**Echinococcus spp.: Tapeworms That Pose a Danger to Both Animals and Humans – A Review**

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Species of the genus *Echinococcus* (Cestoda; Taeniidae) are minute tapeworms of carnivores. Their larvae are known as hydatids (metacestode), which proliferate asexually in various mammals. Like the majority of cestodes, *Echinococcus* spp. require two different host species to complete their life cycle. Definitive hosts harbouring the adult cestodes in the small intestine are exclusively carnivores of the Canidae and Felidae families. A wide range of mammal species including humans is susceptible to infection by the metacestode of *Echinococcus* spp., which develops in their viscera. The disease, caused by species of the genus *Echinococcus*, is called echinococcosis, and it is one of the most dangerous zoonoses in the world. The traditional species *Echinococcus granulosus* and *Echinococcus multilocularis* are agents of significant diseases due to the high number of cases and the wide geographical range. The taxonomy of the genus is controversial; in the current state of ongoing complex revisions, the agent of cystic echinococcosis *E. granulosus* sensu lato is divided into five species (*E. granulosus* sensu stricto, *E. feldii*, *E. equinus*, *E. ortleppi*, *E. canadensis*), in addition to the agents of alveolar echinococcosis (*E. multilocularis*, *E. shiquicus*) and polycystic/unicystic echinococcosis (*E. vogeli*, *E. oligarthrus*). Here we provide an overview of the current situation, which continues to develop.

*Echinococcus granulosus* s.l., *Echinococcus multilocularis*, *Echinococcus oligarthrus*, *Echinococcus shiquicus*, *Echinococcus vogeli*, echinococcosis

**INTRODUCTION**

Echinococcosis is a zoonotic infection caused by adult or larval (metacestode) stages of cestode species belonging to the genus *Echinococcus* (Romig, 2003; Eckert, Deplazes, 2004). This disease remains a significant problem worldwide (Thompson, 2008; Umhange et al., 2016). Of the three types of echinococcosis, alveolar and cystic echinococcosis are of special importance in humans due to their wide geographical distribution as well as to their medical and economic impact. Polycystic echinococcosis is less frequent and is restricted to areas of Central and South America (Eckert, Deplazes, 2004). Species of the genus *Echinococcus* require two different host species to complete their life cycle, the definitive and intermediate host (Romig, 2003). Humans are an aberrant intermediate host for the disease, developing life-threatening tissue cysts predominantly in the liver, lungs, and other organs. Infection follows accidental ingestion of eggs released in faeces of infected dogs, foxes, and wild canines. When a human is infected,
treatment is complicated and carries significant risk (Nasseri-Moghaddam et al., 2011).

The taxonomy of the genus *Echinococcus* has been a controversial issue for decades, and the outcome of recent molecular epidemiological studies reinforces the need to revise the taxonomy of this genus. Several species and subspecies were classified over the last 50 years and subsequently invalidated; however, taxonomic and genetic studies are still ongoing worldwide (Thompson, McManus, 2002; Thompson, 2008; Romig et al., 2015). For instance, Romig (2003) reported four recognized species of the genus *Echinococcus*: *E. granulosus* Batsch, 1786, *E. multilocularis* Leuckart, 1863, *E. oligarthrus* Diesing, 1863, and *E. vogeli* Rausch and Bernstein, 1972 (Fig. 1). Wang et al. (2008) reported seven species in this genus: *E. granulosus*, *E. multilocularis*, *E. oligarthrus*, *E. vogeli*, *E. ortleppi* (Lopez-Neyra and Soler, 1943), *E. equinus* Williams and Sweatman, 1963, and *E. shiquicus* Xiao et al. 2006. Moro, Schantz (2009) reported six recognised species in the genus *Echinococcus*, yet only four of them are of public concern: *E. granulosus*, *E. multilocularis*, *E. oligarthrus*, and *E. vogeli*; the zoonotic transmission potential of the remaining two species – *E. shiquicus* from Tibetian fox and *E. felidis* Ortlepp, 1937 – is unknown. Romig et al. (2015) provided an overview of the following nine species in the genus *Echinococcus* that is still in place: *E. felidis*, *E. equinus*, *E. ortleppi*, *E. canadensis* (these four species cause cystic echinococcosis and belong to the species *E. granulosus* sensu lato), *E. multilocularis*, *E. shiquicus* (agents of alveolar echinococcosis), and *E. vogeli* and *E. oligarthrus* (agents of polycystic/multicystic echinococcosis). Genetically, individual species of *E. granulosus* sensu lato (s.l.) include various strains referred to as G1–G10 (Naka et al., 2007; Huttner et al., 2008; Thompson, 2008; Naka et al., 2013; Boufana et al., 2015; Romig et al., 2015).

