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***ECHINOCOCCUS GRANULOSUS AND
RELATED SPECIES
(CYSTIC HYDATID DISEASE)***

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The infection is acquired by the ingestion of ova from the feces of definitive hosts. The disease in humans and other intermediate hosts is called hydatid disease.

alveolar echinococcosis *Echinococcus multilocularis* polycystic echinococcosis



Most human infections are caused by *E. granulosus* genotype 1 (the sheep strain).

The life cycles of the species of *Echinococcus* are similar, but the geographic distributions, types of hosts, and clinical and morphologic features of the parasites differ significantly.



The adult tapeworm is only 3 to 8 mm long and has two to five segments. Dogs often are hosts to thousands of adult worms. When sheep or humans ingest eggs from the feces of infected dogs, the embryos hatch in the intestine and burrow through the intestinal wall to gain access to the portal circulation. The embryos that survive are able to develop in many tissues, but they do so most commonly in the liver or lungs, where they become the cystic larval structures called *hydatid cysts*.

Transmission

Humans acquire cystic hydatid disease by ingestion of ova shed by the definite host, typically dogs. Close contact with dogs can result in infection because tapeworm eggs can be found on the dog's perianal hair, muzzle, and paws. Contaminated food, drink, or fomites.

Pathology and Pathogenesis

In cystic hydatid disease, the pathologic process is related to compression or displacement of the host's tissue. Cyst growth is variable and depends on (1) the organism involved being slower in the liver than in the lung; (2) the *Echinococcus* species, with *E. canadensis* usually being slow growing; and (3) the age of the subject, with cyst growth being faster in younger individuals.²³⁶ Rates of up to 4 to 5 cm per year have been reported, but growth may be more rapid in the lung. The cyst may exceed 35 cm in diameter in the abdominal cavity.


Cysts may calcify after many years, which usually signifies death of the parasite. Hydatid cysts may form foci for secondary bacterial infection causing liver and lung abscesses.

Clinical Manifestations

Most children with *E. granulosus* infection have a single unilocular cyst, but multiple cysts are seen in 15% to 30% of patients, usually in a single organ system. The most common site of the cysts is in the liver.


Approximately 10% of cysts are found in sites other than the liver or lung, including the spleen, kidney, peritoneum, genitourinary tract, bone, muscles, heart, eye, and brain.

Cyst leakage or rupture is one of the most common complications and may result in seeding of body cavities with fertile protoscolices, local inflammatory reactions (e.g., pneumonitis), infection of the cyst cavity, and type 1 allergic reactions, including hypotension, urticaria, and eosinophilia. Rupture may occur spontaneously or secondary to trauma or surgery



The wide spectrum of symptoms in cystic hydatid disease depends on the number, size, and location of the cysts.

or epigastric pain, enlargement of the liver, or nausea and vomiting.



traumatic event and may cause a range of symptoms, including fever, abdominal pain, hypotension, and allergic manifestations, including eosinophilia, urticaria, and anaphylaxis. Rupture into the biliary tract may cause cholangitis with fever and right upper quadrant pain.

may cause cough, chest pain, dyspnea, hemoptysis, and a salty taste. Fever may be present.

As many as one-third of pulmonary cysts rupture into the pleural space or into a bronchus.

Bone cysts are seen in patients with bone pain or pathologic fracture. Vertebral hydatid disease causes signs and symptoms of spinal cord and radicular compression;

intracranial cysts, 50% to 75% are seen in children. Increased intracranial pressure with headache, vomiting, and focal neurologic symptoms are common findings. Seizures also may occur.

Diagnosis

The diagnosis of cystic echinococcosis usually is suspected on the basis of clinical or radiologic findings plus a history of residence in an endemic area.

liver enzyme results. Eosinophilia typically is low grade or absent. The initial diagnosis of cystic hydatid disease often is based on imaging findings.

surface of the fluid in a ruptured pulmonary cyst has a characteristic “water lily” appearance.

Ultrasound techniques are useful for defining most cysts within the abdomen and can differentiate fluid-filled cysts from solid tumors.

Radiographically apparent cyst wall calcification occurs only in liver or spleen cysts and generally takes more than 5 to 10 years to develop. Bone cysts typically produce radiolucencies without periosteal reaction. MRI offers little advantage over CT for the imaging of cysts except in the CNS and for demonstration of biliary fistulas.¹²⁶ Fine-needle aspiration of cysts for establishment of the diagnosis can cause spillage of hydatid fluid.




The fluid obtained should be examined for evidence of protoscolices, hooks, or antigen.

Multilocularis tested by ELISA, indirect hemagglutination, and fluorescent antibody assays are positive in 50% to 90% of cases. False-negative results are more common in patients with pulmonary, brain, and splenic cysts

treatment of choice for nearly all cases of cystic echinococcosis, but a range of different approaches have been accepted gradually, including surgery, antiparasitic chemotherapy, percutaneous drainage (e.g., PAIR), and “watch and wait” Currently expert opinion suggests that treatment should vary on the basis of the location of the cyst or cysts, the nature and size of the cysts, the condition of the patient

albendazole and mebendazole have been used for chemotherapy Approximately one-third of hepatic cysts will resolve in response to benzimidazoles, and 30% to 50% will improve.



the treatment of choice for small (<5 cm Chemotherapy also has proved to be successful for treatment of small pulmonary cysts.⁶⁵ However, a high failure rate exists with chemotherapy alone for larger lesions in either the liver or the lung.²⁰ Albendazole is thought to be more effective than mebendazole. Albendazole typically is dosed at 10 to 15 mg/kg per day in two daily doses continued for 3 to 6 months. Side effects may include neutropenia, elevated liver enzymes, and alopecia.



PAIR is thought to be the treatment of choice for uncomplicated CE₁ lesions larger than 5 cm and CE_{3a} lesions.

The goal of surgery is to remove the entire cyst without spilling its contents.

Pulmonary cysts have been treated successfully surgically, medically, and with PAIR.

If the cyst wall remains in place after the patient has undergone chemotherapy or PAIR



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