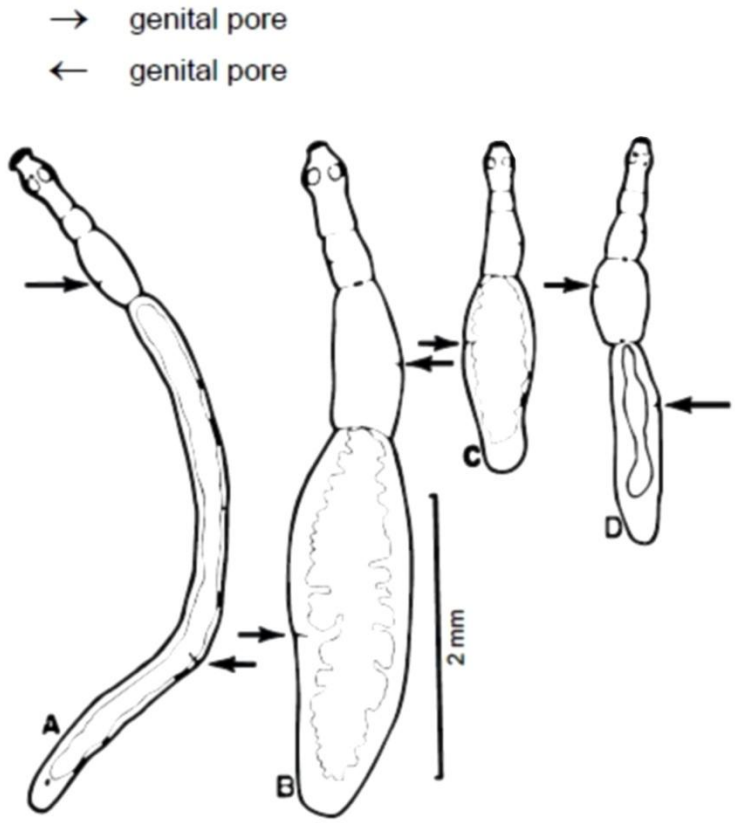
1.1. Introduction

Echinococcosis is a zoonotic infection caused by adult or larval (metacestode) stages of cestodes belonging to the genus *Echinococcus* and the family *Taeniidae*. At present, four species of *Echinococcus* are recognised, namely *Echinococcus granulosus*, *E. multilocularis*, *E. oligarthrus* and *E. vogeli*. The parasites are perpetuated in life-cycles with carnivores as definitive hosts, which harbour the adult egg-producing stage in the intestine, and intermediate host animals, in which the infective metacestode stage develops after peroral infection with eggs.
Within the species *E. granulosus*, genetic heterogeneity is common resulting in a number of intraspecific variants or 'strains'.



A : *Echinococcus vogeli*
B : *Echinococcus granulosus*
C : *Echinococcus oligarthrus*
D : *Echinococcus multilocularis*

Fig.
Comparative general morphology of adult *Echinococcus* species

1.2. General morphology

General features

Echinococcus exhibits certain unique characteristics that set it apart from the other major genus in the family, *Taenia*. An adult *Echinococcus* is only a few millimetres long (rarely more than 7 mm) and usually has no more than six segments, whereas species of *Taenia* can grow to several metres in length and consist of several thousand segments. Like all tapeworms, *Echinococcus* has no gut and all metabolic interchange takes place across the syncytial outer covering, the tegument. The penultimate segment is mature, and the genital pore normally opens posterior to the middle of both mature and gravid segments. The gravid uterus is characterised by welldeveloped lateral sacculations.

