Hydatid cyst

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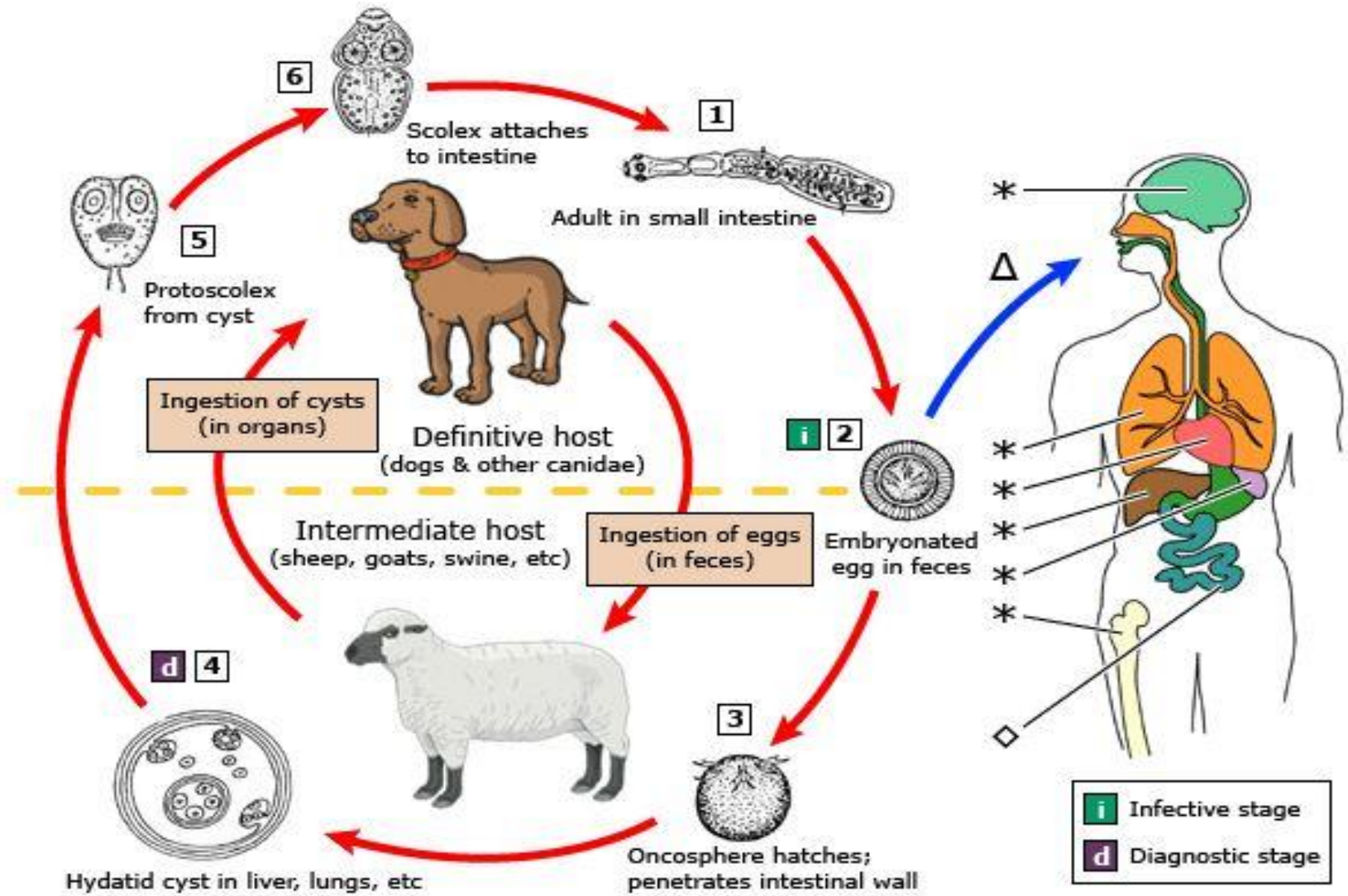
- Echinococcal disease is caused by infection with the tapeworm echinococcus.
- Four species of Echinococcus cause infection in humans. E. granulosus and E. multilocularis are the most common.The two other ,are less frequently associated with human infection.
- Echinococcus granulosus — E. granulosus causes cystic echinococcosis (CE).
- **Life cycle** — The life cycle of echinococcus includes a definitive host (usually dogs or related species) and an intermediate host (such as sheep, goats, camels, cervids, horses, cattle, and swine) .Humans are incidental hosts; they do not play a role in the transmission cycle. E. granulosus adult tapeworms are usually found in dogs or other canids.