### Echinococcus granulosus (Batsch, 1786)

The adult *E. granulosus* (Fig. 2) is a small tapeworm that normally has three segments and a body length of 2–6 mm (Thompson, 1995). The penultimate segment is mature, and the gravid uterus is characterized by well-developed lateral sacculations. The genital pore normally opens posterior to the middle of both the mature and gravid segments (Thompson, McManus, 2001). The metacestode stage (cyst) is a fluid-filled bladder that is usually unilocular, though communicating chambers also occur (Thompson, 1995). This cyst consists of an inner germinal or nucleated layer and an acellular laminated layer of varied thickness. *E. granulosus* typically produces a single-chambered unilocular cyst. In some hosts, especially in humans, large cysts may develop, and daughter cysts may form in the primary mother cyst (Thompson, McManus, 2001). Cystic echinococcosis is caused by the metacestode stage of ten strains or genotypes of *E. granulosus* sensu lato, as is specified below. Two types of hydatid cysts can be observed in various intermediate hosts: (1) fertile cysts, in which brood capsules contain protoscoleces and (2) infertile cysts, which may not produce protoscoleces or are immature and therefore unable to sustain the life cycle of this parasite (Kamenetzky et al., 2000; Lahmar et al., 2004). Hydatid disease infects about 2–3 million humans living in endemic areas of North and South America, Europe, Africa, and Asia (Cummings et al., 2000).
The geographical expansion of *E. granulosus* s.l. is global, and the endemic foci are present on all of the inhabited continents. The highest prevalence of cystic echinococcosis in humans and animal hosts is found in the countries of the temperate zones, including several parts of Eurasia, Australia, some parts of the Americas, and northern and eastern parts of Africa (Dakkak, 2010; Ianggal et al., 2010).

**Echinococcus multilocularis** (Leuckart, 1863)

The body length of the adult tapeworm *E. multilocularis* (Fig. 4) varies between 1.2 and 4.5 mm, and usually it has four or five segments. The antepenultimate segment is characteristically mature and the genital pore is anterior to the middle of mature gravid segments; the gravid uterus is sac-like structure (Thompson, 1995; Thompson, McManus, 2001). The larval stage (metacestode) of *E. multilocularis* is a complex structure and develops quite differently in comparison to *E. granulosus*. The structure consists of small vesicles containing protoscoleces, which are produced by a germinal layer. The mass of larval stage usually contains a semisolid matrix rather than fluid. Proliferation...
occurs endogenously and exogenously (Déplazes, Eckert, 2001; Thompson, McManus, 2001).

Human alveolar echinococcosis is a zoonosis of public health significance as the parasite in a human host behaves like a malignant tumour, predominantly in the liver. Lethality rates had been nearly 100% in untreated patients before the 1970s, when modern treatment methods were not yet established (Eckert, 1998). This disease was first reported in 1852 by Ludwig Buhl and in 1855 by Rudolf Virchow (Tappe, Froesch, 2007). Alveolar echinococcosis is less common in humans. In comparison to cystic echinococcosis, it is more pathogenic, more difficult to treat, and has a higher rate of mortality (Bristow et al., 2012). In Europe, human infections are rare since the tapeworm develops mainly during sylvatic cycle. However, reports show that the parasite is already spreading in Europe (Sreter et al., 2003). Diagnosing human alveolar echinococcosis is based mainly on identifying the parasite lesions using imaging methods (ultrasound examination, computed tomography etc.) and through the detection of specific serum antibodies. Sasaki, Sako (2017) reported that putative serine protease inhibitors (serpins) can influence the mechanisms underlying infiltration into host tissue, as well as immune evasion by larval stage parasites. Treatment is still difficult and complete recovery is only achieved when alveolar echinococcosis is diagnosed early and the entire parasite lesion can be removed surgically; even in such cases, at least two years of chemotherapy is recommended by the WHO (Craig et al., 1996). Symptoms will only occur after long-term incubation (Eckert et al., 2001).

