

Leading Article

Management of hydatid disease of the liver

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Hydatid disease of the liver due to *Echinococcus granulosus* remains an important and challenging medical problem. It has a worldwide distribution and is endemic in sheep-rearing regions like the Mediterranean and Baltic, South America, Australia, New Zealand, Middle East and the Far East. Extensive population migration has led to an increasing incidence of the disease in non-endemic areas like England¹ and North America.² The liver hydatid cyst remains asymptomatic for years and then presents either due to local complications like pressure, rupture or infection or due to systemic hypersensitivity reactions.³ Rupture is usually into the biliary tract, peritoneal cavity or thoracic cavity and rarely into the vascular system, alimentary tract or urinary tract.⁴

A high index of suspicion is essential to initiate a useful array of modern investigating facilities. Immunological testing has come a long way from the unpredictable Casoni's test and has a role not only in primary diagnosis, but also in screening of susceptible populations and post-surgical follow-up for recurrences.⁵ At present, for primary diagnosis, hydatid immunoelectrophoresis (HEP) and enzyme-linked immunosorbent assay (ELISA) are the tests of choice. For post-surgical follow-up for recurrent disease, HEP, and for large-scale screening of the susceptible population, ELISA are preferred. A plain X-ray of the abdomen may reveal curvilinear or ring-like calcification (pericyst) or several rings of calcification (daughter cysts) in up to 20–30% of the patients.⁶ Radionuclide scan reveals a space-occupying lesion, a non-specific finding, in most of the cases.⁷ Ultrasound is a widely used and highly accurate test.⁸ Findings vary from a univesicular cyst without echogenic areas (Gharbi type I—difficult to differentiate from simple liver cysts) to a multiseptate cystic lesion with irregular echogenic areas within it or a complex heterogenous mass or a calcified mass (Type III, IV, V). Cases with cyst rupture into the biliary tree reveal echogenic material without

acoustic shadowing in the bile duct,⁹ and rarely, cyst-biliary communication (pathognomonic of rupture^{10,11}) may also be demonstrated. Computed tomographic (CT) scan has much the same findings, with enhancement of cyst septae on CT after intravenous injection of a cholangiographic contrast material being indicative of biliary communication.¹² Recently, the membrane detachment sign,¹³ seen on CT scan after percutaneous aspiration of the cyst fluid, has been described as typical of hydatid cyst and is helpful in differentiating Gharbi type I hydatid cysts of the liver from simple liver cysts. The investigation of choice for confirmation and direct visualization of cyst biliary communication is endoscopic retrograde cholangiopancreatography (ERCP),^{14–16} which may reveal filiform linear filling defects (laminated membranes), rounded lucent filling defects (daughter cysts) or amorphous irregular material (both combined) in the bile duct,¹⁷ apart from retrograde filling of the cyst itself by contrast from the bile duct. Duodenoscopy in these cases may reveal membranes protruding through the papilla.¹⁸ Percutaneous transhepatic cholangiography also provides the same information¹⁹ but is avoided by most people for fear of anaphylaxis and dissemination of the disease,⁶ though recent reports^{2,13,20–22} prove this fear to be unfounded. In cases where cyst rupture is suspected intra-operatively, a per-operative cholangiogram provides the requisite information.⁵ Hepatic angiography is required only when a major hepatic resection is contemplated. Magnetic resonance imaging,^{23,24} which reveals curvilinear and ring-like low intensity structures within the cyst, is still under evaluation. A judicious combination of these investigative tools today allows us to make an accurate preoperative diagnosis of hydatid cyst as well as presence of complications, if any. This paves the way for a 'planned surgery' in these cases, thereby reducing the number of intra-operative surprises and post-operative complications.

Percutaneous aspiration of liver hydatid cysts for diagnostic and therapeutic purposes is an interesting new development and is being increasingly performed. Fine needle aspiration, performed

transhepatically¹³ (not transperitoneally) has been reported to be safe. Microscopic examination of the fluid reveals basophilic finely granular background material with hooklets or scolices, differentiating it from simple liver cysts.^{2,20} Successful attempts have recently been made to achieve cyst sterilization (definitive treatment) by percutaneous injection of different scolicial agents (hypertonic saline^{2,13,20} or 95% ethanol²¹) after aspiration of the cyst. Side effects were minimal and no early recurrences were observed.² Though the long-term results of the therapy are awaited, it has the potential to emerge as an alternative treatment for small, unilocular (Gharbi type I, II), asymptomatic, uncomplicated cysts or for those unfit for or refusing surgery.^{21,22}

Surgery remains the mainstay of treatment of hydatid disease of the liver, though controversies still exist about the preferred operative technique. The procedures available can be divided into 'conservative' and 'radical' groups.

