CHAPTER 46

Cestodes: Intestinal and Extraintestinal Tapeworm Infections, Including Echinococcosis and Cysticercosis

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Man is the only animal which esteems itself rich in proportion to the number and voracity of its parasites.

George Bernard Shaw (1856-1950)

Infections by cestodes (tapeworms, or flat, segmented worms) in the United States, Canada, and other affluent countries are rare but do occur in specific at-risk populations related to geographic, behavioral, or migration risk characteristics. Clinical suspicion is needed to detect quiescent infections or asymptomatic disease.

Some of the tapeworms affecting humans are endemic in the United States, Canada, and other developed nations. Practically, these can be divided clinically into tapeworm infections that are purely intestinal-dwelling (*Taenia saginata*, beef tapeworm; *Taenia solium*, pork tapeworm; and *Diphyllobothrium latum*, fish tapeworm), purely tissue invasive (echinococcosis), or with both phases (pork tapeworm and cysticercosis). All other human tapeworm infections are usually not clinically significant but reflect poor socioeconomic or immigration status. Rarely, transmission of tapeworm infections introduced by means of infected food handlers under unhygienic circumstances (a public health issue) has been reported. **Table 46.1** shows the common tapeworm infections affecting humans, their usual distribution, and clinical significance.

Humans acquire infective tapeworm larvae or eggs through ingestion of contaminated soil or infected food or by accidentally swallowing the intermediate vectors such as the flea or beetle. *Echinococcus* spp. are tapeworms of carnivorous mammals, usually dogs, that serve as the definitive hosts for the parasite. The extraintestinal larval stages of *Echinococcus* spp. cause cystic mass lesions, most often in the liver of sheep, deer, or moose, but can affect virtually all other organs as well. Humans are an inadvertent and usually "dead-end" intermediate host.

EPIDEMIOLOGY AND DEMOGRAPHICS OF HUMAN TAPEWORM INFECTIONS IN DEVELOPED NATIONS

Intestinal Tapeworms

In the United States and other developed nations, tapeworm infections are uncommon in the general population and are found more frequently in defined migrant, ethnic, and certain cultural groups with specific risks, such as exposures in endemic regions of the world, dietary practices, food choices, and methods of food preparation. Occupational risk exposures include certain animal contacts, veterinary care, and animal control. Humans act as the definitive host for the beef, pork, and fish tapeworm, becoming infected through the

TABLE 46.1 Distribution and Usual Clinical Significance of Tapeworms Affecting Humans

Parasite	Distribution	Usual Clinical Significance
Taenia saginata (beef tapeworm)	*T. saginata* is common in cattle-breeding regions worldwide with humans being a definitive host and cattle the intermediate host. Areas with the highest (i.e., >10%) prevalence are Central Asia, the Near East, and Central and Eastern Africa. Areas with low (i.e., 1%) prevalence are Southeast Asia, Europe, and Central and South America. Pre-patent period: 3-5 months Life span: up to 25 years Length of worms: 4-8 m	Adult tapeworms live in the gastrointestinal tract of the human host. Eggs are excreted in the stools, and motile tapeworm segments can also be expelled from the bowels. The beef tapeworm does not cause invasive disease in humans, but in regions where distribution overlaps with pork tapeworm, must be distinguished from the latter, which does cause tissue infections in people. All ages, races, and genders are susceptible to infection that is acquired by eating larvae-infected undercooked beef meat.
Taenia solium (pork tapeworm)	*T. solium* is endemic in Central and South America, Southeast Asia, India, the Philippines, Africa, Eastern Europe, and China, with humans being a definitive host and pigs the intermediate host. Areas of highest prevalence include Latin America and Africa. In some regions of Mexico, prevalence of infection may reach 3.6% of the general population. Pre-patent period: 3-5 months Life span: up to 25 years Length of worms: 3-5 m	Adult tapeworms live in the gastrointestinal tract of the human host. Eggs are excreted in the stools, and motile tapeworm segments can also be expelled from the bowels. The pork tapeworm larvae causes invasive disease in humans affecting soft tissues and the brain (cysticercosis). All ages, races, and genders are susceptible to infection that is acquired by eating larvae-infected undercooked pork meat or by ingestion of pork tapeworm eggs.