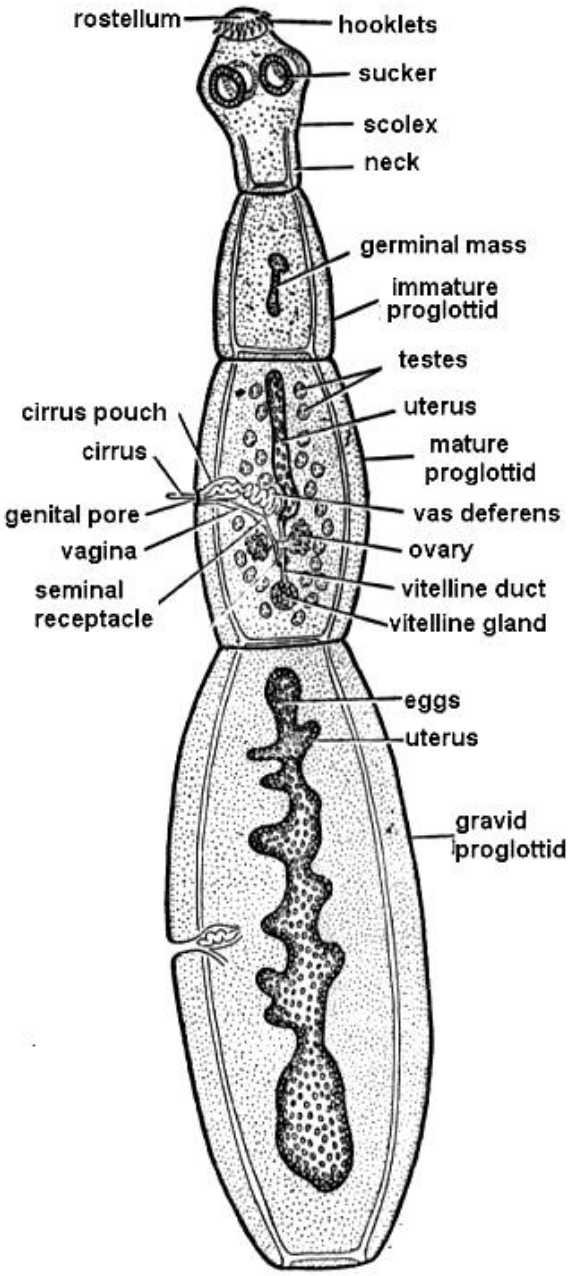


Fig. Adult *E. granulosus*

Scolex and strobila

Anteriorly, the adult *Echinococcus* possesses a specialised attachment organ, the scolex, which has four muscular suckers and two rows of hooks, one large and one small, on the rostellum. The body, or strobila, is segmented and consists of a number of reproductive units (proglottids), which may vary in number
from two to six. The adult worm is hermaphrodite with reproductive ducts opening at a common, lateral,
genital pore, the position of which may vary depending on species and strain. There is a prominent cirrus sac,
which may be horizontal or tilted anteriorly and the vitellarium is globular. The uterus dilates after
fertilisation, eventually occupying most of the terminal segment when the eggs are fully developed.

Eggs

The eggs are ovoid (30 µm-40 µm diameter), consisting of a hexacanth embryo (oncosphere = first larval

stage) surrounded by several envelopes, the most noticeable one being the highly resistant keratinized embryophore, which gives the egg a dark striated appearance. The outer capsule quickly disappears once the eggs are liberated from the host. The eggs of *Echinococcus* are morphologically indistinguishable to those of other tapeworms of the genus *Taenia.*

