonoides*) are also competent definitive hosts. Many rodents can serve as intermediate hosts, but members of the subfamily Arvicolinae (voles, lemmings, and related rodents) are the most typical.

The natural definitive host of *E. vogeli* is the bush dog (*Speothos venaticus*), and possibly domestic dogs. Pacas (*Cuniculus paca*) and agoutis (*Dasyprocta* spp.) are known intermediate hosts. *E. oligarthrus* uses wild neotropical felids (e.g. ocelots, puma, jaguarundi) as definitive hosts, and a broader variety of rodents and lagomorphs as intermediate hosts.

Geographic Distribution

Echinococcus granulosus sensu lato occurs practically worldwide, and more frequently in rural, grazing areas where dogs ingest organs from infected animals. The geographic distribution of individual *E. granulosus* genotypes is variable and an area of ongoing research. The lack of accurate case reporting and genotyping currently prevents any precise mapping of the true epidemiologic picture. However, genotypes G1 and G3 (associated with sheep) are the most commonly reported at present and broadly distributed. In North America, *Echinococcus granulosus* is rarely reported in Canada and Alaska, and a few human cases have also been reported in Arizona and New Mexico in sheep-raising areas. In the United States, most infections are diagnosed in immigrants from counties where cystic echinococcosis is endemic. Some genotypes designated "*E. canadensis*" occur broadly across Eurasia, the Middle

East, Africa, North and South America (G6, G7) while some others seem to have a northern holarctic distribution (G8, G10).

E. multilocularis occurs in the northern hemisphere, including central and northern Europe, Central Asia, northern Russia, northern Japan, north-central United States, northwestern Alaska, and northwestern Canada. In North America, *Echinococcus multilocularis* is found primarily in the north-central region as well as Alaska and Canada. Rare human cases have been reported in Alaska, the province of Manitoba, and Minnesota. Only a single autochthonous case in the United States (Minnesota) has been confirmed.

E. vogeli and *E. oligarthrus* occur in Central and South America.

Clinical Presentation

Echinococcus granulosus infections often remain asymptomatic for years before the cysts grow large enough to cause symptoms in the affected organs. The rate at which symptoms appear typically depends on the location of the cyst. Hepatic and pulmonary signs/symptoms are the most common clinical manifestations, as these are the most common sites for cysts to develop. In addition to the liver and lungs, other organs (spleen, kidneys, heart, bone, and central nervous system, including the brain and eyes) can also be involved, with resulting symptoms. Rupture of the cysts can produce a host reaction manifesting as fever, urticaria, eosinophilia, and potentially anaphylactic shock; rupture of the cyst may also lead to cyst dissemination.

Echinococcus multilocularis affects the liver as a slow growing, destructive tumor, often with abdominal pain and biliary obstruction being the only manifestations evident in early infection. This may be misdiagnosed as liver cancer. Rarely, metastatic lesions into the lungs, spleen, and brain occur. Untreated infections have a high fatality rate.

Echinococcus vogeli affects mainly the liver, where it acts as a slow growing tumor; secondary cystic development is common. Too few cases of *E. oligarthrus* have been reported for characterization of its clinical presentation.