Continued

TABLE 46.1 Distribution and Usual Clinical Significance of Tapeworms Affecting Humans—cont'd

Parasite	Distribution	Usual Clinical Significance
Diphyllobothrium latum (fish tapeworm)	In North America, *D. latum* infections have been previously reported in fish from the Great Lakes. There are six *Diphyllobothrium* species known to reside in Alaskan lakes and rivers, and some saltwater species may also be seen in North America. *Diphyllobothrium* infections are not species specific, and widespread reports describe infection in North American fish-eating birds and mammals. Humans are a definitive host, and crustaceans, followed by fish, are intermediate hosts. The incidence in the USA has been declining recently. Pike, perch, and salmon are among the fish most commonly infected. Reports are commonly made of *D. latum* infection in humans residing in Europe, Africa, and the Far East. Pre-patent period: 3-5 weeks Life span: up to 25 years Length of worms: 4-10 m	Adult tapeworms live in the gastrointestinal tract of the human host. Eggs are excreted in the stools, and motile tapeworm segments can also be expelled from the bowels. The fish tapeworm does not cause invasive disease but due to its length and potential to interfere with vitamin B12 absorption can cause a number of nonspecific symptoms. All ages, races, and genders are susceptible to infection that is acquired by eating undercooked, infected fish flesh. People preparing fresh fish, implements used to prepare fish (e.g., knives and cutting boards), and raw or undercooked fish meals (e.g., sushi, sashimi, ceviche) may be associated with a higher risk of infection.
Dipylidium caninum	*D. caninum* is a cosmopolitan tapeworm infection of dogs with inadvertent human infections occurring through ingestion of the intermediate host, a flea that has fed on the tapeworm eggs contaminating the animal's fur or dog feces. Human infections have been reported in Europe, the Philippines, China, Japan, Argentina, and North America. Pre-patent period: 3-4 weeks Life span: <1 year Length of worms: 10-70 cm	Adult tapeworms live in the gastrointestinal tract of the inadvertent human host, usually a child. Perianal irritation may occur with the passage of motile segments of the tapeworm; small "grain of rice–like" motile segments may be seen in the stools. The proglottids are motile when passed and may be mistaken for maggots or fly larvae.
Hymenolepis nana (dwarf tapeworm)	*H. nana* is a cosmopolitan intestinal tapeworm usually infecting rodents (mice or rats). The intermediate host, a beetle, is not required to complete its lifecycle in definitive hosts. Ingestion of tapeworm eggs by a definitive host, including humans, can reestablish an adult tapeworm infection. Pre-patent period: 2-3 weeks Life span: many years due to autoinfection Length of worms: 2.5-4 cm	Often associated with environments with poor sanitation, the dwarf tapeworm causes few clinical problems with nonspecific abdominal complaints, loosening of the stools, perianal irritation, and the possible presence of small motile segments visible in the stool or on undergarments.

Hymenolepis diminuta	The rat tapeworm requires a grain beetle as an intermediate host, so is most common in grain-producing areas of the world or where grain or other dry foods are stored. Human infections are uncommon. Pre-patent period: 3 weeks Life span: <1 year Length of worms: 20-60 cm	Often associated with environments with poor sanitation, the rat tapeworm rarely infects humans and causes few clinical problems with nonspecific abdominal complaints, loosening of the stools, and perianal irritation and the possible presence of small motile segments visible in the stool or on undergarments.
Echinococcus granulosis and *Echinococcus vogeli*	*E. granulosis* is a tapeworm of canines (dogs) with other vertebrates, most commonly sheep, as the intermedia te host. Ingesting tapeworm eggs passed in the stools of infected dogs infects humans. Once common in all sheep-raising areas of the world (Asia, Europe, the Americas, Africa, and Oceania), animal husbandry practices are resulting in effective disease control in most affected areas.	The larval tapeworm infection in humans causes hydatid disease with large, complex cystic masses occurring most commonly in the liver, but any organ can be affected, presenting with mass effects. Cysts can rupture, causing allergic reactions and anaphylactic shock. Cysts can also present with secondary bacterial infection. Pulmonary cysts can spontaneously rupture and be expelled through the mouth. If intraperitoneal rupture of a primary cyst occurs, multiple secondary cysts can develop in the peritoneal cavity.
Echinococcus multilocularis	*E. multilocularis* is a tapeworm of canines, with foxes and wolves being particularly important definitive hosts with other vertebrates (small rodents such as voles, lemmings, and mice) as intermediate hosts. Humans become infected by ingesting tapeworm eggs passed in the stools of infected canines. This is a rare form of echinococcosis that occurs predominantly in Arctic regions of Europe and North America. It can be prevented by carefully washing strawberries, cranberries, blueberries and other foods that may be contaminated with canine feces.	The larval tapeworm infection in humans causes alveolar hydatid disease with large, complex externally budding cystic masses, occurring most commonly in the liver. Due to its rapid growth and budding characteristics, this form of echinococcosis behaves more like a malignancy displacing liver tissue than an indolent infection, such as cystic hydrosis.

