



## *Echinococcus* across the north: Current knowledge, future challenges☆



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### ABSTRACT

Zoonotic *Echinococcus* spp. cestodes are present in almost all circumpolar nations, and have historically posed a risk to health of indigenous as well as other northern residents. However, surveillance data on both alveolar (AE) and cystic (CE) echinococcosis remains incomplete throughout the circumpolar region: Russia, Fennoscandia, Iceland, Greenland, Canada and Alaska (USA). Prevalence of *Echinococcus* spp. varies considerably in definitive canid hosts, animal intermediate hosts and accidental hosts like humans. Yet despite the high prevalence reported in canids in some geographic locations, human AE and CE are much less common than in endemic Asian and central European countries. This paper explores knowledge gaps and future challenges posed by *Echinococcus* spp. in eight circumpolar countries, a region where rapid environmental and social change are rewriting the boundaries, transmission, and impact of many pathogens, including zoonotic *Echinococcus* spp.

Genotypes G6, G8 and G10 of *Echinococcus canadensis* are causative agents of human CE and have been identified in sylvatic (wild animal) and synanthropic (ecological association with humans) cervid-canine life cycles in the following northern regions: Alaska and northern Canada - G8 and G10; northern Russia - G6, G8, G10; and Fennoscandia - G10 in Finland - with no recent reports from Norway or Sweden. *Echinococcus multilocularis*, which causes AE, has been identified in a sylvatic arvicoline rodent-canine lifecycle in Alaska, Canada, Russia, Sweden and Svalbard (Norway). Asian, Mongolian, European and North American strains of *E. multilocularis* are found in Russia, with the North American N1 strain predominating in the north. The N1 strain is also found in Alaska, as well as Svalbard, whilst Asian strains have been identified in western Alaska. Central North American (N2) strain and European-type strains of *E. multilocularis* are present in Canada. Typing of the strain in Sweden is still pending. Individual human cases of AE with N2 and European-type strains are reported in North America, as well as multiple cases with Asian strains in Russia and historically on St Lawrence Island, Alaska (although genotyping of human cases was not available at the time). *Echinococcus* spp. have not been detected in Greenland and have been eliminated from Iceland.

**Abbreviations:** AE, alveolar echinococcosis; CE, cystic echinococcosis; DALY, Disability Adjusted Life Years; DH, definitive host; IH, intermediate host; NCR, Northern Central Region; NTZ, Northern Tundra Zone; OIE, World Organisation for Animal Health; SLI, St. Lawrence Island-Alaska; USA, United States of America  
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The predominance of *E. multilocularis* N1 strain and *E. canadensis* genotypes, in regions with high prevalence in definitive hosts yet low incidence of human AE and CE, suggests that these genotypes have lower zoonotic potential and pathogenicity than European and Asian strains of *E. multilocularis* and livestock genotypes of *E. granulosus sensu stricto*. The continued monitoring of the emergence of *Echinococcus* genotypes within definitive and intermediate hosts, as well as people, is needed to assess the impact on public health risk, since the introduction of other genotypes could have serious repercussions. Lastly, determining risk factors and source attribution for human cases, including the possibility of food and waterborne transmission and the likelihood of autochthonous transmission, remain challenges.

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## 1. Introduction

Eight countries transcend the Arctic Circle currently at 66°33'46.2", and/or the 10° July isotherm, the meteorological definition of Arctic. These are Russia, Finland, Sweden, Norway, Iceland, Greenland, Canada and the State of Alaska (United States of America, USA). These very diverse countries range from having large metropolitan areas to remote small populations, some with indigenous people practicing a "traditional" lifestyle. Traditional activities such as hunting, fishing, and gathering (e.g. berries, mushrooms) are important culturally, economically, and nutritionally in indigenous peoples in all eight countries, and are also popular in non-indigenous ethnic groups. Untreated surface water is often used as a potable water source in northern communities in these countries. Some remote communities also have limited access to modern health care, such as medical imaging and sensitive and specific diagnostic tests, or veterinary services (Hotez, 2010). These multiple factors combine to contribute to a higher risk of exposure to zoonotic parasites, and more severe health consequences if infected. Tuberculosis screening campaigns in reindeer herders/indigenous people during the mid-20th century incidentally found many cystic echinococcosis (CE) cases in Fennoscandia, northern Canada and Alaska. Alveolar echinococcosis (AE) has been observed in people in western Alaska and Russia. In this paper we explore the two species of the *Echinococcus* that have been identified in northern wildlife and in human populations in these seven countries and Alaska as a whole, not just restricted to the regions north of the Arctic Circle.

Both CE and AE have recently been included on the WHO Neglected Tropical disease list. Echinococcosis has a global estimated disease burden of  $2\text{--}5 \times 10^7$  DALY (Disability Adjusted Life Years) highlighting the considerable medical, social and economic impacts on animal and human populations worldwide (Torgerson and Macpherson, 2011). This estimate includes livestock related as well as sylvatic strains of echinococcosis. We discuss current knowledge and knowledge gaps for future exploration with regard to the life cycles, genotypes, animal hosts, and human cases of *Echinococcus canadensis* (G6, G8, G10) one of the causative agents for cystic echinococcosis (CE) and *Echinococcus multilocularis*, the causative agent of alveolar echinococcosis (AE) in these northern countries.

## 2. Echinococcus canadensis

### 2.1. Distribution, hosts and genotypes

This parasite has a sylvatic and semi-synanthropic lifecycle involving wild cervids as the intermediate host, and canids (dogs (*Canis canis*), coyotes (*Canis latrans*) and wolves (*Canis lupus*)) as the definitive host (Rausch, 2003). Humans serve as accidental hosts upon ingestion of eggs from patent infections in canid hosts. The taxonomy of this group of parasites has been much debated over the years (Nakao et al., 2007, 2009, 2013; Romig et al., 2015). Ten genotypes are recognised and the group is roughly divided into *Echinococcus granulosus* sensu stricto (ss. G1–G3) and sensu lato (sl. G1–G10). The current commonly accepted phylogenetic division includes five species: *E. granulosus* ss. (G1–3), *E. equinus* (G4), *E. ortleppi* (G5), *E. canadensis* (G6, G7, G8, G10) and *E. felidis* (Romig et al., 2015). *Echinococcus canadensis* has been identified during the past 20 years in intermediate (IH) and definitive hosts (DH) in five of the eight countries: Russia, Finland, Sweden, Canada and Alaska (USA). Three genotypes have been recognised in this region: G6, G8 and G10 (Table 1, Fig. 1).

**Table 1**

The different *Echinococcus canadensis* genotypes and *E. multilocularis* strains (Nakao et al., 2007, 2009, 2013; Romig et al., 2015) identified in the circumpolar North, host species involved in circumpolar countries, as well as reports of human cystic echinococcosis (CE), and human alveolar echinococcosis (AE) in the region.

