

Hydatid Disease of the liver

Life cycle

Hydatid disease is due to the larval or cyst stage of infection by the tapeworm *Echinococcus granulosus*, which lives in the dog. Humans, sheep, and cattle are intermediate hosts. The dog is infected by eating the viscera of sheep that contain hydatid cysts. Scolices, contained in the cysts, adhere to the small intestine of the dog and become adult taenia, which attach to the intestinal wall. Each worm sheds approximately 500 ova into the bowel. The infected ova-containing feces of the dog contaminate grass and farmland, and the ova are ingested by sheep, pigs, and humans. The ova have envelopes that are dissolved by gastric juice. The liberated ovum burrows through the intestinal mucosa and is carried by the portal vein to the liver, where it develops into an adult cyst. Most cysts are caught in the hepatic sinusoids, and 70% of hydatid cysts form in the liver. A few ova pass through the liver and are held up in the pulmonary capillary bed or enter the systemic circulation, forming cysts in the lung, spleen, brain, or bones. The most frequent site of hydatid cysts is the liver (50% to 70% of cases), followed by the lungs (20% to 30% of cases).

Three weeks after infection, a visible hydatid cyst develops and then slowly grows in a spherical manner. A pericyst, a fibrous capsule derived from host tissues, develops around the hydatid cyst. The cyst wall itself has two layers: an outer gelatinous membrane (ectocyst) and an inner germinal membrane (endocyst). Brood capsules are small intracystic cellular masses in which future worm heads develop into scoleces. In a definitive host, the scoleces would develop into an adult tapeworm, but in the intermediate host, they can only differentiate into a new hydatid cyst. Freed brood capsules and scoleces are found in the hydatid fluid and form the so-called hydatid sand. Daughter cysts are true replicas of the mother cyst. Hydatid cysts can die with degeneration of the membranes, development of cystic vacuoles, and calcification of the wall. Calcification of a hydatid cyst, however, does not always imply that the cyst is dead.

Clinical features

Because hydatid cysts grow slowly, a protracted asymptomatic stage is typical. Patients with symptomatic liver hydatid cysts present either with uncomplicated disease such as hepatomegaly, with right upper quadrant pain from cyst expansion, or with an acute complication.

Acute complications include:

- ✚ Obstructive jaundice or cholangitis from rupture into a bile duct,
- ✚ Abdominal pain or anaphylaxis from rupture of the cyst into the peritoneal cavity, or
- ✚ Productive cough from rupture into the plural cavity and the lung. However, most patients now present before symptoms occur, with the discovery of a cyst found on an abdominal imaging study.

- ✚ Bacterial superinfection of a hydatid cyst can occur and present like a pyogenic abscess.

DIAGNOSIS

- 1) Various laboratory tests have been used to establish the diagnosis, but none is definitive.
 - ✚ Traditionally, the Casoni and Weinberg skin tests have been employed to aid in the diagnosis. However, their sensitivity is so low that they are no longer used.
 - ✚ The enzyme-linked immunosorbent assay (ELISA) to identify specific antigens (echinococcal antigens and immune complexes) has up to a 90% sensitivity depending on the antigen preparation. This assay is clinically useful because the results can be obtained within hours as opposed to days with more sensitive tests such as Western blotting. The ELISA results may be negative in an infected patient if the cyst has not leaked or does not contain scolices, or if the parasite is no longer viable.
 - ✚ Complement fixation and indirect hemagglutination test results are positive in 85% to 90% of active cysts.
 - ✚ Eosinophilia is present in approximately 40% of patients but is not diagnostic.
- 2) Radiological investigations
 - ✚ Ultrasonography (US) and CT scans are the most widely used imaging studies to diagnose liver cysts. Both studies will show position, size, number of cysts, their proximity to vascular structures, and evidence of extrahepatic cysts. The classic findings for hydatid cysts are thick walls, often with calcifications, and many have daughter cysts. CT scanning may give better information about the location and depth of the cyst than ultrasound. MRI scans also demonstrate cyst characteristics and their relation to vascular and biliary structures. However, MRI does not give additional information and may not be cost effective
 - ✚ Endoscopic retrograde cholangiopancreatography (ERCP) when there is a complication such as cholangitis or jaundice. In these situations, ERCP can show communication between the cysts and bile ducts and can be used to drain the biliary tree before surgery .We advocate the routine use of ERCP to completely define the bile duct anatomy and to visualize any clinically silent connections between the bile ducts and cysts.
 - ✚ In some centers, intraoperative ultrasound also should be used when operative treatment for hydatid disease is undertaken. Operative ultrasound can show the relationship of bile ducts and vessels to the cyst with greater resolution than a transabdominal probe.

