

Patients with hepatic echinococcosis who are symptomatic most often present with abdominal pain or a palpable mass in the right upper quadrant. Compression of a bile duct or leakage of cyst fluid into the biliary tree may mimic recurrent cholelithiasis, and biliary obstruction can result in jaundice. Rupture of or episodic leakage from a hydatid cyst may produce fever, pruritus, urticaria, eosinophilia, or anaphylaxis. Pulmonary hydatid cysts may rupture into the bronchial tree or pleural cavity and produce cough, salty phlegm, dyspnea, chest pain, or hemoptysis. Rupture of hydatid cysts, which can occur spontaneously or at surgery, may lead to multifocal dissemination of protoscolices, which can form additional cysts. Other presentations are due to the involvement of bone (invasion of the medullary cavity with slow bone erosion producing pathologic fractures), the CNS (space-occupying lesions), the heart (conduction defects, pericarditis), and the pelvis (pelvic mass).

The larval forms of *E. multilocularis* characteristically present as a slowly growing hepatic tumor, with progressive destruction of the liver and extension into vital structures. Patients commonly report upper-quadrant and epigastric pain. Liver enlargement and obstructive jaundice may be apparent. The lesions may infiltrate adjoining organs (e.g., diaphragm, kidneys, or lungs) or may metastasize to the spleen, lungs, or brain.

Diagnosis Radiographic and related imaging studies are important in detecting and evaluating echinococcal cysts. Plain x-rays will define pulmonary cysts of *E. granulosus*—usually as rounded masses of uniform density—but may miss cysts in other organs unless there is cyst wall calcification (as occurs in the liver). MRI, CT, and ultrasound reveal well-defined cysts with thick or thin walls. When older cysts contain a layer of hydatid sand that is rich in accumulated protoscolices, these imaging methods may detect this fluid layer of different density. However, the most pathognomonic finding, if demonstrable, is that of daughter cysts within the larger cyst. This finding, like eggshell or mural calcification on CT, is indicative of *E. granulosus* infection and helps to distinguish the cyst from carcinomas, bacterial or amebic liver abscesses, or hemangiomas. In contrast, ultrasound or CT of alveolar hydatid cysts reveals indistinct solid masses with central necrosis and plaque-like calcifications.

A specific diagnosis of *E. granulosus* infection can be made by the examination of aspirated fluids for protoscolices or hooklets, but diagnostic aspiration is not usually recommended because of the risk of

fluid leakage resulting in either dissemination of infection or anaphylactic reactions. Serodiagnostic assays can be useful, although a negative test does not exclude the diagnosis of echinococcosis. Cysts in the liver elicit positive antibody responses in ~90% of cases, whereas up to 50% of individuals with cysts in the lungs are seronegative. Detection of antibody to specific echinococcal antigens by immunoblotting has the highest degree of specificity.

TREATMENT ECHINOCOCCOSIS

Therapy for cystic echinococcosis is based on considerations of the size, location, and manifestations of cysts and the overall health of the patient. Surgery has traditionally been the principal definitive method of treatment. Currently, ultrasound staging is recommended for *E. granulosus* infections (Fig. 260-2). Small CL, CE1, and CE3 lesions may respond to chemotherapy with albendazole. For CE1 lesions and uncomplicated CE3 lesions, PAIR (percutaneous aspiration, infusion of scolicidal agents, and reaspiration) is now recommended instead of surgery. PAIR is contraindicated for superficially located cysts (because of the risk of rupture), for cysts with multiple thick internal septal divisions (honeycombing pattern), and for cysts communicating with the biliary tree. For prophylaxis of secondary peritoneal echinococcosis due to inadvertent spillage of fluid during PAIR, the administration of albendazole (15 mg/kg daily in two divided doses) should be initiated at least 2 days before the procedure and continued for at least 4 weeks afterward. Ultrasound- or CT-guided aspiration allows confirmation of the diagnosis by demonstration of protoscolices in the aspirate. After aspiration, contrast material should be injected to detect occult communications with the biliary tract. Alternatively, the fluid should be checked for bile staining visually and by dipstick. If no bile is found and no communication is visualized, the contrast material is reaspirated, with subsequent infusion of scolicidal agents (usually 95% ethanol; alternatively, hypertonic saline). This approach, when implemented by a skilled practitioner, yields rates of cure and relapse equivalent to those following surgery, with less perioperative morbidity and shorter hospitalization. In experienced hands, some CE2 lesions can be treated by aspiration with a trocar. Daughter cysts within the primary cyst may need to be punctured separately, and catheter drainage may be required.