ingestion of encysted larvae in the muscles of various animals. Humans can also be infected by ingesting tapeworm eggs of *T. solium* and *Echinococcus* spp., both leading to cystic tissue disease, or *Hymenolepis nana*, leading to development of an adult worm and patent infection.

In the typical lifecycle of tapeworm infection, ingesting the cyst-infected meat releases the larval forms in the gastrointestinal tract. The larvae attach to the mucosal lining of the small intestine by the head, or scolex. The tapeworm grows from each scolex, forming proglottids containing both male and female reproductive organs. Adult tapeworms (beef, pork, and fish) may reach lengths of 4.5-10 m. The proglottids may detach from each other, forming a short chain of segments (strobila) containing fertilized eggs. These segments and eggs are passed in the stool. If the scolex and neck of the tapeworm remain attached to the host gut mucosa, it can continue to produce proglottids for many years.

The beef tapeworm (*T. saginata*) infection is most common in people who eat raw or undercooked beef dishes. Recent immigrant groups from areas endemic to *T. saginata* in cattle-rearing Africa (i.e., Ethiopia), cattle and llama-rearing areas in Latin America, and reindeer-rearing areas in the Northern Hemisphere are at greatest risk.

The adult pork tapeworms and cystic forms of the infection (cysticercosis) are found among people exposed to undercooked pork or in contact with the excreted eggs of *T. solium*. This is most common in immigrants and refugees from endemic areas in the developing world.

Infection with the fish tapeworm, *D. latum* and other *Diphyllobothrium* species, occurs when eating raw, smoked, pickled, or undercooked fish. Those at risk are the Inuit, fishermen, Jewish home cooks (uncooked gefilte fish), and consumers of sushi and sashimi, ceviche, or caviar made from infected fresh water fish and roe. The highest risk fish are raw salmon and other anadromous fish (fish ascending rivers to spawn): American shad, blueback herring, short-nose sturgeon, striped bass, and steelhead trout. In Europe, these fish include pike, perch, and turbot.

H. nana and Hymenolepis *diminuta* are rodent tapeworms with rare human infections in developed nations. Human infections are through accidental ingestion of rodent feces containing the tapeworm egg (*H. nana*), or larvae-containing fleas and grain beetles (*H. nana* and *H. diminuta*). Young children in rodent-infested environments are at risk of *H. nana*. Person-to-person transmission of *H. nana* occurs through fecal-oral contamination.

Dipylidium caninum infection usually occurs in children living in households with exposures to a dog, where accidental ingestion of an infected flea can result in a human infection.

Tissue Tapeworms

In the United States, areas endemic to *Echinococcus granulosus* exist in sheep-raising areas of California, Utah, and in Alaska, where dogs, wolves, and other canines are frequently infected. Immigrants from the Mediterranean region, the Middle East, and South America are also at greater risk. Alveolar hydatid disease caused by *Echinococcus multilocularis* is endemic in Alaska and the Arctic regions, central North America involving some parts of Canada, the United States, central Europe, Siberia, and northern Japan.