Treatment

Principles and Options

Although most echinococcal cysts are asymptomatic on presentation, their potential complications such as cholangitis, cyst rupture with anaphylaxis, or pulmonary infection require that all cysts be considered for treatment. However, because most cysts are asymptomatic, a careful well-thought-out plan of treatment should be devised, potentially using a number of modalities. Depending on the patient's state of health and the complicated nature of the cysts, medical, surgical, and percutaneous approaches to the disease should be considered. The principles of treatment are to eradicate the parasite within the cysts, to protect the host against spillage of scolices, and to manage complications.

Medical Therapy

Medical therapy has been limited to the benzimidazoles (mebendazole and albendazole). Albendazole is more readily absorbed from the intestine and is metabolized by the liver to its active form, whereas mebendazole is poorly absorbed and is inactivated by the liver. Therefore albendazole is the drug of choice for medical therapy. However, medical therapy alone for echinococcal cysts has a less than 30% success rate. The response has been shown to be higher in extrahepatic manifestations of the disease and with the alveolar form caused by *E. multilocularis*. Preoperative treatment with albendazole for at least 3 months has been shown to reduce the recurrence when cyst spillage, partial cyst removal, or biliary rupture has occurred. The length of therapy in these circumstances should be at least 1 month postoperatively. Medical treatment with albendazole relies on drug diffusion through the cyst membrane. The concentration of drug achieved in the cyst is uncertain but is better than that of mebendazole, and albendazole can be used as initial treatment for small, asymptomatic cysts. The dose of albendazole is 15mg/Kg/day in 3 divided doses. The duration of medical treatment (Albendazole and Praziquantel) is 2-3 months.

How to avoid recurrence postoperatively

Contamination of the peritoneal cavity at the time of surgery with active hydatid daughters should be avoided by continuing drug therapy with albendazole and adding perioperative praziquantel. This should be combined with packing of the peritoneal cavity with 20% saline-soaked packs and instilling hypertonic 20% saline into the cyst before it is opened.

The dose of praziquantel is 50mg/Kg/day in 3 divided doses.

Surgical Therapy

The treatment of hepatic hydatid cysts is primarily surgical. In general, most cysts are treated, but in elderly patients with small, asymptomatic, densely calcified cysts, conservative management is appropriate. The anesthesiologist has epinephrine and steroids available for the potential of an anaphylactic reaction intraoperatively.

Scolicidal Agents

Considerable controversy exists regarding the use of scolicidal agents. During the early experience with surgical management by cyst evacuation, a high rate of peritoneal implantation occurred. As a result, various scolicidal agents were tried to inject into the cyst before evacuation, to place into the adjacent peritoneal cavity, and to place into the cyst after evaluation.

Formalin was one of the first scolicidal agents, but formalin can cause sclerosing cholangitis when it enters the biliary tract. Therefore *formalin should never be used for this purpose.*

Hypertonic saline (20%) most widely used, but care should be taken to avoid biliary injection and overuse with resultant hypernatremia.

Alcohol 90%

Some authorities recommend **cetrimide or chlorhexidine**, but the safety of any agent entering the biliary tree has not been established. In addition, pre-evacuation injection should be avoided because intracyst pressure is already high. Thus many surgeons prefer a meticulous surgical technique rather than overreliance on scolicidal agents.