*E. multilocularis* typically appears during the sylvatic cycle, which includes foxes (predominantly those of the genus *Vulpes* and *Alopex*) as definitive hosts and small mammals, predominantly rodents, as intermediate hosts. In some endemic areas, other species of wild carnivores, such as coyotes, wolves, and raccoon dogs, serve as definitive hosts. A synanthropic cycle is known to exist in various epidemiological situations with domestic dogs and cats as definitive hosts, which become infected from rodents originating from the sylvatic cycle (Thompson, 1995; Eckert, 1998; Machnicka-Rowinska et al., 2002). The survival time of this parasite in definitive hosts is estimated to be about 5 months (Eckert, 1998). The larval stage in intermediate hosts and aberrant hosts (including humans) is characterized by the formation of multivesicular cysts and tumour like proliferation in the liver or other organs. This cestode is known as a causative agent of dangerous alveolar echinococcosis in humans (Miterpakova, Dubinsky, 2011). In addition to foxes, dogs and cats can occasionally serve as definitive hosts of the parasite (Dyachenko et al., 2008). Proglotids and eggs are shed in the faeces of the definitive host and can be ingested by rodents such as the common vole (*Microtus arvalis* Pallas, 1778), the water vole (*Clethrionomys glareolus* Schreber, 1780) (Eckert, Déplazes, 2004). In Europe the fox population is increasing alongside the prevalence of *E. multilocularis* infection in foxes. Barring historically endemic areas, more and more regions previously considered to be disease-free are now experiencing a higher prevalence of infection (Torgerson, Budke, 2003). Maas et al. (2016) described a new method based on a magnetic capture based DNA extraction, followed by qPCR developed for the detection of the zoonotic parasite *E. multilocularis* in definitive hosts. This method has the advantage of not requiring hazardous chemicals such as phenol-chloroform. Maksimov et al. (2017) demonstrated that the performance of commercial DNA extraction kits for processing *E. multilocularis* eggs in fox faecal samples can vary widely, but certain combinations of DNA extraction kits and PCR protocols can analyze samples containing even a small number of parasite eggs and yield acceptable diagnostic sensitivity as well as diagnostic specificity.

The geographical expansion of *E. multilocularis* is confined to palearctic regions of the northern hemisphere, extending from central Europe throughout northern and central Eurasia to the Far East, including Japan, and to North America (Alaska, Canada, northern and central USA). Prior to the end of the 1980s, *E. multilocularis* endemic areas in Europe were limited to eastern France, Switzerland, southern Germany, and western Austria (Schantz et al., 1995; Eckert et al., 2000, 2001). The occurrence of *E. multilocularis* in Western and Central Europe includes regions in Austria, Switzerland, France, Germany, Liechtenstein, Luxembourg, Belgium, the Netherlands, Poland, the Czech Republic, the Slovak Republic, Denmark, and the Norwegian Islands of Svalbard (Schantz et al., 1995; Pavelsek et al., 1997; Dubinsky et al., 1999; Kolarova, 1999; Romig et al., 1999; van der Giessen et al., 1999; Eckert et al., 2000; Vervaekte et al., 2003). The presence of this tapeworm in their definitive or intermediate hosts

Fig. 4. Adult *Echinococcus multilocularis* (Photo: Adéla Brožová)

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196

**SCIENTIA AGRICULTURAE BOHEMICA, 48, 2017 (4): 193–201**
has not been evidenced in five European countries: Finland, mainland Norway, Great Britain, Ireland, and Malta (Miterpakova, Dubinsky, 2011; Learmount et al., 2012; Murphy et al., 2012). Using a program of anthelmintic bait distribution, Budgey et al. (2017) developed a model to predict the probability of successful eliminating a focal outbreak of E. multilocularis.