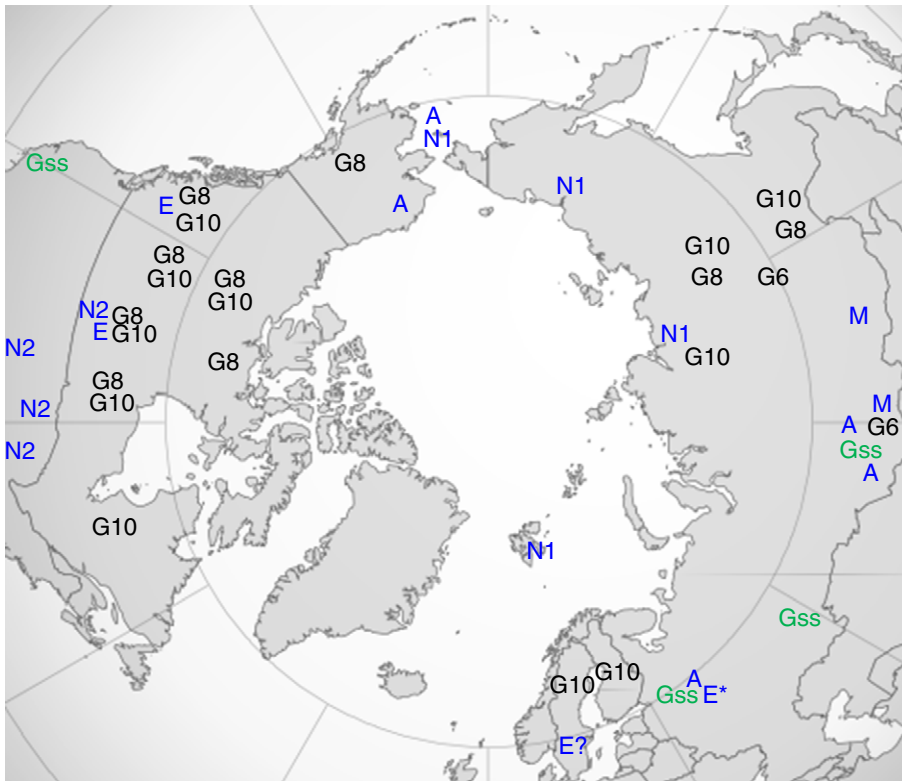
Species	Genotype/strain	Country	Definitive hosts	Intermediate hosts	Isolated from Human CE/AE cases	References	
<i>Echinococcus canadensis</i>	G6	Russia	Wolves	Reindeer	Yes (Russia)	Bowles et al. (1994); Bryan et al. (2012); Hämäläinen et al. (2015); Hirvelä-Koski et al. (2003); Konyaev et al. (2013); Lavikainen et al. (2006); McManus et al. (2002); Oksanen and Lavikainen (2015); Rausch (2003); Schurer et al. (2013, 2014a, 2016); Thompson et al. (2006).	
	G8	Russia Canada Alaska	Wolves, Coyotes <sup>a</sup>	Moose Elk Muskox	Yes (Alaska)		
	G10	Russia Finland Sweden Canada	Wolves, Coyotes <sup>a</sup> Domestic dog	Moose Reindeer/Caribou Elk	Yes (Russia, 1 case, Finland)		
<i>Echinococcus multilocularis</i>	North American	N1	Russia	Foxes	<i>Microtus</i> spp. <i>Myodes</i> spp. Shrews	No	Catalano et al. (2012); Christiansen et al. (2015); Gesy et al. (2013); Gesy et al. (2014); Gesy and Jenkins (2015); Henttonen et al. (2001); Holt et al. (2005); Jenkins et al. (2012, 2013); Kirk (2011); Knapp et al. (2012); Konyaev et al. (2013); Liccioli et al. (2014); Massolo et al. (2014, 2015); Miller et al. (2016); Nakao et al. (2009); Osterman Lind et al. (2011); Peregrine et al. (2012); Schurer et al. (2014a, 2016); Skelding et al. (2014); Yamasaki et al. (2008).
		N2	North Central Region (Canada, USA)	Foxes Coyotes Wolves <sup>a</sup>	<i>Peromyscus</i> sp. <i>Microtus</i> spp. <i>Myodes</i> spp.	Yes (1 case, USA)	
	Asian	Russia Alaska	Foxes Wolves	<i>Microtus</i> spp. <i>Myodes</i> sp. Shrews	Yes (Alaska, Russia)		
		Mongolian	Russia	Coyotes	Lake Baikal voles	Yes <sup>b</sup>	
	European	Russia <sup>c</sup> Canada			Senegalese bushbaby <sup>d</sup> Domestic dogs <sup>d</sup> Goeldi's monkey <sup>d</sup> ( <i>Callimico goeldii</i> )	Yes (1 case, Canada)	
		To be determined	Sweden	Red foxes	<i>Microtus</i> sp. <i>Arvicola</i> sp.		

<sup>a</sup> Identified in these hosts in Canada but genotyping information not available.

<sup>b</sup> Isolated from human cases in other countries but no autochthonous reports from countries in the circumpolar North.

<sup>c</sup> Considered to be an imported case.

<sup>d</sup> These species are not normally involved as intermediate hosts (IMH) in the lifecycle. The sylvatic IMH for the European strain of *E. multilocularis* in Canada has yet to be identified.



**Fig. 1.** A map of the circumpolar region ([www.wikipedia.com](http://www.wikipedia.com)) with the 60°N latitude line shown. The map shows the approximate geographic origin where different genotypes of *Echinococcus canadensis* (G6, G8, G10) and *E. multilocularis* (North American (N1, N2), Asian (A), European (E) and Mongolian (M)) have been isolated from definitive or intermediate hosts, or from people. Also shown are reports of *E. granulosus sensu stricto* (G1–G3). In Sweden the genotype has yet to be determined (indicated by ?) and in Russia the European genotype was thought to be imported (indicated by \*).

### 2.1.1. Russia

*Echinococcus granulosus* ss. has been identified in humans, domestic sheep and a cat whilst G6 has been identified in humans, wolves and reindeer (*Rangifer tarandus*), as has G10 which in addition to these hosts has also been identified in a moose (Konyaev et al., 2012, 2013). G8 has only been identified in moose (*Alces alces*) and wolves. In Yakutia, it is thought that spill over from the sylvatic cycle into domestic dogs is responsible for human CE given the absence of sheep in the region, which would act as IH for *E. granulosus* ss. (Konyaev et al., 2013).

### 2.1.2. Fennoscandia

*Echinococcus granulosus* ss. has been reported in human patients in all three countries, generally in immigrants or returning travelers (Hämäläinen et al., 2015; Norwegian Public Health Institute, 2016; Public Health Agency of Sweden, 2016). *Echinococcus canadensis* (G10) has been detected in a sylvatic wolf-moose lifecycle in Eastern Finland as well as in reindeer (Hirvelä-Koski et al., 2003; Lavikainen et al., 2006) and more recently in a domestic dog (Hämäläinen et al., 2015). To date only G10 has been identified in this region. *Echinococcus canadensis* (G10) has also been identified in historical samples from Sweden (Lavikainen et al., 2006) in reindeer but has yet to be confirmed in Norway despite historic evidence of a reindeer-herder dog cycle present in the Finnmark region (Heier et al., 2014; Oksanen and Lavikainen, 2015). During the 1950's it was estimated that up to 10% of the reindeer in Kautokeino, northern Norway were infected. Sporadic cases have been reported since then, based on pathological findings in reindeer at the abattoir (Heier et al., 2014) but cysts were not genotyped. It is thought that the decline in use of herder dogs, improved meat control restricting feeding dogs with viscera, as well as canine deworming have contributed to the decline in human and animal cases in this region.