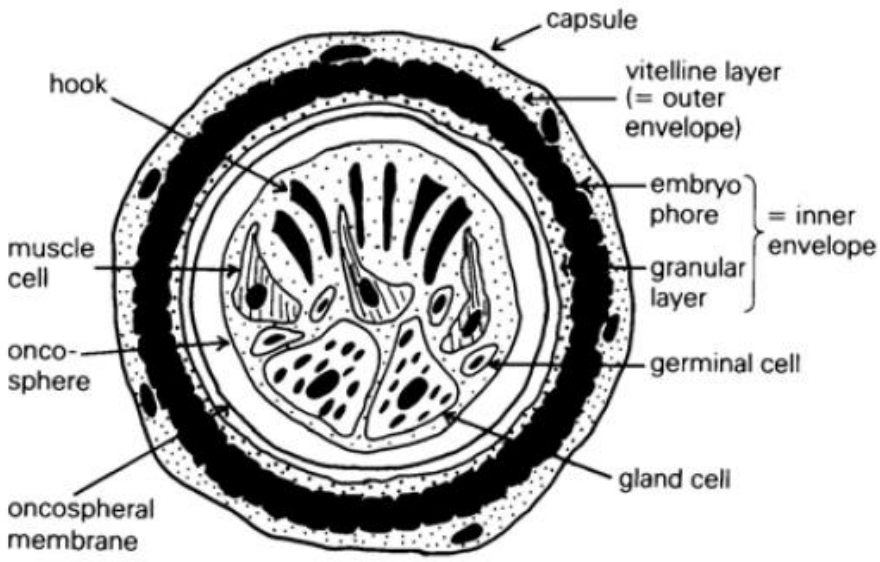


Fig.
Diagram of the egg of ***Echinococcus***

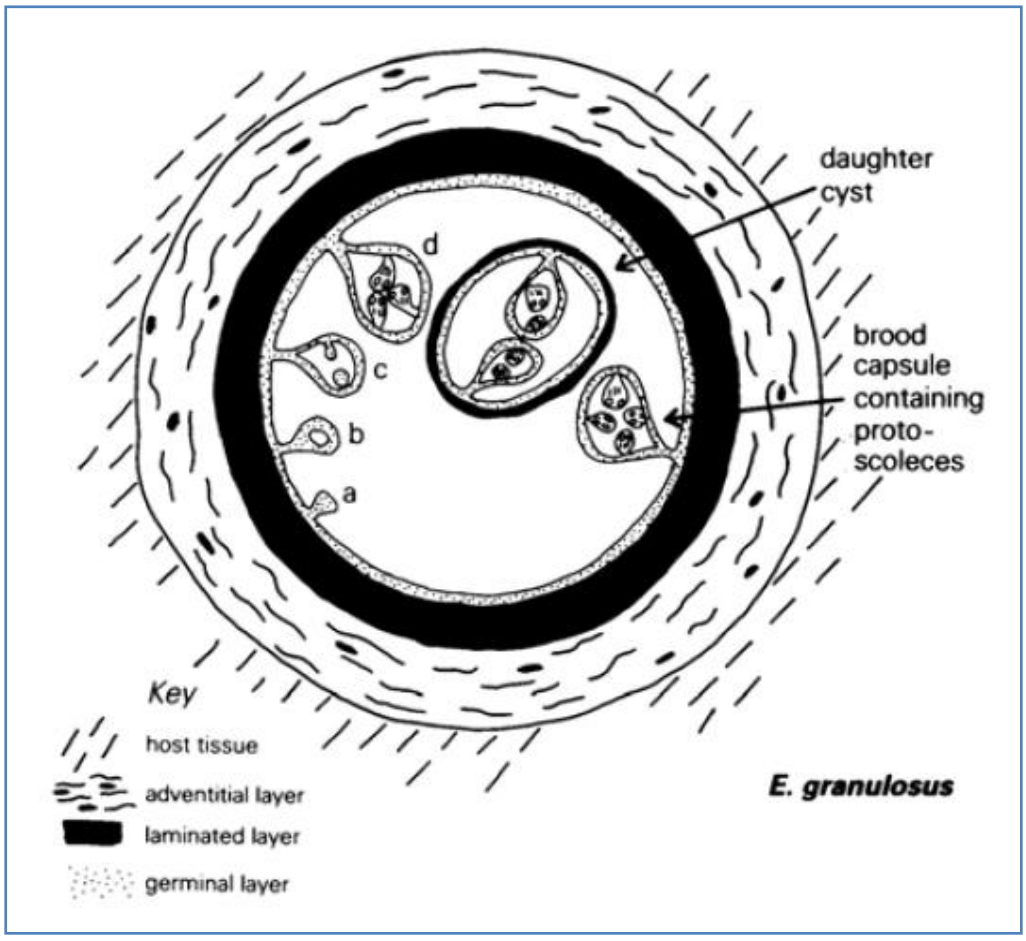


Fig.
Diagrammatic representation of the metacestode of ***Echinococcus granulosus***

Metacestode

The metacestode (= second larval stage) basically consists of a bladder with an outer acellular laminated layer
and an inner nucleated germinal layer, which may give rise by asexual budding to brood capsules. Protoscoleces arise from the inner wall of the brood capsules. The structure and development of the metacestode differs between the four species of *Echinococcus* .