Surgical options

Open Cyst Evacuation

A number of operations have been used, but in general, the abdomen is completely explored, the liver mobilized, and the cyst exposed. Packing off of the abdomen is important because rupture can result in anaphylaxis and diffuse seeding. Usually, the cyst is then aspirated through a closed-suction system and flushed with a scolicidal agent such as hypertonic saline. The cyst is then unroofed, which can then be followed by a number of possibilities, including excision (or pericystectomy), marsupialization procedures, leaving the cyst open, drainage of the cyst, omentoplasty, or partial hepatectomy to encompass the cyst. Total pericystectomy or formal partial hepatectomy can also be performed without entering the cyst. Radical (resection) and conservative (drainage and evacuation) surgical approaches appear to be equally effective at controlling disease.

When bile duct communication is diagnosed at operation or preoperatively, it must be meticulously sought out. Simple suture repair is often sufficient, but

major biliary repairs, approaches through the common bile duct, or postoperative ERCP may be necessary. Recurrence rates after surgical treatment range from 1% to 20% but are generally 5% or less in experienced centers.

Pericystectomy

Pericystectomy involves complete resection of the cyst wall without entering the cyst cavity itself. This procedure can be done either along a plane outside the pericyst or along the cyst wall itself. Pericystectomy, like cyst evacuation, is best performed on cysts that are accessible along the periphery of the liver. Proper preoperative evaluation of the location of bile ducts and vascular structures with either CT or ultrasound should be obtained. If a bile duct connection is suspected, then preprocedure ERCP should be performed. The use of intraoperative ultrasound also is very helpful in identifying and avoiding key structures. The advantage of this procedure over simple cyst drainage is that it decreases the risk of content spillage into the peritoneal cavity, which can cause anaphylaxis, as well as the risk of recurrence. The disadvantage of pericystectomy is an increased risk of bleeding or damage to bile ducts in proximity to the cyst wall.