### 2.1.3. Iceland

This country is currently considered free from *E. granulosus* sl. having eliminated it in sheep, cattle and dogs during the second half of the 20th-century (Skirnisson et al., 2003). The genotype involved in the historical cases was not determined.

### 2.1.4. Greenland

This country is considered free from *E. granulosus* sl., as no autochthonous human or animal cases have been observed (Eckert et al., 2001; Rausch, 2003).

### 2.1.5. North America

In northern Canada and Alaska, *E. canadensis* is most commonly maintained by wolf-moose and wolf-caribou host assemblages. Historically, domestic dogs replaced wolves as primary definitive hosts in agricultural regions and in areas where sled-dogs were common (Rausch, 2003). Recent surveys for *E. granulosus* sl. in dogs are very limited; however, infection levels are thought to be low due to the decline of sled-dogs, the availability of effective anthelmintics (praziquantel), and public health education efforts to dissuade owners from feeding cervid viscera to dogs (Lantis, 1980).

Two genotypes are present in Canada: G8 and G10. Molecular characterisation of hydatid cysts using PCR to target mitochondrial genes (cox1, NAD1) has identified G8 in elk (*Cervus canadensis*) (Alberta) and muskoxen (*Ovibos moschatus*, Nunavut), and G10 in elk, moose and caribou (*Rangifer tarandus*) (Alberta, Saskatchewan, Manitoba, Quebec) (Schurer et al., 2013; Thompson et al., 2006). Studies of eggs and adult cestodes collected from wolves and characterised at the same gene targets as cervids, provided evidence for sympatric distributions of G8 and G10 in British Columbia, Saskatchewan, Manitoba, and the Northwest Territories (Bryan et al., 2012; Schurer et al., 2016). G10 is also present in wolves from Alberta (Thompson et al., 2006). No molecular data is available for CE in coyotes in Canada, and information on dogs is also limited. Infected coyotes were previously observed in Alberta, Manitoba, and Ontario (Freeman et al., 1961; Holmes and Podesta, 1968; Samuel et al., 1978), but recent surveys have not detected the parasite. *Echinococcus granulosus* sl. was observed in faecal samples collected from shelter dogs in British Columbia, Alberta, and Ontario (Villeneuve et al., 2015), and G10 was reported in dogs from Saskatchewan (Himsworth et al., 2010). Overall, G10 appears to be more common than G8 in Canada, but additional data is required from central and eastern Canada to determine if this trend is consistent across the country (Schurer et al., 2013, 2016). In Alaska, hydatid cysts characterised at multiple loci demonstrated G8 in a human and a moose (McManus et al., 2002).

## 2.2. Surveillance activities and human CE

### 2.2.1. General common to all regions

Surveillance programs for *E. canadensis* tend to be limited and based upon passive surveillance during meat inspection (cervid hosts) or studies investigating cervid health or parasitism in canid hosts. Echinococcosis is notifiable in human patients in Fennoscandia and Alaska but not in Canada (at the national level) or Russia. Echinococcosis is currently only nationally notifiable in animals in Fennoscandia. The World Organisation for Animal Health (OIE) however requires the veterinary authorities of member countries (including Canada, Russia, United States of America and the Fennoscandian countries) to provide reports on *Echinococcus* (OIE listed disease) if there is a first occurrence, re-occurrence, new strain, change in distribution or host species (OIE, 2015). Genotyping is not routinely carried out in most of the countries and the diagnosis often based on macroscopic pathological changes identified during diagnostic imaging or surgery, serological diagnosis or confirmatory microscopy of cystic fluid (Norwegian Public Health Institute, 2016; Public Health Agency of Canada, 2014; Public Health Agency of Sweden, 2013).

### 2.2.2. Russia

The number of human echinococcosis cases increased substantially after the dissolution of the Soviet Union. *Echinococcus granulosus* ss. and sl. are considered to be widespread in Russia (Konyaev et al., 2013; Nikulina et al., 2003). Earlier studies have indicated regional variations in *Echinococcus* spp. seroprevalence in different human populations from 2.3%–12.3% (Martynenko et al., 1984; Vol'fson, 1968). The average incidence of echinococcosis, in 2000–2011, varied from 0.04 to 11.55 per 100 000 across the Russian Arctic, Siberia and the Far East (Dudarev et al., 2013) but the authors stressed that since sampling was not evenly distributed regional incidence results should be interpreted with caution. More recent data is lacking as is information regarding incidence and prevalence in animal hosts. Recognised risk factors for seropositivity were being involved in reindeer herding and hunting (Vol'fson, 1968).

### 2.2.3. Fennoscandia

The prevalence of infection in reindeer in Finland has been less than 0.1% since the 1990's with the majority of cases detected in the eastern part (Eastern Lapland and North-Eastern Ostrobothnia) of the reindeer husbandry area (Hirvelä-Koski et al., 2003; Raulo et al., 2012). The number of cases has been decreasing since 2004. There are occasional reports from moose in eastern and north-eastern Finland with prevalence in the endemic area recorded between 1 and 2%, more than a decade ago. Selective sampling diagnosed CE in 15% of the 46 moose sampled from northern Karelia in 2009 (EVIRA, 2010, 2011) but it was not found in animals outside the endemic region. The prevalence in the definitive host remains low in Finland. Oksanen and Lavikainen (2015) reported that *E. canadensis* prevalence in wolves in Finland, during 2000–2010, was 10%. The parasite has not been detected in wildlife in Sweden or Norway since 1997 and 2003 respectively (Heier et al., 2014; Lavikainen et al., 2006; SVA, 2015a).

During the early to mid-twentieth century echinococcosis was endemic in northern Norway (Heier et al., 2014) and Sweden (Söderhjelm, 1969) as well as being sporadically reported in Finland. Cases continue to be reported annually throughout the region, but virtually all are thought to have been acquired abroad. The disease is notifiable in Finland, Sweden (since 2004) and Norway (since 2003). Records however do not differentiate between CE and AE, nor is genotyping a requirement for diagnosis. Sweden reports the highest annual number of echinococcosis cases of the three countries, ranging between 15–30 per annum with the average incidence during the last decade (2006–2015) of 0.207/100 000 per inhabitants per annum (Public Health Agency of Sweden, 2016). Nearly all the cases have been acquired abroad with only a small percentage of unknown origin (Norwegian Public Health Institute, 2016; Public Health Agency of Sweden, 2016; unpublished data Helsinki University Hospital Laboratory (HUSLAB)). Some of these human CE cases have identified *E. granulosus* ss. by genotyping (Hämäläinen et al., 2015;

Norwegian Public Health Institute, 2016; Public Health Agency of Sweden, 2016) and a case of G6 was identified in an immigrant (A. Lavikainen; unpublished archival specimen from 1998). A single autochthonous CE case caused by G10 was recently confirmed by genotyping in Finland (Hämäläinen et al., 2015). Before this recent case, the most recent autochthonous cases were diagnosed in 1963 in Northern Finland, in the late 1960's in Sweden and 1977 in Norway (Heier et al., 2014; Lavikainen, 2005; SVA, 2015a).

