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Armando E. Gonzalez ESSENTIALS Cystic hydatid disease, caused by Echinococcus granulosus, is a zoo- notic disease principally transmitted between dogs and domestic livestock, particularly sheep. Humans are infected when they ingest tapeworm eggs, with disease occurring in most parts of the world where sheep are raised and dogs are used to herd livestock. Clinical features, diagnosis, and treatment—the most common clin- ical manifestations are cysts in the liver (typically presenting with hepatomegaly) and/or lung (presenting with cough, haemoptysis, and dyspnoea). Diagnosis is usually made on the basis of imaging techniques, supported by serological tests. Treatment options include
surgery, chemotherapy with anthelminthic agents, or—for liver cysts— PAIR (puncture–aspiration–injection–reaspiration). Prevention—echinococcosis is a major public health problem in several countries. Control programmes have been aimed at educating dog owners to prevent their animals from having access to infected offal, along with praziquantel treatment of dogs. Vaccines against sheep hydatidosis and the dog tapeworm stage are prom- ising alternatives. Introduction Cystic hydatid disease is a zoonotic disease caused by infection with the larval stage (hydatid cyst) of the tapeworm Echinococcus granulosus. Hydatid cysts in liver and lung are frequent causes of human morbidity in endemic zones. Aetiology The lifecycle of
E. granulosus requires two hosts. The adult tape- worm is found in the small intestine of the definitive host, usually dogs or other canids. It consists of only three to five proglottids, and measures between 3 and 7 mm long when fully mature. E. granu losus has remarkable biological

potential; there may be as many as 40 000 worms in a heavily infected dog, each one of which sheds about 1000 eggs every 2 weeks. Dogs infected with echinococcus tapeworms pass eggs in their faeces that contaminate the soil and vegetation and remain viable for long periods in cold humid places. Intermediate hosts (sheep, cattle, horses, pigs, and other mammals, including humans) acquire hydatid disease by ingesting viable eggs of E. granulosus. Eggs hatch in the intestine, freeing oncospheres which penetrate the intestinal mucosa and are transported by the blood and lymphatic systems to the liver, lungs, and other organs, where they develop into cysts. Molecular studies using mitochondrial DNA sequences have iden- tified 10 distinct genetic types within E. granulosus. These include two sheep strains (G1, G2), two bovid strains (G3, G5), a horse strain (G4), the camelid strain (G6), a pig strain (G7) and the cervid strain (G8). A ninth genotype (G9) has been described in swine in Poland, and a tenth genotype (G10) in cervids. The sheep strain (G1) is the most cosmopolitan form that is most commonly associated with human infections. The other strains appear to be genetically distinct. The presence of distinct strains of E. granulosus may affect clinical aspects and control strategy. The risk of human infection differs as does its localization in the body, clinical expression, and geograph- ical distribution. Shorter maturation time of a given strain in dogs might reduce the duration of infection by the adult intestinal form so that shorter intervals might be required between rounds of ad- ministration of antiparasite drugs for control. Epidemiology Hydatid disease is an important cause of human morbidity, requiring costly surgical treatment. The infection is widely distributed in most parts of the world where sheep are raised and dogs are used to herd livestock. In the Americas, most cases have been reported from Argentina, Chile, Uruguay, Peru, and southern Brazil. Studies in Peru have revealed a prevalence of hydatid disease ranging from 5.7 to 8.9% in highland villagers, and as high as 32 and 89% in dogs and sheep, respectively. High prevalences of liver hydatid disease, with rates of up to 5.6%, have also been reported in north-western Turkana in Kenya. Echinococcus is widespread in the Old World, particularly in Greece, Cyprus, Bulgaria, Lebanon, and Turkey. In the United States of America, most infections are seen in immigrants from endemic countries; however, sporadic autochthonous transmission is currently recognized in Alaska, California, Utah, Arizona, and New Mexico. Communities at higher risk of infection include those where sheep are raised extensively and where dogs are used to care for large flocks of livestock. Known risk factors for infection include feeding dogs with raw offal and access of dogs to sheep that die in the field (Fig. 8.10.2.1). The risk of infection is also linked to poor hygiene and intimate contact with dogs. In north-western Turkana, dogs are allowed to stay within the house, and are used to clean up women’s menses and lick vomit from faces and diarrhoea from the Fig. 8.10.2.1 Epidemiological conditions for completion of the life cycle of echinoccocus: stray dogs waiting for sheep offal outside a slaughterhouse in Peru.