Cysticercus cellulosae are the tissue lesions caused by larval dissemination of *T. solium*. This disease occurs in Mexico, Central and South America, Africa, India, China, Eastern Europe, and Indonesia. Cysticercosis infection of the central nervous system is a common cause of seizures among immigrants from Latin America to the United States.

People are usually infected with *T. solium* when the eggs passed in the stool are ingested in contaminated food or water. Person-to-person transmission in household settings has been documented in non-endemic areas. Autoinfection by refluxing *T. solium* eggs within the human gut may be a means of larval infection. The larvae are found in subcutaneous connective tissue. The second most common site is the eye, followed by the brain, muscle, heart, liver, and lungs. Cysts may cause irreparable damage to the eye.

Sparganosis has been reported from many countries of the world but is most common in eastern Asia and rarely elsewhere. It is almost impossible to identify spargana to the species level, making it unclear as to which species actually infect humans. Infection in humans is caused by swallowing the first intermediate infected copepod hosts in contaminated drinking water or eating raw or undercooked amphibians, reptiles, or mammals, which are second intermediate hosts in endemic areas. The spargana can migrate to virtually any part of the body and grow up to 6 cm in length. Asymptomatic soft tissue or internal organ infection is most common in humans. Surgical removal, if necessary, is the usual treatment.

Clinical Presentation of Tapeworm Infections

Intestinal tapeworm infections usually asymptomatic. Nonspecific abdominal pain, cramps, and diarrhea have been described. Discovery of infection occurs when a motile segment or a "tape" of proglottids is passed during a bowel movement or spontaneously appears in the undergarments. Perianal or vaginal irritation may also occur. Chronic *D. latum* infection may rarely cause vitamin B12 deficiency and megaloblastic anemia.

Tapeworm infections are confirmed by the presence of eggs or segments found in stool specimens during parasitological examination.

Tissue infection with *Echinococcus* spp. (*E. granulosis* or *E. vogeli*) causes hydatid disease and most commonly presents with a mass effect of the slow-growing cystic lesions, usually in the liver, taking decades to reach symptomatic size. Primary cysts may reach 25 cm or more in diameter. Rupture of a cyst can release antigenic material, causing an acute allergic or anaphylactic reaction. Secondary bacterial infection of liver cysts can present as a liver abscess. Previous leakage of a liver cyst into the peritoneal cavity may cause multiple secondary cystic growths throughout the abdominal cavity. Pulmonary echinococcal cysts are usually asymptomatic, but if they rupture into the bronchial tree they can cause hemoptysis and coughing out of daughter cysts that look like grape skins. Echinococcal cysts in other organs (e.g., skin, bone, brain, eye, and visceral organs) have been described. Alveolar echinococcal infections (*E. multilocularis*) present as a rapidly progressive solid tumor mass in the liver. Due to their aggressive presentations, these infections clinically behave like a malignancy.

Tissue infection with cysticerci of *T. solium* infection is usually asymptomatic, even when the cysts are in the brain. Neurocysticercosis is a common cause of seizure disorder in Mexico, Central and South America, Africa, India, China, Eastern Europe, and Indonesia. A rare form of central nervous infection, called racemose cysticercosis, is associated with grape-like cystic growths in the brain causing obstructive hydrocephalus, arachnoiditis, and cerebellitis.

Diagnosis

A clinical diagnosis to the species level cannot be made from general history, physical examination of the patient, or the gross examination of expelled tapeworm segments in the stool. Patient demographics can be very suggestive of a specific adult tapeworm or tissue cystic presentation, but definitive diagnosis requires expert parasitologic laboratory testing, radiographic imaging, and, on occasion, pathologic specimen examination for parasitic elements.

Parasitologic Testing

Note: tapeworm segments or proglottids containing eggs of *T. solium* are infectious.

1. *Stool specimens*: eggs of *Taenia* spp., *Diphyllobothrium* spp., *D. caninum*, and *Hymenolepis nana* may be found in concentrated stool preparations made from fresh or preserved stool samples. The eggs of *T. saginata* and *T. solium* are morphologically indistinguishable; identification depends on examination of proglottids and scolices, if available. *Taenia* eggs may also be found on adhesive tape preparations taken near the anus. A copro-antigen enzyme-linked immunosorbent assay (ELISA) has been developed to detect *Taenia* species antigens from feces. Polymerase chain reaction is highly sensitive, specific, and an easy technique to detect *T. saginata*.