#### 2.2.4. Iceland

*Echinococcus granulosus* sl. was endemic in Iceland until the second half of the 20th century. Current evidence indicates that the parasite has been eliminated with no new cases reported since 1979 (Skírnisson et al., 2003).

Harald Krabbe (1865) detected *E. granulosus* sl. adults in 28% of 100 dogs from various parts of Iceland in 1863. A century later, during 1950–1960, examinations of 200 dogs did not find this tapeworm, nor was it found in 80 Arctic foxes (*Vulpes lagopus*) caught between 1960–1975 (Pálsson, 1984), and recently a further 50 foxes sampled in the western part of the island, in the late 1980's also tested negative (Skírnisson et al., 1993).

Surveys and reports from meat inspectors in slaughterhouses indicate that CE in sheep and cattle was first brought under control in the 1940's (Pálsson, 1984). Previously, the maintenance of the life cycle of *E. granulosus* sl. seems mainly to have been restricted to the eastern part of the country as all the 62 CE cases reported from sheep in Iceland during 1953–1979 originated from 8 or 9 farms in the Múlasýslur area (Pálsson, 1984). In sheep the latest confirmed CE case was reported from a farm in Stöðvarfjörður, East Iceland in 1979 whilst in cattle the last CE case was reported in 1961 (Pálsson, 1984). Earlier in the 20th century sporadic CE cases had been reported in pigs (Pálsson, 1984). CE has never been reported from reindeer or horses.

The infection prevalence in humans born during 1861–1870 was estimated to be 22%, based on autopsy reports from 1932 to 1982. During the late 19th century public education programs, reduction of the dog population, and a ban on feeding raw offal to dogs, complimented later in the 20th century by prohibiting the import of dogs from other countries, meat inspection and targeted intervention on infected farms, successfully reduced this high prevalence (Dungal, 1957; Ólafsson, 1979; Pálsson, 1984). Autopsies of humans born in Iceland in the 20th century revealed only eight CE cases, indicating that new infections were uncommon by the end of the 19th century (Pálsson, 1984). Locally, however, the cestode maintained its life cycle for some decades. During 1984–1988 four cases of hydatid disease were reported and treated in the Akureyri Regional Hospital. The youngest patient was believed to have acquired the disease in the late 1950's; the other patients contracted the infections in the earlier half of the century (Arinbjarnar, 1989).

#### 2.2.5. Greenland

There are no definitive records of *E. granulosus* sl. in Greenland animals. One survey of 423 wolves reported taeniid eggs in 5% of faecal samples; however, molecular methods were not used to identify genera or species (Marquard-Petersen, 1997).

#### 2.2.6. North America

G8 and G10 are the predominant genotypes present in circumpolar North America, and these occur in sympatric distribution. The livestock strain (*E. granulosus* ss.; G1) is present in the contiguous United States, but not in Canada or Alaska. Definitive hosts for the sylvatic strains include wolves, coyotes and dogs. Most recently, molecular analysis of *Echinococcus* cestodes in wolves from western Canada found G8 and G10 in 6% and 24% of 191 animals, respectively. Co-infections between G8–G10 (5%) and G10-*E. multilocularis* (6%) were observed in multiple locations (Schurer et al., 2016). The overall prevalence of *E. granulosus* sl. is similar to previous reports showing the parasite in 20–24% of wolves in the Ontario, and the Yukon and Northwest Territories, but lower than the 72% prevalence reported in Alberta (Freeman et al., 1961; Holmes and Podesta, 1968; Marquard-Petersen, 1997). These older surveys suggested that *E. granulosus* sl. was less prevalent in coyotes (0.5–9%) than in wolves (Freeman et al., 1961; Holmes and Podesta, 1968; Samuel et al., 1978), and recent negative findings in coyote surveys indicate that this may still be true (Bridger et al., 2009; Catalano et al., 2012; Thompson et al., 2009). Surveillance for this parasite in dogs is limited to four surveys where taeniid eggs collected from faecal samples were characterised using molecular methods. These studies found *E. canadensis* G10 in one of three communities in rural Saskatchewan (6% of 155 faecal samples), and *E. granulosus* sl. in British Columbia, Alberta, and Ontario (Himsworth et al., 2010; Schurer et al., 2014b, 2015a; Villeneuve et al., 2015). In Alaska, *E. granulosus* sl. in canids was last estimated in the 1960s, and reported infection of 30% in wolves (N = 200) and 4–22% in sled dogs (Rausch, 1960; Rausch and Williamson, 1959).

Intermediate hosts for *E. canadensis* most commonly include moose, caribou, elk, and deer (Schurer et al., 2013; Sweatman and Williams, 1963), although sporadic infections are observed in mountain goats, muskoxen, and American bison (Rausch and Williamson, 1959; Sweatman and Williams, 1963). Over the last 50 years, post-mortem surveys of cervid hosts (N > 100) suggest that prevalence levels range between 42–47% in moose, and 1–21% in caribou, 11–38% in elk, and 0–0.3% in white tailed deer (Schurer et al., 2013). Surveys in Alaska are also limited to older data, which report infection levels of 20% (N = 124) in moose, and 0–6% of caribou (Rausch, 1959; Sweatman, 1952). Infected moose appear to experience higher levels of predation by wolves and human hunters than non-infected animals possibly due to compromised lung function (Joly and Messier, 2004; Rau and Caron, 1979). Cystic echinococcosis in animals is reportable to the OIE but is not notifiable nationally in Canada and the United States, with one exception: in Alaska immediate reporting is required (State of Alaska, 2003).

Human CE cases are not nationally reportable in North America, but they are reportable at state level in Alaska (USA) and in the Northwest Territories (Canada). In Canada, disease burden has been estimated by examining national hospitalisation records, sero-surveillance studies in northern and remote communities, and from case reports. Cystic echinococcosis was first reported in Canada in 1883 (Cameron, 1960). Up until the mid-twentieth century, the disease was most commonly diagnosed in immigrants

from countries that were endemic for the pastoral biotypes (e.g. Ireland, Italy, and Germany) (Cameron, 1960; Finlayson and Fergus, 1963). Public health initiatives to control CE abroad also decreased the prevalence in immigrants arriving in Canada, so that in the latter twentieth century, CE was more commonly diagnosed as an accidental finding during TB screening. Today, CE is considered rare, with only 0.072–0.14 cases per 100,000 people diagnosed per year (Gilbert et al., 2010; Schurer et al., 2015b). Human cases are widely distributed across the country; however, northern populations experience higher levels of infection (Gilbert et al., 2010; Schurer et al., 2015b). Sero-surveillance in northern Indigenous communities indicates that exposure levels vary by location (0–48%, Jenkins et al., 2013). To date, no cysts from Canadian patients with CE have been genotyped.