Echinococcus shiquicus (Xiao et al., 2006)

The cestode E. shiquicus was found exclusively in Tibetan foxes Vulpes ferrilata Hodgson, 1842 in China (Qinghai – Tibet plateau region) in 2005. This species has two types of adult tapeworms (Fig. 5) containing gravid segments. The first type consists of premature and gravid segments. The second type consists of immature, mature, and gravid segments. Total body length ranges between 1.3 and 1.7 mm. The larval stage (metacestode) has only been found in plateau pikas Ochotona curzoniae (Hodgson, 1858). DNA sequencing revealed that five (5%) of 101 pikas have hydatid cysts in their livers. A pulmonary hydatid cyst was also found in one pika. Most of the larval forms were unilocular cysts approximately 10 mm in diameter, and one appeared an oligovesicular form. E. shiquicus infections have not been detected in humans (Xiao et al., 2005, 2006). Ma et al. (2012) collected 70 Echinococcus samples (larval and adult) from sheep, yak, pikas, voles, dogs, and humans in the Qinghai-Tibet Plateau. The majority of Echinococcus samples were identified as E. multilocularis and E. granulosus s.l. while E. shiquicus was determined only in pikas; it is suggested that E. shiquicus had restricted intermediate host specificity.

Echinococcus vogeli Rausch and Bernstein, 1972

E. vogeli (Fig. 6) has the longest strobila of any member of the genus Echinococcus, indicating a maximum length of 12 mm in experimentally infected dogs (D’Alessandro, Rausch, 2008). The adult tapeworm length ranges between 3.9 and 5.6 mm, and usually has three segments. The penultimate segment is mature and the genital pore is situated posterior to the middle of both the mature and gravid segment. The gravid uterus has no lateral branches or sacculations, and is relatively long and tubular in form (Thompson, McManus, 2001).

The life cycle of the South American species E. vogeli involves the bush dog (Speothos venaticus Lund, 1842) as the natural definitive host, and pacas (Cuniculus pacu Linnaeus, 1766) and agoutis (Dasyprocta spp.) as intermediate hosts. Intestinal stages of E. vogeli evidently do not develop in cats or in other species of felids (D’Alessandro et al., 1981; D’Alessandro, Rausch, 2008). E. vogeli is a causative agent of polycystic echinococcosis and this disease has been reported in humans in four countries: Colombia, Ecuador, Panama, and Venezuela (D’Alessandro et al., 1979; D’Alessandro, Rausch, 2008).

Echinococcus oligarthrus Diesing, 1863

It was noted that the frequently used species name E. oligarthrus is nomenclatorically incorrect (see Huttner, Romig, 2009; Nakao et al., 2013) as E. oligarthra is the correct term. However, we have chosen to keep the most commonly used form. The strobila is 2–3 mm long with three proglottids (Fig. 7). The rostellar hooks are similar to those of E. vogeli in length but differ in shape (Rausch, Bernstein, 1972). The genital pore is anterior to the middle in mature segments and approximately at the middle in gravid segments. The gravid uterus is sac-like (Thompson, McManus, 2001).
Neotropical felids serve as definitive hosts of *E. oligarthrus*. Out of ten species indigenous to Central and South America, the following six species have been identified as definitive hosts of this parasite: the pampas cat (*F. colocolo* Molina, 1782), Geoffroy’s cat (*F. geoffroyi* d’Orbigny and Gervais, 1844), the ocelot (*F. pardalis* Linnaeus, 1758), the jaguarundi (*F. yagouaroundi* Geoffroy Saint-Hilaire, 1803), the jaguar (*P. onca* Linnaeus, 1758), and the puma (*P. concolor* Linnaeus, 1771). Moreover, *E. oligarthrus* was found in northern Mexico in one bobcat (*L. rufus* Schreiber, 1771), and therefore on agoutis (*Dasyprocta* spp. Allen, 1899), and pacas (*Cuniculus pacu* Linnaeus, 1766) are known as definitive hosts. Diseases caused by *E. oligarthrus* are known as unicyclic or cystic echinococcosis (D’Alessandro, Rausch, 2008). Basset et al. (1998) observed retroocular cystic tumor in the left eye, which caused this disease in a six-year-old boy. The child had a uniocular cystic echinococcosis (D’Alessandro, Rausch, 2008). Basset et al. (1998) observed retroocular cystic tumor in the left eye, which caused this disease in a six-year-old boy. The child had a uniocular cystic echinococcosis (D’Alessandro, Rausch, 2008). Basset et al. (1998) observed retroocular cystic tumor in the left eye, which caused this disease in a six-year-old boy. The child had a uniocular cystic echinococcosis (D’Alessandro, Rausch, 2008)

CONCLUSION

The taxonomy of *Echinococcus* is not yet to be completely clarified. Diseases caused by agents of this genus remain among the most dangerous zoonoses in the world.

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