What is Echinococcosis or Hydatidosis?

Echinococcosis, or Hydatid disease, is an infection caused by tapeworms of the genus *Echinococcus*, a tiny tapeworm just a few millimetres long. Five species of *Echinococcus* have been identified which infect a wide range of domestic and wild animals. Echinococcosis is a zoonosis, a disease of animals that affects humans.

Like all tapeworms the life cycle involves two animals. A carnivore is the definitive host – where the adult worms live in the intestines – and almost any mammal, including humans, can be the intermediate host - where the worms form cysts in various organs.

The disease symptoms are caused by the cysts, which are slow growing fluid-filled structures that contain the larvae and are most often located in the liver or lungs. Called hydatid cysts, for *E. granulosus*, they act like tumours that can disrupt the function of the organ where they are found, cause poor growth, reduced production of milk and meat, and rejection of organs at meat inspection. In humans the disease can be severe, occasionally fatal, and the treatment is lengthy and expensive. *Echinococcus* is benign in the intestine of the carnivorous definitive host.

Echinococcus infection is a disease listed in the OIE *Terrestrial Animal Health Code* and must be reported by Member Countries and Territories according to the OIE *Code*. 
Where is the disease found?

Of the five species, *Echinococcus granulosus* is found all over the world. *E. multilocularis* is widespread in the northern hemisphere, while two species, *E. oligarthus* and *E. vogeli* are found only in Central and South America. The fifth, *E. shiquicus*, was discovered in 2006 in the People’s Republic of China. *E. granulosus* and *E. multilocularis* are most important for zoonotic risk. In contrast the Latin American species rarely infect humans, and the zoonotic status of *E. shiquicus* is unknown.

How is the disease transmitted and spread?

The most widespread cycle exists for *E. granulosus* between dogs and sheep. When dogs are fed fresh offal or scavenge infected sheep carcasses containing cysts, they become infected, contaminate the pasture with their faeces, and sheep are re-infected as they graze. There is a similar cycle between dogs and horses, dogs and camels etc, and in wildlife, for example between wolves and moose/deer, and dingoes and kangaroos/wallabies. *E. multilocularis* is mainly transmitted within the predator-prey relationship between foxes and small mammals especially voles; cattle, sheep and pigs although sometimes exposed to infection only develop small non-viable lesions of *E. multilocularis* and are therefore not involved in transmission.

The life cycle:

Adult worms live in the small intestine of the definitive host. They reproduce releasing eggs into the environment in the faeces of the host animal. The eggs are well adapted to survive in the environment for as long as a year in cool moist conditions, but are susceptible to desiccation. Fresh eggs are sticky and may adhere to the fur of definitive hosts facilitating their spread.

The intermediate host ingests the eggs incidentally while grazing, foraging or drinking. The eggs hatch in the small intestine, become larvae which penetrate the gut wall, and are carried in the circulatory system to various organs. There the cysts, called hydatid cysts or metacestodes, are formed. The cysts, which contain larvae, either comprise fluid filled bladders, which contain larval pre-tapeworms (protoscoleces), and cause the disease cystic echinococcosis due to *E. granulosus* or alternatively, for *E. multilocularis* a multivesiculated lesion or mass containing protoscoleces that grows rapidly by exogenous budding and causes alveolar echinococcosis in rodents and other small mammals. Though slow growing in humans and long-lived animals (e.g. camels or horses) cysts of *E. granulosus* can reach a size of 10-20 centimetres, but in sheep are usually 2-6 cms.

The life cycle is completed when the cysts are ingested by a carnivore definitive host (e.g. dog, fox, or wolf), the larvae (protoscoleces) are released from the cyst into the small intestine, and develop into adult tapeworms that produce eggs which are released into the environment in the faeces of the host animal within 25-80 days depending on the species and strain of *Echinococcus*. 
What are the clinical signs of the disease?

Tapeworms in the small intestine of the definitive host cause few ill effects. In the intermediate host, the cysts gradually displace or induce fibrosis in normal tissue, and result in disease manifestation. The symptoms in humans depend on where in the body the cyst develops, and the size and numbers of cysts or metacestode mass.

In infected livestock with *E. granulosus* there can be reduced growth, decreased production of milk, meat and wool, reduced birth rate and losses due to condemnation of organs at post mortem examination. However, the cysts grow slowly so that many infected animals are slaughtered before the cysts ever cause disease problems.

There may however be multiple cysts of *E. granulosus* which can also occur in the brain, kidneys, bones, or testes causing more severe illness. Without control measures, infection rates can be very high in livestock and dogs, with associated significant incidence in humans.

E.*multilocularis* (and the other species) rarely infect cattle, sheep and pigs and when exposure occurs the cysts may not be viable.

How is the disease diagnosed?