Historic records indicate that approximately 193 human cases of CE were diagnosed in Alaska prior to 1980 (Hueffer et al., 2013). Similar to northern Canada, CE was over-represented in indigenous populations and was associated with sled-dogs, which were fed discarded viscera. Rates of CE increased as Indigenous communities transitioned from nomadic to sedentary lifestyles, because new town sites became quickly contaminated with dog excrement (Deplazes and Eckert, 2001). Today, CE is considered rare, and only 8 cases have been reported since 1990 (Hueffer et al., 2013). This rapid decline has been attributed to the replacement of sled-dogs with motorised transport, and public health programs to increase awareness of the necessity of canine deworming (Lantis, 1980; Rausch et al., 1990a, 1990b). Molecular data is limited to a single study, which reported G8 in an Alaskan patient. Case reviews of patients in both Alaska and northern Canada suggest that the sylvatic biotype is less virulent than pastoral strains, and that a non-surgical approach (watch and wait) is often warranted for cervid strains of CE (Finlay and Speert, 1992; Pinch and Wilson, 1973).

### 3. Echinococcus multilocularis

#### 3.1. Distribution, hosts and genotypes

This parasite has a sylvatic lifecycle involving a wild rodent intermediate host and a canid definitive host (Table 1). In the circumpolar region, the most common wild canid hosts are the red fox (*Vulpes vulpes*) and the Arctic fox (*Vulpes lagopus*), and the most common intermediate hosts are arvicoline rodents (e.g. *Microtus* spp.) (Eckert and Deplazes, 2004). Domestic dogs are not considered an important part of the sylvatic lifecycle, but may play an important role in human infections. As accidental hosts, human AE is caused by the ingestion of *E. multilocularis* eggs from environments contaminated with canid faeces. Symptoms arise from the growth of the parasite into a metacystode in the liver which can metastasize to other organs (Ammann and Eckert, 1995). Without treatment, mortality rates approach 90% within ten years of diagnosis (Ammann and Eckert, 1995; Kern et al., 2003). Risk factors for infection include dog ownership, a farming or rural lifestyle and, to a lesser extent, ingestion of contaminated food from forests or gardens and drinking contaminated water (Kern et al., 2004; Yamamoto et al., 2001). Molecular analysis of *E. multilocularis* has revealed a number of strains which can be strictly grouped into Asian, Mongolian, European and North American strains (Nakao et al., 2009). Microsatellite analysis is able to show fine scale difference between the different *E. multilocularis* populations (Knapp et al., 2008, 2012, 2015).

##### 3.1.1. Russia

In Russia, the Asian and North American strains have been identified in red foxes, wolves and rodents (*Microtus gregalis*, *Microtus oeconomus*, *Myodes rufocanus*), whilst the Mongolian strains have only been identified in intermediate hosts to date (Lake Baikal voles: *Aticola strelzowi*, *Alticola olchonensis*). A European strain was identified in a Senegal bushbaby at Moscow zoo. It is thought that imported mulch used in the cage may have been the source of infection in this captive bred animal. The human AE cases in Russia which have been genotyped to date have been linked to Asian strains (Konyaev et al., 2013).

##### 3.1.2. Fennoscandia

In Fennoscandia, *E. multilocularis* has been identified in southern and central Sweden in red foxes and water (*Arvicola amphibius*) and field voles (*Microtus agrestis*) (Miller et al., 2016; Osterman Lind et al., 2011; Wahlström et al., 2015) as well as on the Norwegian high Arctic island of Svalbard, Grumant area (Fuglei et al., 2008; Henttonen et al., 2001; Knapp et al., 2012; Stien et al., 2010) in Arctic foxes and sibling voles (*Microtus levis*). Finland and mainland Norway are considered free from *E. multilocularis* based on annual surveillance (Wahlström et al., 2015). Microsatellite analysis of the *E. multilocularis* from samples from Sweden is yet to be finalised. The North American N1 strain has been identified in Svalbard (Knapp et al., 2012).

##### 3.1.3. North America

In North America, the distribution of *E. multilocularis* has historically been divided into the Northern Tundra Zone (NTZ, consisting of the northwestern coastal regions of Alaska and of the western Canadian Arctic, essentially corresponding to the range of Arctic fox and the North Central Region (NCR, consisting of the prairie ecozone in Canada and northcentral states in the contiguous USA). However, recent findings of the parasite in wolves in the taiga region, between the NTZ and NCR, suggest that these two populations may not be disjunct (Schurer et al., 2014a, 2016). The parasite has not been reported in the Yukon Territory or in the Canadian Arctic east of Hudson's Bay, possibly due to lack of sampling effort and/or low prevalence (Jenkins et al., 2013; Rausch, 1995). Definitive hosts reported for *E. multilocularis* are Arctic fox and red fox on islands in the Bering Sea and coastal regions of the western Arctic, and wolf in subarctic Taiga regions of the mainland western Arctic (Choquette et al., 1962, 1973; Eaton and Secord, 1979; Fay and Williamson, 1962; Gesy et al., 2014; Kirk, 2011; Rausch et al., 1990a, 1990b; Schurer et al., 2014a, 2016). Coyotes also serve as important DH for *E. multilocularis* in North America although they are

considered rare north of 65°N (Naughton, 2012). Arvicoline rodents and shrews serve as primary IH; the parasite has been reported in the ground squirrel (*Citellus undulatus*), red-back vole (*Clethrionomys* [= *Myodes*] *rutilus*), tundra vole (*Microtus oeconomus*) and shrew (*Sorex jacksoni*) from St Lawrence Island, (SLI) (Rausch and Schiller, 1956; reviewed in Jenkins et al., 2013). The life cycle of *E. multilocularis* in northern North America is considered primarily sylvatic, with spillover into domestic dogs and people recognised only on SLI (Rausch and Fay, 2002). Once considered a hyperendemic focus of AE, control programs and social change have successfully decreased transmission of AE in western Alaska, such that no cases have been detected since 1986 (Jenkins et al., 2013; Rausch et al., 1990a, 1990b).

It was once thought that *E. multilocularis* was genetically uniform in North America, and that the NCR population represented a relatively recent colonisation event from Arctic fox moving south into central Canada. Fine scale genetic differences and distinct genetic groupings in *E. multilocularis* are now recognised across north-western Canada (Gesy et al., 2014; Gesy and Jenkins, 2015), as well as larger genetic differences (using multiple mitochondrial DNA loci) between strains of the parasite on SLI, Alaska (North American N1 and Asian haplotypes A2 and A4) and the NCR (North American N2) (Nakao et al., 2009). In the NCR, coyotes and deer mice (*Peromyscus maniculatus*) are important DH and IH for *E. multilocularis* (Catalano et al., 2012; Gesy et al., 2013; Leiby et al., 1970), rather than foxes and arvicoline rodents in the NTZ and elsewhere in the world. In addition, there are distinct developmental differences between NTZ and NCR strains of the parasite in experimentally infected animals (Bartel et al., 1992; Rausch and Richards, 1971).

Together these genetic, ecological and biological differences, between the NCR and NTZ strains of *E. multilocularis* (Bartel et al., 1992; Rausch and Richards, 1971), suggests that we may need to revisit their taxonomic status. Finally, recent detection of European type strains of *E. multilocularis* in wild canids, a domestic dog, and a captive primate in Canada (Table 1) may reflect relatively recent introduction with imported wild or domestic canids, such that populations of *E. multilocularis* may be present as a complex mosaic of introduced and native haplotypes (Gesy et al., 2013, 2014; Gesy and Jenkins, 2015).