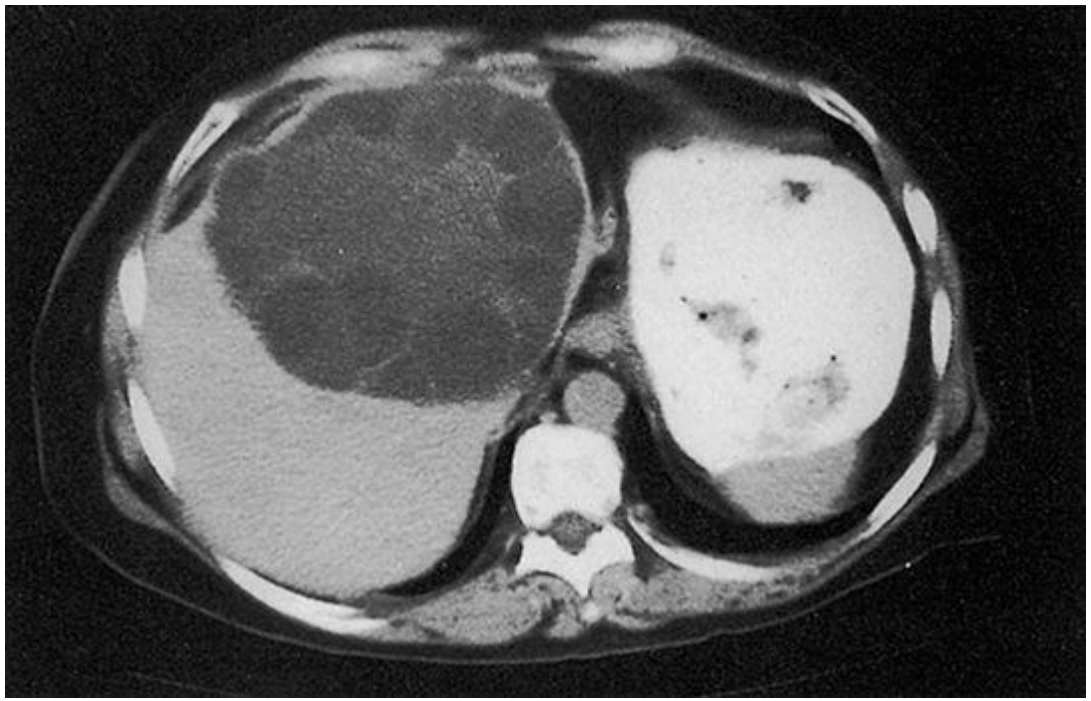

6
Scolex attaches to intestine
1
Adult in small intestine
5
Protoscolex from cyst
Ingestion of cysts (in organs)
Definitive host
(dogs & other canidae)
i 2
Embryonated egg in feces
Intermediate host
(sheep, goats, swine, etc)
Ingestion of eggs (in feces)
d 4
3
Hydatid cyst in liver, lungs, etc
Oncosphere hatches; penetrates intestinal wall
Δ
i Infective stage
d Diagnostic stage

- Epidemiology - The overall prevalence of echinococcal infection is underestimated in many series . The number of recognized cases is increasing, however, which may be due in part to better diagnostic technology and surveillance programs.
- In endemic rural areas, prevalence rates of 2 to 6 percent or higher have been recorded.
- Pathology — The hydatid cyst is usually filled with fluid .The inner layer is the germinative layer that gives rise to the hydatid fluid and small secondary cysts (brood capsules), which bud internally from this layer. Fragmentation of the germinative layer and brood capsules gives rise to daughter cysts.

Cysts may contain liters of fluid and thousands of protoscolices.

- CLINICAL MANIFESTATIONS
- Echinococcus granulosus — The initial phase of primary infection is always asymptomatic. Many infections are acquired in childhood but do not cause clinical manifestations until adulthood(more than 50 years). While approximately 50 percent of detected cases occur in asymptomatic patients, many more cases remain undiagnosed or are found incidentally at autopsy.
- The clinical presentation of E. granulosus infection depends upon the site of the cysts and their size. Small and/or calcified cysts may remain asymptomatic indefinitely. However, symptoms due to mass effect within organs, obstruction of blood or lymphatic flow, or complications such as rupture or secondary bacterial infections can result.

- Cysts typically increase in diameter at a rate of one to five centimeters per year.
- The liver is affected in approximately two-thirds of patients, the lungs in approximately 25 percent, and other organs including the brain, muscle, kidneys, bone, heart, and pancreas in a small proportion of patients.
- Single organ involvement occurs in 85 to 90 percent of patients with E. granulosus infection, and only one cyst is observed in more than 70 percent of cases.



A computed tomogram shows a multilocular cyst in the liver of a patient with hydatid disease.

- Liver involvement — frequently produces no symptoms. The right lobe is affected in 60 to 85 percent of cases. Significant symptoms are unusual before the cyst has reached at least 10 cm in diameter. If the cysts become large, hepatomegaly with or without associated right upper quadrant pain, nausea and vomiting can result .