In view of the life cycle, the best means to diagnose echinococcosis in definitive hosts is the demonstration of the adult worm in the intestine at post mortem or in the mucus after a diagnostic test (arecoline purgation), or finding the proglottids (tapeworm segments) in faeces. In wild carnivores necropsy is commonly undertaken, for example foxes in *E. multilocularis* endemic areas. There are also tests for specific antigens in faeces (coproantigens) which are highly genus specific for Echinococcus and exhibit greater sensitivity than arecoline purgation.

In intermediate hosts, diagnosis depends on the post mortem detection of the cysts particularly in the liver and lungs for *E. granulosus*. They are often detected during meat inspection, and can also be found with ultrasound examination but specificity of imaging is poor. Serological tests for cystic echinococcosis in cattle, sheep and pigs are not used routinely because of variable sensitivity and specificity. Post mortem examination of small mammals can be used to detect *E. multilocularis* cysts but prevalences are usually low, and fox prevalence data is therefore more useful.

Eggs can be sampled from soil (or faeces) for specific amplification of DNA to detect the presence of Echinococcus.

The diagnosis is performed according to the guidelines found in the OIE *Manual of Diagnostic Tests and Vaccines for Terrestrial Animals*.

What is being done to prevent or control the disease?

The best control measure is to interrupt the life cycle of the parasyte. For *E. granulosus* this can be done by:

- preventing access of dogs to livestock carcasses or slaughter wastes from farms, households, abattoirs or butchers,
- treating dogs with an anthelmintic (praziquantel) to kill the adult tapeworm,
- detecting cysts at meat inspection, thus targeting infected farms or communities,
- vaccinating sheep (or other livestock) to protect against the development of the larval stage of *E. granulosus*.
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In the laboratory, strict hygiene is necessary to prevent infection of workers.

The cycle of *E. granulosus* in wildlife is not amenable to control, but by discouraging scavenging, and implementing hygiene, the infection of domestic animals and the subsequent spread to humans can be reduced.

Control of *E. multilocularis* is much more difficult because of the wildlife cycle between foxes and rodents, but reduction in transmission has been achieved by use of praziquantel baits for foxes and dosing of owned dogs where spill-over into the dog population occurs.

**What is the public health risk associated with this disease?**

Echinococcosis is a serious zoonosis, with rates of human cystic echinococcosis infection ranging from less than 1 per 100,000 to more than 200 per 100,000 in certain rural populations where there is close contact with domestic dogs. Incidence of human alveolar echinococcosis is usually < 0.5 per 100,000 but may be >100 per 100,000 in certain communities (e.g. Tibetan herdsmen).

Laboratory workers, animal handlers, veterinarians, dog owners are all at higher risk of infection. Since the eggs are shed in the environment, they can contaminate fruits, vegetables or water, or can stick to the fur of an animal and be transferred on hands to the mouth.

In humans the cysts of *E. granulosus* usually develop in organs such as the liver or lungs, so the signs of disease are due to liver or lung deficiency. Rarely, cysts form in bones causing spontaneous fractures, or in the brain causing neurological signs. Cysts or lesions of *E. multilocularis* occur primarily in the liver and grow slowly but with eventual serious liver pathology and high risk of mortality if untreated. As well, the cysts occasionally rupture and cause severe allergic reactions in humans.

Treatment includes surgery to remove or drain cysts or liver resection, and use of long term chemotherapy with parasiticides (eg. albendazole, mebendazole) to kill larvae or prevent them from growing back after surgery.

Since the route of infection is hand to mouth, frequent hand washing constitutes an important preventative measure.
More Information?

References:

1. OIE Terrestrial Animal Health Code:
   www.oie.int/en/international-standard-setting/terrestrial-code/access-online/

2. OIE Manual of Diagnostic Tests and Vaccines for Terrestrial Animal:
   www.oie.int/en/international-standard-setting/terrestrial-manual/access-online/

3. OIE Technical Disease Card:

4. The Center for Food Security and Public Health, Iowa State University
   www.cfsph.iastate.edu/

5. Merck Veterinary Manual:

6. Atlas of Transboundary Animal Diseases Animales Transfronterizas
   P. Fernandez, W. White; Ed.: 2011

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List of Reference Laboratories:
www.oie.int/en/our-scientific-expertise/reference-laboratories/list-of-laboratories/

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www.oie.int/en/our-scientific-expertise/collaborating-centres/list-of-centres/
Key Facts

• Over four centuries BC, Hippocratus reported that hydatid cysts in humans could sometimes burst open into the abdominal cavity. Furthermore, he compared these cysts to ‘water-filled tumours’ which he observed on post-mortem examination of cattle and pigs.

• In an endemic area where cystic hydatid disease is uncontrolled the infection rate in sheep can be >50% and human prevalence rates >5%. Pastoral areas with livestock, dogs and people help maintain the transmission of E.granulosus through home slaughter and feeding of offal to dogs.

• Control programs for cystic echinococcosis can be successful but require a long period of intervention (>5-10 years) based primarily on dog-targeted control measures. The advent of a new vaccine for livestock may help reduce the time required to interrupt transmission between dogs and sheep and the risk of human exposure.