### 3.2. Surveillance activities and human AE

#### 3.2.1. Russia

The surveillance data in definitive hosts and intermediate hosts is incomplete in Russia and no active surveillance programs are currently in place. Data from studies of definitive hosts, in Yakutia 1981–2011, found relatively high *E. multilocularis* prevalence levels in arctic foxes, 50% (n = 54), compared to red foxes, 4.4% (n = 68) (Odnokurtsev and Sadalishchev, 2012). *E. multilocularis* cysts have been registered in multiple IH throughout Yakutia, Taimyr peninsula and Yamal peninsula (Bessonov, 1998; Gubanov, 1964; Gubanov and Fedorov, 1970; Shakhmatova, 1981): voles (*Microtus* spp., *Arvicola terrestris*, *Myodes* spp.) squirrels (*Citellus* spp., *Sciurus vulgaris*); lemmings (*Lemmus sibiricus*, *Dicrostonyx torquatus*, *Lagurus lagurus*), musk rats (*Ondatra zibethica*) and hares (*Lepus timidus*). Adult cestodes were observed in foxes, wolves and dogs in Yakutia, the Taimyr and Yamal peninsula (Gubanov, 1964; Shakhmatova, 1981). Ninety five percent of the foxes examined on the Yamal peninsula had *E. multilocularis*. The level of infection in rodents in the tundra zone was found to decrease from north to south (Shakhmatova, 1981).

Multiple human AE cases have been reported in the Altai republic (Asian strain) which borders to the Asian countries. Historically there has also been a single report from a patient in Yakutia (Martynenko et al., 1984) which is considered endemic for the North American strain (Konyaev et al., 2012) although genotyping was not carried out in this case. The prevalence of AE in humans in Yakutia is very low despite the high infection prevalence in IH and DHs. This also seen on the Taimyr Peninsula, where there are no registered cases of AE in humans but records in animal hosts (Shalaev, 1972 in Shakhmatova, 1981). Likewise, on the Yamal Peninsula up to 95% of the foxes are infected, whilst AE is not observed in the local human population (Luzhkov, 1962, 1963; Klebanoffskiy and Shpil'ko, 1969).

#### 3.2.2. Fennoscandia

Surveillance of foxes and fox faeces in mainland Norway and surveillance in raccoon dogs, foxes, and rodents in Finland have yielded no positive results (Wahlström et al., 2015). Surveillance efforts in these countries are designed to meet the EFSA standard for freedom from disease, which requires sampling efforts to show with 95% confidence that prevalence of *E. multilocularis* in definitive hosts is less than 1% (EFSA, 2010) and more than 3000 foxes/country have been examined since the start of the programs since the early 2000s. In Sweden subsequent to initial finding of *E. multilocularis* in foxes in 2011, during the annual surveillance program, surveillance efforts were intensified across the region (Wahlström et al., 2015). Two nationwide surveys of shot foxes and fox faeces have estimated overall prevalence in foxes in Sweden to be approximately 0.1% (SVA, 2015b; Wahlstrom et al., 2012). However, recent research has demonstrated the presence of micro-foci, clusters of positive fox faeces and rodents (as described by Giraudoux et al., 2002), in the Swedish environment (Miller et al., 2016; unpublished). Overall, prevalence in rodents is low (0–3.8%), but, in these localised areas, the prevalence can be as high as 30% (3/10 water voles collected 2013–2014) (Miller et al., 2016). Prevalence in sibling voles on Svalbard has been reported as high as 51% (23/45 voles) in 2000 (Henttonen et al., 2001) and was 8.5% in 353 arctic foxes collected over 8 trapping seasons, 1996–2004 (Stien et al., 2010).

There have been no recorded autochthonous cases of *E. multilocularis* in humans in mainland Norway, Sweden, or Finland. Although two human cases were identified in Sweden in 2012, these infections were thought to have originated abroad (Wahlström et al., 2015). Two cases of seroconversion without clinical disease have been reported from Svalbard (Wahlström et al., 2015) and one imported case of AE has been recorded in Finland (unpublished data, Antti Lavikainen).



### 3.2.3. North America

In northern North America, there is no systematic surveillance in place, and prevalence studies in DH largely rely on detection of adult cestodes on necropsy, although more recent studies are using molecular characterisation of taeniid eggs shed in faeces. Prevalence of *E. multilocularis* in Arctic foxes has historically been much higher on Alaskan islands (32–100%) than the Alaskan mainland (2–15%) and the western Canadian Arctic (1–2%). In other canids, prevalence was 2% of 100 and 55% of 11 red foxes in Alaska, and 8% of 73 wolves in the Northwest Territories of Canada (Gesly et al., 2014; Jenkins et al., 2013; Kirk, 2011; Schurer et al., 2014a). Interestingly, wolves in the coastal Northwest Territories were negative for *E. multilocularis* (Schurer et al., 2016), possibly due to dietary differences (consumption of ungulate and aquatic prey rather than rodents). While Arctic fox may be the most important definitive host on Arctic islands and along the Arctic coast, wolves may be the most important definitive host in subarctic and boreal regions given the absence of arctic foxes from this region and lack of *E. multilocularis* findings in red foxes (Rausch, 1995). With large home range sizes (2500–75,000 km<sup>2</sup> in northern Canada and Alaska) and dispersal distances (50–800 km) (Naughton, 2012), wolves may serve to bridge the “boreal gap” between the NTZ and NCR, and disseminate introduced European-type strains.

Apart from SLI there has been little effort to examine rodent intermediate hosts in northern North America. Rausch did not find lesions in 2500 rodents on the Alaskan mainland (Rausch, 1956); likewise, AE was not detected in 117 rodents examined in the central Canadian Arctic (Gesly et al., 2014). On SLI, prevalence in voles was highest in late May, reaching 80% in some locations prior to the “wash-out” effect from recruitment of uninfected young of the year (Rausch et al., 1990a, 1990b). In addition to seasonal variation, prevalence varies annually, regionally, and among IH species, with reports of 0.6% in brown lemmings (*Lemmus trimucronatus*) on the Alaskan mainland, 2–63% in tundra voles, 5–18% in red-backed voles, and 23–25% in shrews on SLI (Holt et al., 2005; Rausch and Schiller, 1956; reviewed in Jenkins et al., 2013). Deer mice are an important IH in the NCR, with prevalence up to 22%, and are present in the Yukon and Northwest Territories of Canada (Holmes et al., 1971; Naughton, 2012). Finally, AE in domestic dogs is increasingly detected in Canada, in both historically and newly endemic regions; in at least one dog and a captive primate cyst material proved to be a European strain (Christiansen et al., 2015; Jenkins et al., 2012; Peregrine et al., 2012; Skelding et al., 2014). This may reflect enhanced awareness, increasing overlap of dogs with infected wild canids in urban and rural areas, and/or emergence of introduced European strains of the parasite.

In northern North America, AE has not been considered a mainstream human health issue apart from western Alaska (Massolo et al., 2014). Reports are generally limited to case reports, reviews of hospitalisation data, and occasional studies, often serological, and subject to both false positives and negatives (Jenkins et al., 2013). While not nationally notifiable to public health authorities, Alaska (USA) and the Northwest Territories (Canada) mandate routine reporting of cases of human hydatid disease to public health authorities (Castrodale, 2003; Northwest Territories Health and Social Services, 2007).