1.3. Geographic distribution

E. granulosus s. l. occurs worldwide, with the exception of a few countries such as Iceland and Greenland.

1.4. Hosts

Echinococcosis is a zoonotic disease caused by *Echinococcus* spp. tapeworms. The definitive hosts, which include dogs, other canids, hyenas and cats, carry the adult tapeworms subclinically. Dogs are particularly important in zoonotic transmission due to their close relationships with humans. Intermediate hosts are initially asymptomatic; however, the growth of the larvae, which form cysts in vital organs such as the liver and lungs, can lead to illness and death.

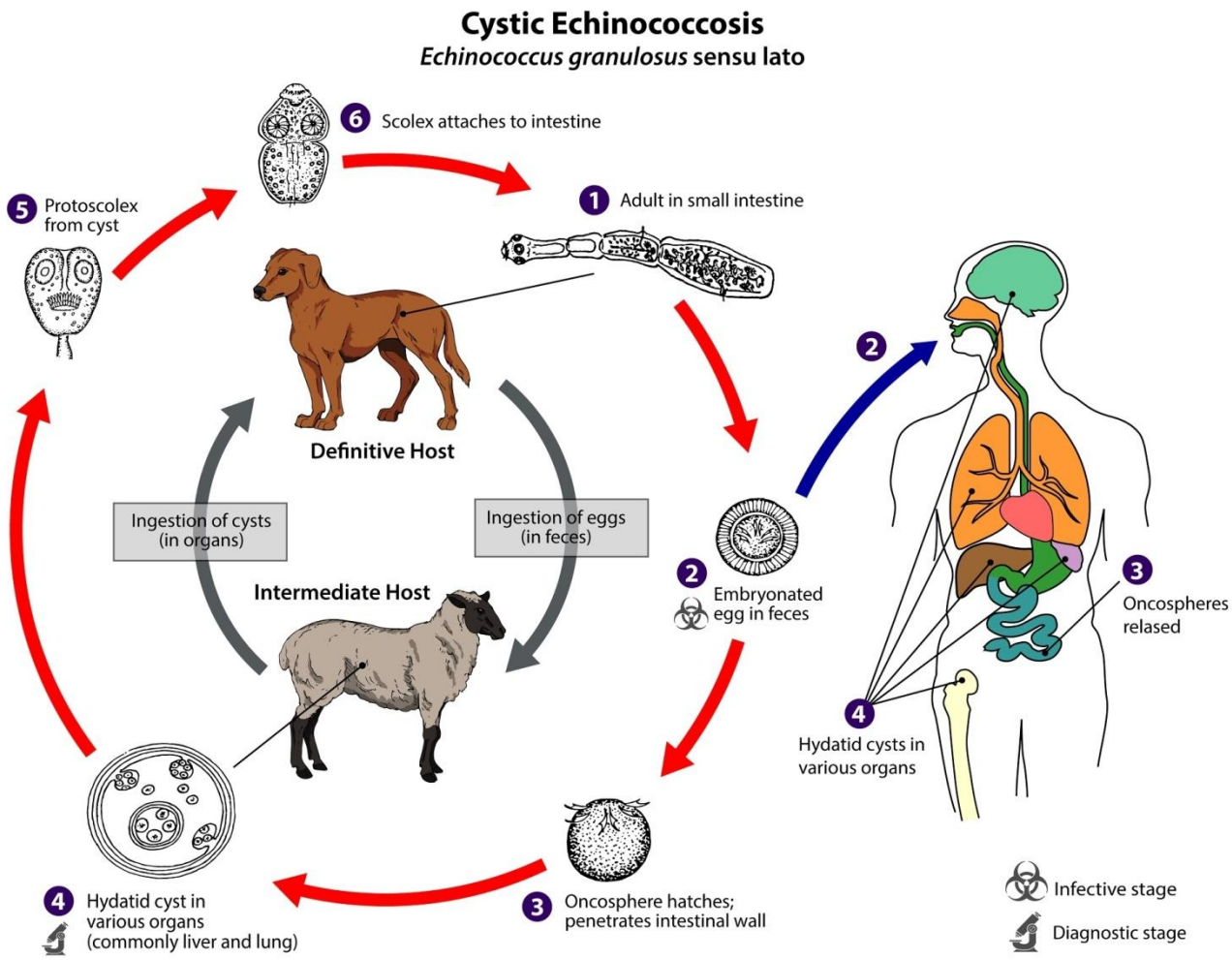
1.5. Life cycle

Echinococcus species have an indirect life cycle, and must develop in both an intermediate and a definitive host. In many cases, the parasite cycles through specific predators or scavengers, and their prey.

The definitive hosts for *E. granulosus s. l.* (canids, felids, and hyaenids) become infected when they ingest cysts (metacestodes) in the tissues of the intermediate hosts. Feeding the viscera of intermediate hosts to dogs perpetuates cycles in domesticated animals. The cysts develop into tapeworms, which mature in the host's small intestine. Gravid proglottids or eggs are shed in the feces, and are immediately infective. *Echinococcus* eggs have a sticky coat that will adhere to an animal's fur and other objects. Insects such as flies and beetles, or birds, can also act as mechanical vectors. In addition, the shed proglottids may perform rhythmic contractions that help to disperse the eggs widely on pastures.

Under ideal conditions, *E. granulosus s. l.* eggs remain viable for several weeks or months in pastures or gardens, and on fomites. They survive best under moist conditions and in moderate temperatures. Viable eggs have been found in water and damp sand for three weeks at 30°C, 225 days at 6°C and 32 days at 10-21°C. The eggs survive for only short periods of time if they are exposed to direct sunlight and dry conditions.