- E. granulosus cysts can rupture into the biliary tree and produce biliary colic, obstructive jaundice, cholangitis, or pancreatitis.
- Pressure or mass effects can result in cholestasis, portal hypertension, venous obstruction, or the Budd-Chiari syndrome.
- can also rupture into the peritoneum, causing peritonitis, or transdiaphragmatically into the pleural space or bronchial tree, causing pulmonary hydatidosis or a bronchial fistula. Secondary bacterial infection of the cysts can result in liver abscesses.

- Lung involvement — The most common symptoms include cough (53 to 62 percent), chest pain (49 to 91 percent), dyspnea (10 to 70 percent), and hemoptysis (12 to 21 percent). Less-frequent symptoms include malaise, nausea and vomiting, and thoracic deformations . The majority of children and adolescents with lung lesions are asymptomatic .
- Cysts can break or develop secondary bacterial infection. complication is cyst rupture, into the bronchial tree or the pleural cavity. Bronchial tree involvement can lead to cough, chest pain, hemoptysis, or emesis; pleural cavity involvement can cause pneumothorax, pleural effusion, or empyema. Secondary bacterial infection of the cyst can manifest as a pulmonary abscess .
- Approximately 60 percent of pulmonary hydatid disease affects the right lung, and 50 to 60 percent of cases involve the lower lobes .Multiple cysts are common. Approximately 20 percent of patients with lung cysts also have liver cysts .The ratio of lung to liver involvement is higher in children than in adults.

- Other organs —
- Infection of the heart can result pericardial tamponade .
- Central nervous system involvement can lead to seizures or signs of raised intracranial pressure; infection of the spinal cord can result in spinal cord compression .
- Cysts in the kidney can cause hematuria or flank pain [14]. Immune complex-mediated disease, glomerulonephritis.
- Bone cysts are usually asymptomatic until a pathologic fracture develops.
- Ocular cysts also occur

- Cyst rupture — Fever and acute hypersensitivity reactions.

Hypersensitivity reactions are related to the release of antigenic material and secondary immunologic reactions.

- Outcome — Approximately 15 percent of patients had undergone surgery 10 to 12 years after the initial diagnosis. Among patients who did not undergo surgery, 75 percent remained asymptomatic; 57 percent did not have a change in the size of the cyst by imaging.
- Calcification usually requires 5 to 10 years to develop and occurs most commonly with hepatic cysts, but rarely with pulmonary or bone cysts. Total calcification of the cyst wall suggests that the cyst may be nonviable.

- DIAGNOSIS — Both cystic and alveolar echinococcus may be diagnosed with a combination of imaging and serology .
- Serologic assays for E. multilocularis infection are more sensitive and specific than for E. granulosus .
- E. granulosus — leukopenia or thrombocytopenia, mild eosinophilia, and nonspecific liver function abnormalities may be observed but are not diagnostic. Eosinophilia is observed in fewer than 15 percent of cases and generally occurs only if there is leakage of antigenic material.
- Imaging — Hydatid cysts may be visualized and evaluated with ultrasonography, computed tomography (CT), or magnetic resonance imaging (MRI).

- The sensitivity of ultrasonography for evaluation of echinococcus is 90 to 95 percent .
- The most common appearance on ultrasound is an anechoic, smooth, round cyst, which can be difficult to distinguish from a benign cyst. mixed echoes can be confused with an abscess or neoplasm. In the presence of daughter cysts, characteristic internal septation can be seen.
- Computed tomography — (CT) has higher overall sensitivity than ultrasonography (95 to 100 percent) .CT is the best mode for determining the number, size, and anatomic location of the cysts, and is better than ultrasound for detection of extrahepatic cysts.
- CT may also be used for monitoring lesions during therapy and to detect recurrences.
- may be superior to ultrasonography in assessing for complications such as infection and intrabiliary rupture

- Magnetic resonance imaging — has no major advantage over CT . may be better at diagnosing complications, particularly for cysts with infection or biliary communication. However, MRI is usually not required and in most instances, is not cost effective.
- Serologic and antigen assays — Serology is useful for primary diagnosis and for follow-up after treatment .Antibody detection is more sensitive than antigen detection for diagnosis of E. granulosus.
- Clinical factors — A negative serologic test generally does not rule out echinococcosis. There is no correlation between serologic results and the number or size of cysts .
- Overall, approximately 85 to 95 percent of liver cysts and 65 percent of lung cysts are associated with positive serology.
- Brain, eye, and splenic cysts often do not produce detectable antibodies, whereas bone cysts frequently are associated with positive serology.
- Serology is less likely to be positive with cysts at any site if the cysts are intact, calcified, or nonviable.
- Children and pregnant women more frequently have negative serology .