No autochthonous cases of AE have been officially reported in northern Canada, and in Canada as a whole, only two autochthonous cases are reported (James and Boyd, 1937; Massolo et al., 2015). The latter was an immunocompromised person infected with a European-type strain despite the absence of travel history to Europe, suggesting a local source. Unfortunately, in human cases, AE is not always distinguished from CE. For example, in 251 cases of hydatid disease detected in Canada based on hospitalisation data, the type of echinococcosis was not reported. In addition travel histories are often incomplete, and a very low expected prevalence generally lead to assumptions that most cases are foreign acquired. However, a recent review of hospitalisation data in Canada found 16 AE cases between 2002 and 2011, six of which occurred in patients from northern regions (three northern territories and northern regions of the prairie provinces) where immigration is minimal (Schurer et al., 2015b). Where the geographic distribution of human cases of AE overlaps with the known distribution of *E. multilocularis* in wildlife in North America, the possibility of endemic transmission should be explored through genotyping of cysts along with epidemiological case investigation.

In the USA, autochthonous cases of human AE have historically been limited to SLI, with relatively few cases on the North Slope of the Alaskan mainland (Castrodale, 2003) and one case in the continental USA that was determined to be the N2 strain (Gamble et al., 1979; Yamasaki et al., 2008). Reports based on serology, skin tests, medical imaging, and biopsy are reviewed in Jenkins et al. (2013); Rausch and Schiller (1956); and Wilson et al. (1995). At its peak, seropositivity for AE on St. Lawrence Island reached 98/100,000 (Schantz et al., 1995; Wilson and Rausch, 1980). Risk factors for human AE on SLI included lifetime dog ownership, tethering dogs near the home, and living in a home built directly on the tundra (Stehr-Green et al., 1988). A total of 54 human cases of AE were reported between 1940 and 2010 in Alaska, and no cases of AE were reported from 1986 to 2010 (reviewed in Jenkins et al., 2013). Between 2010 and 2014, five cases of human hydatid disease were reported to the State of Alaska; of these, four were from interior and southeastern regions of Alaska and were more likely to be associated with CE rather than AE (Castrodale, 2003).

## 4. Current challenges and future needs for *E. canadensis* and *E. multilocularis* in the north

### 4.1. Reporting- surveillance

Perhaps one of the biggest challenges across all circumpolar countries is obtaining reliable data on the genetic diversity, incidence, prevalence, disease severity, health and economic burden of zoonotic *Echinococcus* spp. (Conraths and Deplazes, 2015). In animals, cervid IH for *E. canadensis* are not routinely inspected at harvest in many circumpolar nations, and *E. multilocularis* generally has low prevalence in IH. As a result, animal surveillance has largely relied on opportunistic terminal studies in definitive hosts, using animals trapped for fur harvest or predator control. The ability to detect the parasite in ground collected faeces

using molecular characterisation of taeniid eggs will enable more non-invasive studies that are ethically and logistically more feasible, especially in species of conservation concern in remote regions (Gesby et al., 2014; Kirk, 2011). Molecular identification of adult cestodes, recovered at necropsy, and not just for the differentiation of taeniid eggs can enable detection of mixed infections with *E. multilocularis* and *E. canadensis* in the intestines of DH, such as wolves and coyotes in North America, who harbour both species (Schurer et al., 2014a).

In people, both AE and CE are reportable diseases in some of the countries, but recognition of these diseases, their diagnosis, and compliance with reporting regulations remain a problem. In most parts of the circumpolar regions, these diseases are considered rare or even non-existent. In addition to under-reporting, CE is considered under-diagnosed because more than half of all cases are thought to be asymptomatic (Deplazes and Eckert, 2001). Hospitalisation records often do not differentiate between CE and AE and are often reported simply as “Echinococcus”, nor do they always report the travel history of the patient, adding to ambiguity in population level reporting (Schurer et al., 2015b). In addition, the metacestode lesion of AE can be misdiagnosed as hepatic carcinoma by practitioners unfamiliar with the disease (Jenkins et al., 2013; Massolo et al., 2014; Wilson and Rausch, 1980). This is problematic not only for documenting occurrence, but also for directing treatment.

Given what is already known about the distribution of the different *Echinococcus* genotypes (Fig. 1) the use of the host and geographically derived names for the different genotypes; such as camel strain for G6, Fennoscandian cervid strain for G10 as well as classifying *E. multilocularis* into North American, Asian, European and Mongolian strains; requires more extensive geographic survey and better understanding of the history of host biogeography, including anthropogenic translocations. There remains a great deal of controversy over the taxonomic, biological and health significance of genetic diversity within *Echinococcus* that can only be resolved by more universally accepted classification schemes, and molecular epidemiological investigation of human cases. This is difficult because CE caused by G6, G8, or G10 is often managed medically and thus cyst material is not available for genotyping and many medical practitioners see no obvious benefit to the patient or their case management. For human AE, it is possible that the low prevalence (relative to endemic regions of Europe and Asia) of human infection with AE in the North (outside Alaska and the Altai Republic of Russia) is due to a decreased opportunity for human exposure. However, this seems unlikely given the prevalence in DH in many circumpolar countries, and that many of the inhabitants hunt, consume untreated surface water, keep dogs as pets and working animals, and harvest foods that could be contaminated with faeces of wild canids. The geographical overlap between foci of human AE in the circumpolar north (in northwestern Alaska and the Altai republic of Russia) and the location of Asian strains of *E. multilocularis* in wildlife raises the possibility of higher zoonotic potential of Asian strains of the parasite, and not simply unique ecological and behavioural risk factors for exposure. It is fortunate perhaps, that the *E. multilocularis* strain (N1) and *E. canadensis* genotypes (G6, G8, G10) that are more prevalent in the circumpolar North would appear to have lower zoonotic potential. The human health significance of introduced European type strains of *E. multilocularis* in North America remains to be determined, although one case has been detected in an immunocompromised person (Massolo et al., 2015).

Immunocompromised people are at greater risk of infection with *Echinococcus*, often with more severe outcomes than immunocompetent patients (Chauchet et al., 2014; Massolo et al., 2015; Sailer et al., 1997). Increasing numbers of patients on immunosuppressive treatments (transplant, chemotherapy) or living with other cause of immunosuppression (such as HIV), in North America, Europe and Russia warrant greater consideration with regard to CE and AE. Indigenous people are at particular risk given the higher HIV diagnosis rates in this group compared to non-indigenous people in some regions (Negin et al., 2015). Therefore, targeted surveillance and enhanced awareness in medical professionals in high risk communities is advisable.