The intermediate hosts include a large number of domesticated and wild animals, particularly herbivores. Humans can also be infected. If an intermediate host ingests the eggs, the larvae are released, penetrate the intestinal wall, and are carried in blood or lymph to the target organs.



Life Cycle

The adult *Echinococcus granulosus* (sensu lato) (2—7 mm long) ❶ resides in the small intestine of the definitive host. Gravid proglottids release eggs ❷ 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 ❸ 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 ❹ 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 ❺ evaginate, attach to the intestinal mucosa ❻ , and develop into adult stages ❶ in 32 to 80 days.

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

Parasites can develop into cysts in many different organs, but they are found most often in the liver and, less frequently, the lungs. The rate of development varies with the intermediate host and species of parasite, but the cysts usually grow slowly. Their diameter generally increases from less than 1 cm to 5 cm each year. Some cysts may persist unchanged for years. Most *E. granulosus s. l.* cysts are 1-7 cm in diameter when they are discovered, but some may eventually reach 20 cm. Each fluid-filled cyst is surrounded by a fibrous wall from the host and contains two walls derived from the parasite: an outer laminated membrane and an inner membrane called the germinal layer. Brood capsules develop from the germinal membrane. Each brood capsule contains one to several invaginated heads (protoscolices) that can develop into adult tapeworms if they are ingested by the definitive host. Capsules and protoscolices either float freely in the hydatid fluid or adhere to the wall with a peduncle; the capsules and protoscolices that float freely are known as "hydatid sand." If a cyst ruptures, the hydatid sand can develop into new cysts. Some cysts are sterile; they either never produce brood capsules, or they become sterile after bacterial infection or calcification. The percentage of sterile cysts varies with the intermediate host and its susceptibility to a particular strain/ species.