- False-positive reactions: other helminth infections (such as Taenia saginata, Taenia solium, and particularly neurocysticercosis), cancer, and immune disorders.

- Cyst aspiration or biopsy — In the absence of a positive serologic test, percutaneous aspiration or biopsy may be required to confirm the diagnosis by demonstrating the presence of protoscolices, hooklets, or hydatid membranes.
- Percutaneous aspiration of liver cyst contents is associated with very low rates of complications, but this method of diagnosis is generally reserved for situations when other diagnostic methods are inconclusive because of the potential for anaphylaxis and secondary spread of the infection
- Polymerase chain reaction — Polymerase chain reaction (PCR) techniques are limited to research settings.

- Differential diagnosis —such as cystadenoma, cystadenocarcinoma, hepatic abscess, a necrotic malignant tumor, hemangioma, and hamartoma.
- Treatment — The majority of simple cysts do not require treatment. However, monitor large cysts (≥4 cm in diameter) periodically with ultrasonography to assure that they remain stable. We suggest an initial follow-up study in three months after the diagnosis and then again at 6 to 12 months. Further monitoring is usually unnecessary if the cyst remains unchanged for two to three years.
- Percutaneous needle aspiration with injection of sclerosing agents is generally safe, effective, and relatively non-invasive .however, it may occasionally have serious complications.

CYSTIC ECHINOCOCCOSIS (E. GRANULOSUS)

- Management options for cystic echinococcosis (CE) include surgery, percutaneous management, drug therapy, and observation . Surgery has been the traditional approach for treatment of CE.
- cysts that are <5 cm may be treated with albendazole alone .In settings where albendazole treatment with follow-up monitoring is not feasible, definitive management with percutaneous treatment via PAIR (puncture, aspiration, injection, and reaspiration) is an acceptable alternative approach.
- cysts that are >5 cm may be treated with albendazole in combination with PAIR or surgery. In situations where albendazole treatment is not feasible, percutaneous treatment with PAIR is an alternative.

PAIR should not be performed in the following circumstances :

- Cyst with nondrainable solid material or echogenic foci
- Superficial cyst at risk of rupture into the abdominal cavity
- Cyst that has ruptured into the peritoneum
- Cyst with biliary communication
- Inactive or calcified cyst

- Multiseptated, "rosette-like" "honeycomb" cyst, Cyst with daughter cysts with any size = surgery
- Complicated cyst= surgery
- Other indications for surgery= cyst diameter >10 cm, superficial cyst at risk of rupture due to trauma, and extrahepatic disease (lung ,bone ,brain ,kidney, or other site) .
- Surgery is also appropriate in settings where percutaneous treatment is not available.
- Adjunctive drug therapy should be administered to minimize risk of secondary echinococcosis from seeding of protoscolices in the abdominal cavity in the event of fluid spillage. Albendazole is generally administered beginning one week prior to surgery and continued for at least four weeks postoperatively.

- Drug therapy — Drug therapy may be used for definitive management in selected cases; it is also a useful adjunctive therapy to surgery and percutaneous treatment. Albendazole is the primary antiparasitic agent for treatment of E. granulosus . Albendazole is poorly absorbed and should be ingested with food, ideally with a fatty meal to increase bioavailability (15 mg/kg/day, divided into two doses, to maximum 400 mg orally twice daily with food). In the absence of albendazole, mebendazole may be used as an alternative therapy; it is less well absorbed than albendazole.
- optimal duration is uncertain; one to three months may be appropriate, depending clinical factors; up to six months may be required.

- Drug treatment alone is usually not effective for treatment of cysts with diameter >5 cm, or for treatment of WHO stage CE2 or CE3b cysts .
- Other circumstances in which drug treatment alone may be warranted include management of multiple liver cysts <5 cm, management of cysts deep in liver parenchyma that are not amenable to percutaneous treatment.
- Mebendazole and praziquantel are less effective agents; in the absence of albendazole, mebendazole may be used as an alternative therapy.
- Mebendazole is dosed 40 to 50 mg/kg per day in three divided doses.
- Albendazole should be avoided during pregnancy because of potential teratogenicity; if feasible, treatment should be delayed until after delivery

- Follow-up — Cystic echinococcus (CE) can relapse years after treatment.
- The optimal approach to monitoring is uncertain and must be individualized according to patient characteristics and available resources .
- Follow-up usually consists of ultrasound or other imaging (CT or MRI) at three- to six-month intervals until the findings are stable, followed by yearly monitoring. Follow-up for up to 5 years is usually warranted to evaluate for recurrence; in some cases, 3 years may be sufficient if radiographic findings are stable .
- Serology — The optimal serologic test for monitoring patients on treatment for hydatid disease is uncertain. Frequently, serologic titers fall one to two years following successful surgery, and rise again in the setting of recurrence. However, antibodies may remain elevated even many years after successful cyst removal.

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