2. *Tapeworm segments*: tapeworm segments may be found in the fresh or preserved stool by macroscopic examination or directly as a segment in formalin. If the head of the tapeworm is found, the species can be identified by the scolex.
   The tapeworm species can also be identified by the characteristics of the proglottid seen on staining in the parasitology laboratory with India ink. The proglottid can also be stained with hematoxylin-eosin (H&E).
3. *Cystic fluid*: cystic fluid obtained during surgery or by aspiration can be examined for the hydatid "sand" of *Echinococcus* hooklets or protoscolices. Microscopic wet mounts and smears are stained by an acid-fast procedure. Microscopic examination of H&E-stained smears of the hydatid cyst shows a wall surrounded by host tissue capsule, a laminated acellular layer, and an inner, nucleated, germinal layer that gives rise to brood capsules. These capsules can degenerate or form "daughter" cysts containing many viable protoscolices.
4. *Parasite serology:* serologic testing for tissue infection with *T. solium* and *Echinococcus* spp. is performed by the Centers for Disease Control and Prevention (http://www.cdc.gov/dpdx/).

For hydatid disease, specific immunoglobulin G (IgG) ELISA is the most sensitive test (83.5% sensitivity in one study). The CDC's immunoblot assay with purified *T. solium* antigen is the test of choice by the World Health Organization and the Pan American Health Organization. It has 100% specificity and a high sensitivity. These tests do not differentiate between active and inactive infections and cannot be used to evaluate the outcome and prognosis of medically treated patients.

Radiology

Barium contrast studies of the bowel can show filling defects of the long ribbon-like bodies of adult tapeworms. Any radiologic imaging technique (ultrasound, computed tomography, or magnetic resonance) will demonstrate the classical features of hydatid disease with primary cyst and multiple daughter cysts. Hydatid cysts in the lung may not have discernible daughter cysts, and they rarely calcify in the outer membrane. Alveolar echinococcal disease of the liver appears as a solid mass with external extensions that spread to adjacent organs, making it difficult to distinguish from other invasive tumors in the liver. Cysticerci in the soft tissues appear as small calcifications. Neuroradiology of cysticerci in the brain will show typical lesions very suggestive of the diagnosis in the cystic form of the disease. Racemose cysticercosis of the brain can be very challenging to diagnose with radiology alone.

Pathology

Pathologic examination is not indicated in intestinal tapeworm infections. In tissue examinations of an echinococcal cyst and its contents, cysts tend to be round or oval. The wall consists of a fibrous capsule of host tissue; a tough, elastic, laminated, acellular layer; and an inner nucleated germinal layer from which arises the brood capsule. The fluid or "sand" in the cysts contains protoscolices, daughter cysts, and degenerated brood capsules that have liberated protoscolices and hooklets. In alveolar hydatid disease, the laminated layer is invasive and diffusely spread in the tissue; brood capsules are not produced.

Brain or muscle tissue examination for cysticerci is generally not required for a diagnosis of disseminated *T. solium* infection.

Clinical Management

Treatment

Antihelminthic drugs and their dosages for treatment of tapeworm infections are listed in **Table 46.2**. Praziquantel and albendazole are the drugs of choice for most intestinal infections. Niclosamide tablets must be taken on an empty stomach and thoroughly chewed and swallowed. Niclosamide kills the parasite by direct contact with the tapeworms. If the scolex is protected from contact with the drug, the adult tapeworm infection may survive