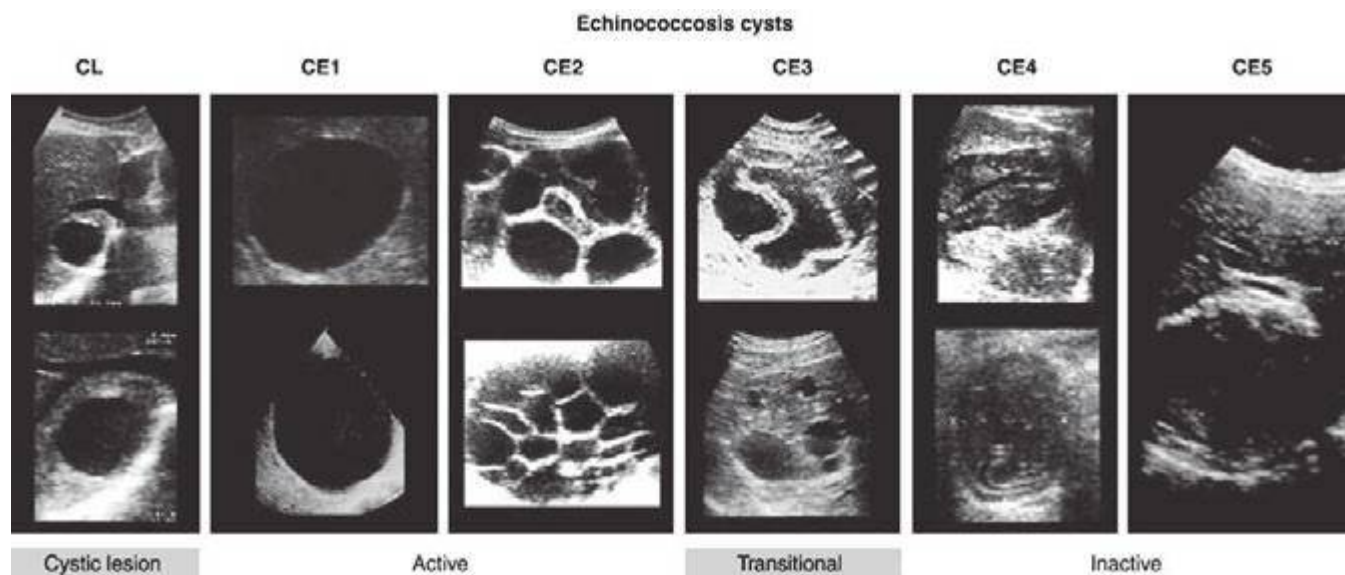


FIGURE 260-2 Management of cystic hydatid disease caused by *Echinococcus granulosus* should be based on viability of the parasite, which can be estimated from radiographic appearance. The ultrasound appearance includes lesions classified as active, transitional, and inactive. *Active* cysts include types CL (with a cystic lesion and no visible cyst wall), CE1 (with a visible cyst wall and internal echoes [snowflake sign]), and CE2 (with a visible cyst wall and internal septation). *Transitional* cysts (CE3) may have detached laminar membranes or may be partially collapsed. *Inactive* cysts include types CE4 (a nonhomogeneous mass) and CE5 (a cyst with a thick calcified wall). (Adapted from RL Guerrant et al [eds]: *Tropical Infectious Diseases: Principles, Pathogens and Practice*, 2nd ed, p 1312. © 2005, with permission from Elsevier Science.)