In some hosts, few or no cysts of a given parasite species/ strain appear to be fertile and viable. These hosts serve as indicators for the presence of that organism, but are unimportant in perpetuating it. The importance of a host species, and the proportion of sterile cysts, seems to vary with the geographic region in some cases. For example, sheep are the primary intermediate hosts for the G1 strains in some countries, and many or most cysts in cattle are not viable. In other locations, livestock such as cattle and/or camels have a high proportion of fertile cysts and participate in the G1 parasite's life cycle. In one study, the percentage of cattle with fertile and viable G1 cysts was considerably higher among cattle in Argentina than Spain. It has not been determined whether host immune responses, management factors or parasite genetic differences are responsible for this difference.

1.6. Clinical Signs

Definitive hosts

Echinococcus spp. are usually carried asymptomatically in their definitive hosts. Large numbers of parasites may be able to cause enteritis and diarrhea, but this seems to be rare; thousands of adult parasites have been found in asymptomatic dogs and foxes.

Intermediate hosts

There is little information on natural infections with *E. granulosus s. l.* in domesticated animals. The cysts grow slowly and are usually asymptomatic until they are large enough to put pressure on adjacent tissues and organs. Livestock are often slaughtered before this occurs. If clinical signs are seen, they are those of a mass lesion and vary with the organ affected. Most cysts are found in the liver and lungs, but they may also occur in many other organs. Symptoms that have been reported occasionally in sheep include hepatic disorders with ascites and jaundice, as well as bronchopneumonia, heart failure, poor growth, weakness and lameness.

Disease in Man

Persons with cystic echinococcosis often remain asymptomatic for years until hydatid cysts containing the larval parasites grow large enough to cause discomfort, pain, nausea, and vomiting. The cysts grow over the course of several years before reaching maturity and the rate at which symptoms appear typically depends on the location of the cyst. The cysts are mainly found in the liver and lungs but can also appear in the spleen, kidneys, heart, bone, and central nervous system, including the brain and eyes. Cyst rupture is most frequently caused by trauma and may produce a host reaction manifesting as fever, urticaria, eosinophilia, and potentially anaphylactic shock, even death, as a result of the release of cystic fluid or cyst dissemination.

1.7. Diagnosis

The presence of a cyst-like mass in a person with a history of exposure to sheepdogs in an area where *E. granulosus* is endemic suggests a diagnosis of cystic echinococcosis. Imaging techniques, such as CT scans, ultrasonography, and MRIs, are used to detect cysts. After a cyst has been detected, serologic tests may be used to confirm the diagnosis.

1.8. Treatment

In the definitive host, *Echinococcus* spp. can be treated with anthelminthic drugs. Praziquantel, which is effective against both juvenile and adult *Echinococcus* parasites, is often used.
In intermediate hosts, surgery is often the treatment of choice. Long-term anthelminthic treatment may also suppress some cysts. Long term daily albendazole treatment, after surgical resection of the cyst masses, has suppressed parasite growth in some dogs with *E. multilocularis* cysts.

In man surgery was the only treatment for cystic echinococcal cysts in the past. Chemotherapy, cyst puncture, and PAIR (percutaneous aspiration, injection of chemicals and reaspiration) have been used to replace surgery as effective treatments for cystic echinococcosis. However, surgery remains the most effective treatment to remove the cyst and can lead to a complete cure. Some cysts are not causing any symptoms and are inactive; those cysts often go away without any treatment.

1.9. Prevention & Control

Cystic echinococcosis is controlled by preventing transmission of the parasite. Prevention measures include limiting the areas where dogs are allowed and preventing animals from consuming meat infected with cysts.

- Prevent dogs from feeding on the carcasses of infected sheep.

- Control stray dog populations.
- Restrict home slaughter of sheep and other livestock.
- Do not consume any food or water that may have been contaminated by fecal matter from dogs.
- Wash your hands with soap and warm water after handling dogs, and before handling food.
- Teach children the importance of washing hands to prevent infection.