TABLE 46.2 Treatment for Human Tapeworm Infections

Parasite	Drug	Dosage
Taenia saginata	Praziquantel	Single dose of 5-10 mg/kg
	Alternative: niclosamide	2.0 g once (50 mg/kg once)
Taenia solium (intestinal)	Praziquantel	Single dose of 5-10 mg/kg
	Alternative: niclosamide	2.0 g once (50 mg/kg once)
T. solium (tissue larval)	Seizure control	See specific anticonvulsants or seek specialist's consultation
	Alternatives:	
	Albendazole	400 mg p.o. (15 mg/kg, max. 400 mg b.i.d.) b.i.d. × 8-30 days (repeat if necessary)
	or	
	Praziquantel	50-100 mg/kg per day in three divided doses ×30 days
Diphyllobothrium latum	Praziquantel	Single dose of 5-10 mg/kg
	Alternative: niclosamide	2.0 g once (50 mg/kg once)
Dipylidium caninum	Praziquantel	Single dose of 5-10 mg/kg
	Alternative: niclosamide	2.0 g once (50 mg/kg once)
Echinococcus granulosis	See indications for surgery or percutaneous drainage. Albendazole	400 mg b.i.d. × 1-6 months (15 mg/kg per day; max. 400 mg b.i.d.)
Echinococcus multilocularis	See indications for excisional surgery	Albendazole or mebendazole may be adjunctive to curative surgery or, if inoperable, supportive therapy
Hymenolepis nana	Praziquantel	Adults and children: single dose of 25 mg/kg
	Alternative: nitazoxanide	500 mg × 3 days (1-3 years old: 100 mg b.i.d. × 3 days; 4-11 years old: 200 mg b.i.d. × 3 days)
Hymenolepis diminuta	Praziquantel	Adults and children: single dose of 5-10 mg/kg
	or	
	Nitazoxanide	500 mg × 3 days (1-3 years old: 100 mg b.i.d. × 3 days; 4-11 years old: 200 mg b.i.d. × 3 days)

b.i.d., Twice per day; *p.o.*, by mouth.

treatment. Drinking orange juice, lemonade, or similar acidic, mucolytic beverage 30 min before taking the tablets is recommended.

Presumptive mass treatment may be chosen over individual assessment in some settings.

Surgical excision is the only reliable means of cure of hydatid disease and is the primary therapy for alveolar echinococcal disease. In nonresectable cases, treatment with albendazole may stabilize and sometimes cure infection. Hydatid cysts may be treated with albendazole for an extended period of time (1-6 months) with good clinical outcomes. Affected patients may also benefit from surgical resection or percutaneous drainage of cysts. Percutaneous aspiration–injection–reaspiration with ultrasound guidance plus albendazole or mebendazole

therapy has been effective for management of hepatic hydatid cyst disease. Praziquantel is indicated preoperatively or in case of surgical spillage of cyst contents.

Initial therapy for patients with inflamed central nervous system parenchymal cysticercosis should focus on symptomatic treatment with antiseizure medication. Treatment of central nervous system parenchymal cysticerci with antiparasitic drugs (albendazole or praziquantel) remains controversial due to the potential of an exacerbated inflammatory response in the brain to dead and dying parasites. If treatment is undertaken, patients with live parenchymal cysts and seizures should be treated with combination drug therapy, for example, albendazole together with steroids (6 mg dexamethasone or 40-60 mg prednisone daily) and an antiseizure medication.

Public Health and the Role of Screening

An identified index case of intestinal tapeworm infection may have community or public health implications, depending on the species of tapeworm, the socioeconomic and geographic setting, and whether institutionalized care is a potential co-factor. Gastrointestinal infection with *T. solium* has been associated with larval infection in communities not usually associated with the consumption of pork meat products. The diagnosing clinician may wish to consult with the regional public health officials on recommendations for reporting, surveillance, or investigation of unusual parasitologic findings.

Prognosis

Untreated intestinal and tissue tapeworm infections may persist for years and may be associated with mild eosinophilia. Megaloblastic anemia is rare in chronic *D. latum* infection. *T. solium* infections may result in a disseminated extraintestinal larval infection called cysticercosis.

Management of associated seizure disorders in neurocysticercosis is generally all that is required for affected patients. Very rarely is a persistent adult *T. solium* tapeworm infection found, but this diagnosis should prompt consultation with a specialist before antiparasitic treatment is initiated, due to the possibility of exacerbating adverse consequences of undiagnosed larval infection of the central nervous system, as noted previously.

The prognosis of both hydatid and alveolar echinococcal disease has very much improved with modern surgical, percutaneous, and therapeutic management. Long-term follow-up is required to monitor clinical improvement and to observe for recurrences.

FURTHER READING

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