section 8 Infectious diseases 1530 anal regions of their children. Global losses due to hydatid disease have been estimated to be US $763 980 979. Pathogenesis The incubation period of human hydatid infections is highly vari- able and often prolonged for several years. Cysts have been reported to grow continuously. However, recent studies suggest that cyst growth is highly variable. Some cysts grow as much as 1 cm per year while other viable cysts showed no growth during 3 to 12 years of follow-up. Most human infections remain asymptomatic; hydatid cysts are found incidentally at autopsy much more frequently than the re- ported local morbidity rates. The locality of the cysts, their size, and their condition determine the particular manifestations. Clinical features Hydatid cysts are most frequently seen in the liver (60–70%) fol- lowed by the lungs (30–40%). Signs of hepatic hydatid disease in- clude hepatomegaly with or without the presence of

a mass in the upper right quadrant. Obstructive jaundice, mild epigastric pain, indigestion, and nausea might occur occasionally. Hydatid cysts can become secondarily infected with bacteria presenting as a hep- atic abscess. Features of lung involvement (Fig. 8.10.2.2) are cough, haemoptysis, dyspnoea, and fever. The ratio of liver to lung cysts varies from one geographical region to another: a liver to lung ratio of 1.4:1 has been observed in Peru, in contrast to the 3:1 to 13:1 ratio reported in Argentina and Uruguay. Brain cysts produce intracra- nial hypertension and epilepsy. Vertebral cysts compress the spinal cord causing paraplegia; bone cysts produce spontaneous fractures (Figs. 8.10.2.3 and 8.10.2.4) and deformity. Sudden rupture of cysts in the peritoneal cavity might result in peritonitis (Fig. 8.10.2.4), and rupture in the lungs can cause pneumothorax and empyema. Rupture can also cause allergic manifestations such as pruritus, oe- dema, dyspnoea, anaphylactic shock, and even death. Diagnosis Clinical findings, such as a space-occupying lesion, and residence in an endemic region are suggestive of hydatid disease. Abdominal ultrasonography is the most widely used imaging technique for echinococcosis because of its widespread availability and useful- ness for defining number, location, dimensions, and vitality of cysts. Ultrasonography is the basis for the World Health Organization Informal Working Group on Echinococcosis (WHO-IWGE) clas- sification of hydatid cysts and is the basis for treatment decisions involving hepatic cysts. Portable ultrasonography machines are used with good results in field surveys. Chest radiography is useful for diagnosis of lung cysts. CT scanning is very helpful, especially for diagnosis of atypical lesions (Fig. 8.10.2.4b). Serology Several serological tests have been developed for diagnosis of hydatid disease, including an enzyme immunoassay, which iden- tifies antibodies against antigen B or components of this antigen. Fig. 8.10.2.2 Plain chest radiograph showing a lung hydatid cyst displacing the heart. (a) (b) Fig. 8.10.2.3 (a) Pathological fracture of the femur caused by hydatid infection. (b) Hydatid cyst in muscle excised from around the femoral head (same case as shown in (a)). Copyright D. A. Warrell.

 A western blot assay based on the identification of three specific antigens of 8, 16, and 21 kDa is currently used. Major drawbacks in serological diagnosis are low sensitivity for detection of lung hydatid cysts and cross-reactivity with sera of patients with Taenia solium infection. Cyst rupture or secondary infection are strongly associated with a positive result in hydatid serology. In field sur- veys, serological tests should be used in combination with imaging techniques in order to detect most cases of hydatid disease. Parasitological diagnosis When serological assays are negative, identification of protoscolices, hooklets, or hydatid membranes can be done through percutaneous aspiration of liver cyst contents. It might also be possible to identify these structures from sputum samples of patients whose lung cysts have recently ruptured. Treatment Treatment of hydatid cysts should be stage-specific and should take into account cyst size, stage, number, localization, presence of complications, as well as characteristics of the patient (e.g. com- pliance with long-term follow-up). The WHO-IWGE reached a consensus on treatment of cystic echinococcosis which is an image-based, stage-specific approach and follows these options:

1. surgery, 2) chemotherapy, 3) percutaneous treatment, 4) watch and wait. An imaging-based classification system is recommended when using this approach (Box 8.10.2.1). The benefits and limi- tations of current treatment options have been reviewed by the WHO-IWGE and others. Determining the radiologic stage of the cyst, using the WHO classification of the cyst stage, and the size of the cyst are the first steps in determining the treatment options. Surgery Surgical removal of hydatid cysts remains the treatment of

choice in many countries. Surgery is the preferred treatment for (1) large cysts with multiple daughter vesicles (e.g. CE2, CE3b cysts of the WHO- IWGE classification); (2) single superficial hepatic cysts which may rupture spontaneously or through trauma; (3) infected cysts or cysts located in certain organs (i.e. lung, brain, kidney) or (4) cysts that communicate with the biliary tree. Surgery is contraindicated in pregnant women, patients with pre-existing medical conditions that put them at operative risk, or those who have multiple cysts that are difficult to access. The usual surgical approach involves aspiration of cyst fluid and injection of a protoscolicidal agent into the cyst, usually 20% hypertonic saline solution or 90% alcohol, followed by evacuation of the fluid, prior to surgical excision. Major risks of sur- gical treatment include accidental spillage of fluid and scolices into the peritoneal cavity, which may lead to anaphylaxis or secondary peritoneal hydatidosis. Recurrence rates following surgery can be as (b) (a) Fig. 8.10.2.4 (a) Numerous subcutaneous, peritoneal, and renal hydatid cysts in an Argentine patient. (b) Contrast CT scan of the same patient. Courtesy of Professor Olindo Adriano Martino, Buenos Aires. Box 8.10.2.1 The World Health Organization 2001 classification of hepatic hydatid cysts is a useful for assessing stage of a hepatic hydatid cyst on ultrasound and to decide on appropriate management for it depending on the stage of cyst. Classification • CL — unilocular anechoic cystic lesion without any internal echoes and septations • CE 1 — uniformly anechoic cyst with fine echoes settled in it representing hydatid sand • CE 2 — cyst with multiple septations giving it multivesicular appearance or rossette appearance or honey comb appearance with unilocular mother cyst — this stage is the active stage of the cyst • CE 3 — unilocular cyst with daughter cysts with detached laminated mem- branes appearing as ‘water lily sign’ — this is the transitional stage of the cyst • CE 4 — mixed hypo and hyperechoic contents with absent daughter cysts, these contents give an appearance of ball of wool sign indicating the degenerative nature of the cyst • CE 5 — arch-like thick partially or completely calcified wall — this stage of cyst is inactive and infertile

section 8 Infectious diseases 1532 high as 30%. Intraoperative mortality can vary from 0.5% to 4% and can be even higher with repeated interventions. Antihistamines are given as prophylaxis and suction cones have been used to prevent spillage. The efficacy of these methods is uncertain. Some experts recommend prophylactic chemotherapy prior to surgery Chemotherapy Benzimidazole compounds have been shown to be effective against some forms of hydatid disease. They can be used alone in patients with CE1 and CE3a cysts that are less than 5 cm in diameter. They are used in combination with percutaneous interventions CE2 and CE3b as well as larger CE1 and CE3a cysts. It is not indicated for CE4 or CE5 cysts. They can also be used in patients who are not candidates for, or refuse, surgery and those with multiple cysts. Albendazole should be administered in a dose of 10–15 mg/kg per body weight per day without interruption. This regime cures approximately one-third of cases of liver hydatid disease and causes partial regression of cysts in another one-third of patients. Because of its high scolicidal activity, albendazole is recommended as a prophylactic agent 1 to 3 months before surgical intervention. Albendazole is indicated when surgery is contraindicated. Mebendazole can also be used, although it is less effective than albendazole. Albendazole, mebendazole, and other benzimidazole compounds should not be used during early preg- nancy because of their potentially teratogenic effects at the doses and regimes used for hydatid disease. Since benzimidazoles are potentially hepatotoxic, liver enzymes should be monitored before and during treatment, every 2 weeks during the first 3 months and then monthly for one year. The optimal duration of treatment with albendazole is