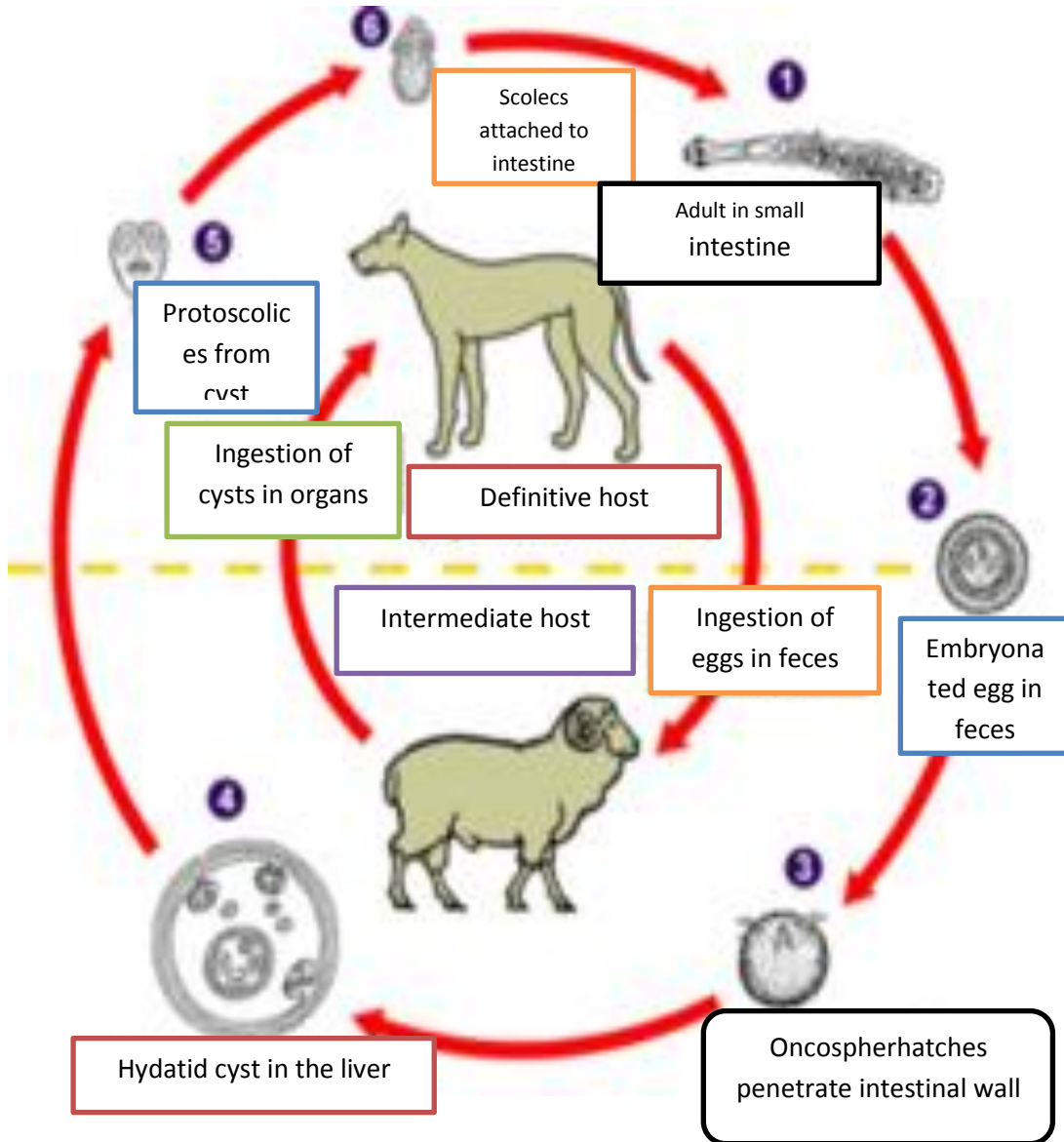
Laparoscopic Cyst Evacuation

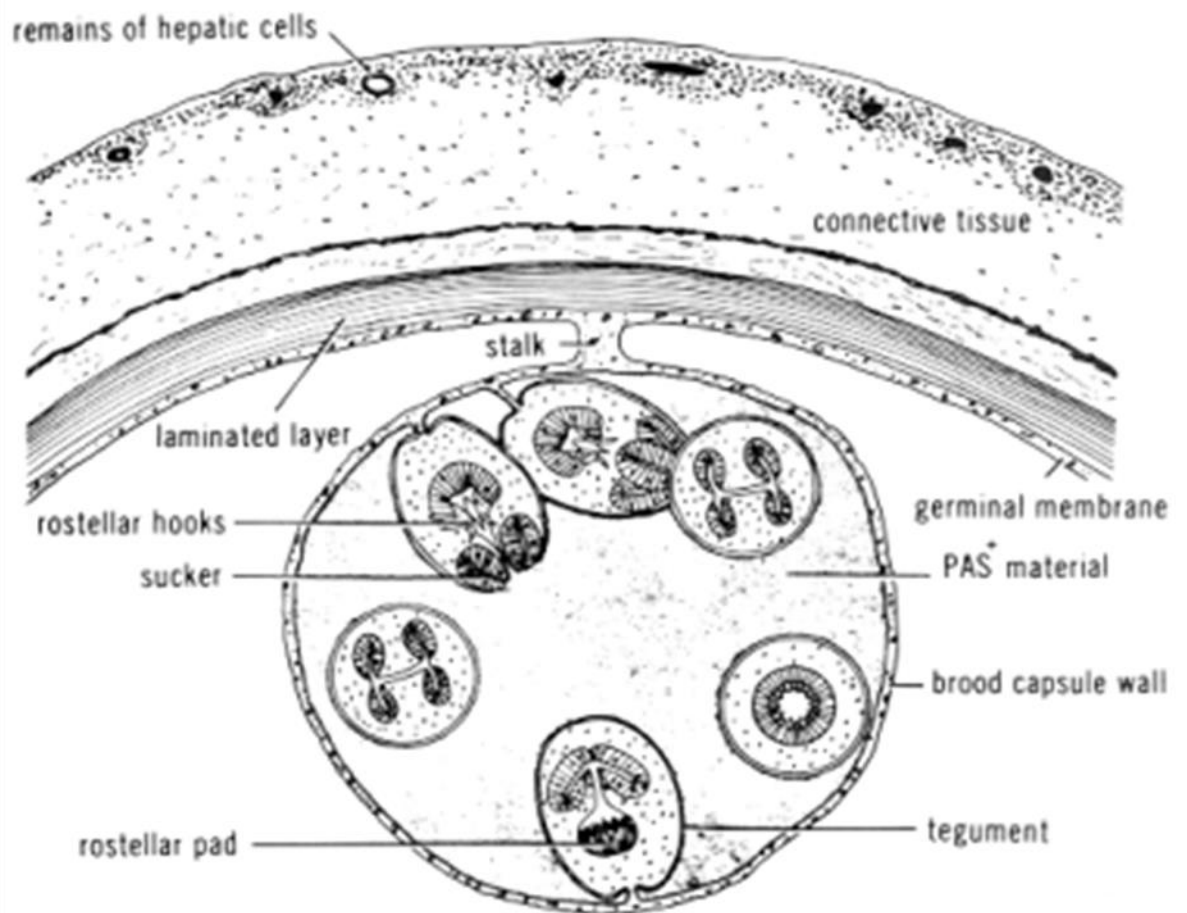
Several recent reports suggest that carefully selected patients with peripherally located echinococcal hepatic cysts may be safely managed by laparoscopic cyst evacuation. The laparoscopic approach is best suited for anterior cysts without thick calcified walls, but cysts in segments VI and VII may be managed by a right lateral approach. One technique uses an 11-mm trocar placed just above the cyst, through which 10% povidone iodine-soaked sponges are placed to act as ascoleoidal agent. The cyst is then punctured with a 14-gauge needle and aspirated. The endocyst will then shrink from the wall and rest at the bottom of the cyst. Next, the 11-mm trocar is exchanged for an 18-mm one, and the germinal membrane is then aspirated. The telescope is then inserted directly into the cyst to explore for remaining daughter cysts or biliary fistulae. The cyst cavity is then irrigated with 20% saline solution, and excision of the cyst wall is performed. Finally, omentoplasty or closed suction drainage may be performed as described above.

In properly selected patients with uncomplicated cysts, the conversion rate should be less than 5%. Operative times of less than 90 minutes have been reported, and complication rates have been low. A major disadvantage of the laparoscopic approach is the relative inability to avoid peritoneal spillage, especially with high intraabdominal pressures due to the pneumoperitoneum. Advocates of this approach recommend oral albendazole for 10 days preoperatively and for 3 months postoperatively. Obvious advantages for the laparoscopic approach, on the other hand, are reduced hospital stay, reduced hospital cost, and earlier return to productive activity.

Percutaneous Therapy

Albendazole was administered before the procedure for 10 days to those undergoing percutaneous drainage. In the percutaneously managed patients, the cyst was rapidly aspirated with a 5-Fr catheter. If daughter cysts were present, they were aspirated with a 20-cm long cholangiography needle. Then the cyst was filled with 20% saline for 20 minutes. The cyst was then aspirated completely, irrigated with 0.9% saline, and partially filled with 0.9% saline. The patients were then monitored and discharged from the hospital on average within 48 hours. In the percutaneously managed patients, the mean hospital stay was shorter than that for those who underwent the open procedure. No differences were observed between the two treatment groups in the percentage of patients with cyst disappearance, the percentage with subsequent negative antibody titers, or the residual cavity size 17 months after the procedure.





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