#### 4.2. Transmission

Environmental contamination with eggs, and subsequent ingestion in food and water, is thought to be the main route of transmission between definitive and intermediate animal hosts for *Echinococcus* spp. The risk of exposure for people via food and water has been harder to quantify, in large part due to the long temporal delay between time of infection and the development of clinical signs of AE/CE. The majority of studies have not been able to link risk factors for human AE infection with the consumption of certain types of fresh produce or water sources. Chewing grass and eating unwashed strawberries were risk factors for AE, in Germany whilst eating unwashed raw berries, mushrooms, vegetables or herbs was not (Kern et al., 2004). It was hypothesised that this could be due to the greater consumption of strawberries (relative to other food products) and/or reduced environmental contamination with eggs in woodlands compared to farmed/cultivated areas. In Poland, *E. multilocularis* DNA was detected on 23% of fruit (including 20% of raspberries), vegetable and mushroom samples collected from forests, gardens and plantations, in a highly endemic region using a nested-PCR method (Lass et al., 2015). This is surprising given that raspberries grow well above the ground, where natural contamination with canine faeces is expected to occur. There are also some significant technical criticisms to this paper, as highlighted by Robertson et al. (2016), like a poor limit of detection and lack of sufficient suitable controls, which mean that the results of this study should be interpreted with caution. Never the less further environmental investigation is warranted. Finally, the role of waterborne transmission of *Echinococcus* spp. is not well understood for the North, although elsewhere drinking well water has been found to be a significant risk factor (Yamamoto et al., 2001). In many northern regions, water treatment infrastructure is not optimal, and may well be overwhelmed seasonally or due to increased frequency and severity of extreme weather events as well as changes in regional hydrology in a future of rapid climate change. The role that fresh produce and water could have in the transmission of *Echinococcus* eggs needs to be elucidated as well as the need to develop environmental sampling methods with high sensitivity.

The potential for the introduction or reintroduction of different strains or genotypes of *E. multilocularis* or *E. canadensis* into new regions needs to be considered. These emergence events are associated with changing risk levels and should be addressed through revision of public health recommendations and guidelines for animal translocation. In Canada, introduction and establishment of European type *E. multilocularis* strains (possibly through importation of infected dogs) may be increasing the risk of AE in both dogs and people (Geszy et al., 2013; Jenkins et al., 2011; Massolo et al., 2015). In Fennoscandia the spread of *E. multilocularis* to Finland and Norway seems to be merely a question of time given that *E. multilocularis* occurs in Sweden, Russia and Estonia. Likely mechanisms of introduction include illegal importation of dogs from high endemic countries and non-compliance with existing deworming regulations (Maijala et al., 2001; Wahlström et al., 2015), dispersal of infected foxes from Sweden, Russia and Estonia, and establishment of the parasite in raccoon dogs (*Nyctereutes procyonoides*) (Schwarz et al., 2011), a suitable DH already established in Finland. Eradication efforts for raccoon dogs in Norway and Sweden have largely been successful (Mårdhundsprojektet, <https://jagareforbundet.se/vilt/Mardhundsprojektet/>), but these hosts could be important for the distribution and parasite prevalence on the Scandinavian peninsula in the future. Similarly, translocation of wildlife species could unknowingly introduce or enable the parasite lifecycle, as illustrated with beavers (*Castor fiber*) in Great Britain (Barlow et al., 2011) and the sibling vole in Svalbard (Henttonen et al., 2001). Finally the wildlife/human interface is constantly changing around the circumpolar North and will be altered by both environmental and social change, such as increasing urbanisation and encroachment into wildlife habitat. In Canada, coyotes, which are suitable DH for both *E. canadensis* and *E. multilocularis*, are increasing in abundance and geographic range and establishing and thriving in urban areas which may lead to increased risk of exposure of domestic animals and people (Catalano et al., 2012; Geszy et al., 2013; Naughton, 2012).

#### 4.3. Control methods

As transmission is mediated mostly through sylvatic reservoirs, efforts to control CE and AE have not been deemed practical or necessary in most of North America or Russia. Some of the genotypes involved in northern communities, especially native North American strains of *E. multilocularis* and *E. canadensis* are considered to be less pathogenic than other strains, but human cases are sporadically reported (Table 1). Spillover of *E. multilocularis* into domestic dogs and people on SLI was significant enough to warrant intervention. A 10 year program of monthly deworming of dogs with praziquantel resulted in an 83% reduction in prevalence in voles, which served as an indicator of environmental contamination and therefore human risk of exposure (Rausch et al., 1990a, 1990b). Unfortunately, such programs have since been discontinued (Hueffer et al., 2013); disease elimination programs are often victims of their own success.

Iceland, Norway and Finland have specific import regulations regarding deworming of dogs prior to entering the country (Eydal and Skirnisson, 2016; Wahlström et al., 2015). In Sweden deworming of imported dogs is a recommendation only (Wahlstrom et al., 2012). In Canada and the USA there are no deworming requirements for import of dogs. In Canada, praziquantel dosing of the national dog population is currently not considered cost-effective for the control of CE in people, in large part due to the low incidence of CE in people and the relatively benign nature of infection with *E. canadensis*, which is often managed through a “watch and wait” approach (Schurer et al., 2015b). However, it may be cost effective at a local scale; for example, in a community with cases of CE where dogs are demonstrated to be a source of environmental contamination (Himsworth et al., 2010). As well, AE is a considerably more pathogenic disease. Schurer et al. (2015a, 2015b) estimated that the average cost to treat a single human CE case was \$8,842 (CDN), whilst another study estimated the median treatment cost per case of AE in Switzerland to be €90,230–€118,146 (Euros) (Torgerson et al., 2008). If AE cases are detected in which dogs are determined to be a source of human exposure (rather than direct exposure to eggs shed by wild canids), then programs for dog treatment may well be effective at reducing the human burden of disease as well as economically justified for some communities.

Future recommendations for enhanced surveillance and potential control efforts for CE and AE across the north could include the following: development of serological tests that are optimised for the strains/species of *Echinococcus* currently circulating, molecular characterisation of cyst material and epidemiological follow-up to determine source of and risk factors for human CE and AE (especially to determine if cases are autochthonous), increased awareness of clinical and pathological signs among physicians and veterinarians in endemic regions, and addition of these parasites to the list of nationally notifiable pathogens for both animals and people (Schurer et al., 2015b). Distinguishing AE from CE infections in humans is a clinical and epidemiological necessity. In addition, regulatory bodies for animal health need to consider genetic diversity within the *Echinococcus* genus and marked regional differences in prevalence, testing and/or treatment of imported dogs, translocated wild canids and potential intermediate hosts to develop policies that prevent importation of foreign strains or dissemination into non-endemic regions. One policy solution might include zoning portions of North America (for example, the Atlantic provinces and some eastern states) as *Echinococcus*-free, and applying pet mobility restrictions and deworming regulations accordingly as is currently done in Finland, Norway and Iceland.

## 5. Conclusions

Our knowledge regarding *E. canadensis* and *E. multilocularis* in wildlife in Russia, Fennoscandia and North America, is continuously growing. However, without systematic surveillance in humans, animals and environmental samples obtaining reliable data on incidence, prevalence, genetic diversity and human health burden from across the circumpolar region remains problematic. We are fortunate that *E. canadensis* appears to have considerably lower pathogenicity compared to *E. granulosus* ss. and that

the “North American” *E. multilocularis* strains may have lower zoonotic potential than European and Asian genotypes. However this situation could be changing due to rapid climate and landscape change, anthropogenic changes in the wildlife/human interface, and globalisation of trade and travel. Increasing survey effort and more powerful molecular tools are already bringing to light a much more complicated picture of genetic diversity with *Echinococcus* including the presence of Asian and European type strains of *E. multilocularis* in animals around the circumpolar north. It may be a decade or more before we are able to assess the effects in the human populations in the region.

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