uncertain; one to three months might be appropriate, depending on clinical factors; up to six months can sometimes be re- quired. Fat fat-rich meals facilitate absorption and bioavailability of albendazole. Adverse reactions (neutropenia, liver toxicity, alopecia, and others), have been observed in a few patients but are reversible with cessation of treatment. In addition to early pregnancy, contra- indications to chemotherapy include chronic hepatic diseases, and bone marrow depression. The combination of praziquantel and albendazole seems to show better efficacy than albendazole alone in a few human case series and experimental animal studies but further studies are needed before this approach can be recommended. Recent experimental studies in animals have shown that another benzimidazole compound, oxfendazole, has strong parasiticidal activity. Intermittent weekly therapy with oxfendazole was ef- fective in sheep hydatid disease, suggesting the possibility that daily therapy as currently used with albendazole might not be needed. Current studies are evaluating the safety and pharmacokinetics of oxfendazole in the treatment of humans. Percutaneous treatment There are two categories of percutaneous treatment. There is Percutaneous aspiration, injection, reaspiration (PAIR) and there is non-PAIR percutaneous treatment. PAIR consists of percutaneous puncture using sonographic guid- ance, aspiration of substantial amounts of the cyst fluid, and injec- tion of a protoscolicidal agent, usually hypertonic saline for at least 15 min, followed by reaspiration of cyst contents. PAIR is indicated for patients with CE1 and C3a cysts that are larger than 5 cm in diameter. It can also be considered for those who cannot undergo surgery and for patients who refuse surgery, or those who have single or multiple cysts in the liver, abdominal cavity, spleen, kidney, and bones. PAIR is contraindicated for inaccessible or superficially located liver cysts and for inactive or calcified cystic lesions. It is also contraindicated for cysts that communicate with the biliary tree, CE2, CE3b, CE4, CE5, and lung cysts. Complications of PAIR in- clude secondary infection of the cavity, acute allergic reactions, and recurrence; however, these have been rare. Albendazole should be administered before PAIR treatment and one month after treatment. During PAIR antihistamines should be given to reduce the risk of allergic reactions if there is spillage of fluid. Good results have been reported with this procedure with no major complications. A meta- analysis comparing the use of PAIR and surgical treatment for liver hydatid cysts found fewer complications and a shorter hospital stay in the PAIR-treated group. CE2 and CE3b cysts that are not surgically resected require non- PAIR percutaneous treatment in combination with chemotherapy. These two types of cyst respond poorly to PAIR. The goal of the non- PAIR percutaneous treatment is the removal of the entire endocyst and all daughter cysts. This requires a large bore catheter or a cutting device with an aspiration apparatus. The long-term outcomes are unclear but studies of medium term outcomes suggest that treat- ment is successful. As with PAIR, this treatment is usually accom- panied by chemotherapy. Watch and wait Uncomplicated asymptomatic inactive cysts of the liver can be left untreated and monitored regularly using imaging techniques. The rationale for this approach is based on the observation that up to 20% of cysts become spontaneously inactive and remain so over time. CE4 and CE5 stage cysts are managed with this approach. Prevention and control Control programmes have been aimed at educating dog owners to prevent their animals from having access to infected offal. This ap- proach includes periodic treatment (every 45 days) of sheepdogs with 5 mg/k of praziquantel, reduction in the dog population, close veterinary inspection of slaughterhouse facilities for the presence of dogs, and cremation of infected offal. Control programmes are in force in Argentina, Chile, and Uruguay. Partial success has been achieved. Control programmes in Cyprus, the Falkland Islands, New Zealand, and Tasmania have reduced the number of infected animals and the incidence of human infection. Serological tests such as the western blot for diagnosis of sheep hydatidosis and the coproantigen enzyme-linked immunosorbent assay (ELISA) for canine echinococcosis are potentially useful for measuring the

burden of disease and monitoring control pro- grammes in endemic regions. A recent major advance has been the development of a recombinant vaccine (EG95) which seems to confer 96–98% protection against challenge infection with oncospheres. Recent trials in Australia and Argentina using this vac- cine have reported that 86% of immunized sheep were completely free of viable hydatid cysts when examined 1 year later. The number of viable cysts was reduced by 99.3%. Similarly, a vaccine against the dog tapeworm stage has been developed and conferred 97 to 100% protection against worm growth and egg production in immunized dogs. A recent field trial of the EG95 vaccine in Argentina found that

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