The 'conservative procedures' achieve neutralization of the parasite and evacuation of cyst contents, leaving the pericyst intact. The essential steps are: packing of the peritoneal cavity with sponges soaked in scolicial solution,²⁵ partial decompression of the cyst before opening it using a wide bore needle or a catheter needle device²⁶ reported recently (cyst fluid is often under high pressure) and careful evacuation of all cyst contents without spillage. Use of Aaron's cone⁵ and multiple wide-bore suction tubes help prevent spillage. The inside of the cyst cavity is now thoroughly swabbed with sponges soaked in scolicial agent. Intracystic injection of scolicial agents prior to cyst evacuation is not always useful and has been termed by Saidi²⁷ as an 'ineffective ritual which is neither standardized nor reliable'. *After cyst evacuation, it is essential to carefully look for and under-run any biliary communications.* The residual cyst cavity can be filled with normal saline and closed primarily without a drain (capsulorrhaphy),⁵ or an omentoplasty^{3,28-30} may be performed. A small cavity can be obliterated by capittonage³¹ also. External drainage and marsupialization are to be condemned. Internal drainage is not recommended as a primary procedure.

The 'radical procedures' achieve complete removal of both the cyst and the pericyst. These consist mainly of total cystopericystectomy. Cases with pericyst adherent to vital structures like hepatic veins or intrahepatic vena cava should have subtotal cystopericystectomy, leaving behind only the adherent part of the pericyst.³² Cystopericystectomy is done with or without prior controlled evacuation of the cyst ('open' or 'closed' cystopericystectomy³³). Also included in this group are wedge liver resections (cases with superficial,

peripheral cyst) and major liver resections (single large or multiple cysts occupying an entire lobe).

Postoperative bile leak and recurrence rates are the parameters used to compare the two groups. The conservative procedures, easier to perform, have a reported bile leak rate of 14%³²-27.5%³⁴, longer hospital stay and recurrence rates up to 16%³². The radical procedures, on the other hand, have a lower bile leak rate (3.5%³⁴-6.5%³²), shorter hospital stay and lower postoperative recurrence rates (2%)³². Many surgeons (authors included), however, consider them inappropriate, claiming that the intra-operative risks may be too high for a benign disease.^{29,35} Unlike conservative procedures, these may be hazardous in inexperienced hands, although several measures like temporary hepatic pedicle occlusion,³⁶ the use of a Lin liver clamp,³⁷ intra-operative ultrasound and the ultrasonic surgical aspirator³³ have recently been suggested to decrease the risks involved. Bile leaks after conservative procedures may be greatly reduced (as low as 1.5%³⁰) by meticulously looking for and under-running the cyst biliary connections at surgery, supplemented by an omentoplasty.³⁰ Local recurrence is now being increasingly recognized to be due to the presence of exogenous cysts (outside the pericyst)³⁸ which supposedly remain undetected and untreated during conservative procedures.³² However, these cysts, being reported in up to 23% of the patients undergoing total cystopericystectomy,^{38,39} can be picked up during conservative surgery also by the use of an intra-operative ultrasound.³² These observations negate the stated advantages of the radical procedures. Thus, without recommending any one of these as the 'standard' procedure for hydatid disease of the liver, we endorse the concept that the choice of the operative procedure is best left to the operating surgeon, who should take into consideration the site, size and number of the cyst(s), presence or absence of complications, condition of the patient and the surgical expertise available. No single procedure will suit all hydatid cysts.

Complicated liver hydatid cysts deserve separate mention here. Infected cysts should have external drainage³³ with or without omentoplasty,⁵ after evacuation of its contents. Cysts with biliary rupture should have cholecystectomy and exploration of the common bile duct with meticulous clearance of all migrated hydatid material from it, preferably aided by an intra-operative choledochoscopy.⁴⁰ Instillation of scolicial substances directly into the duct should be avoided.^{41,42} After exploration, the duct can be closed over a T-tube^{43,44} or a choledcho-duodenostomy⁴⁵ or transduodenal sphincteroplasty⁴⁶ can be performed. Alternatively, duct clearance can be achieved by pre-operative endoscopic papillotomy,⁴⁷⁻⁴⁹ obviating the need for duct exploration with its attendant complications. Some

authors have reported endoscopic papillotomy, extraction of hydatid contents from the duct and the cyst, and endoscopic irrigation of the main cyst by scolical substances as the definitive treatment for complicated hydatid cysts of liver—avoiding surgery altogether.⁵⁰ Cysts with intrathoracic rupture, which may be into a bronchus or pleura or pericardium, are approached by thoraco-abdominal route and should have removal of all hydatid contents, lower lobectomy (if broncho-biliary fistula is there), lavage of pleural cavity by a safe scolical solution and closure of diaphragm with drainage of the pleural cavity.^{5,32} Cysts with intraperitoneal rupture usually present with peritonitis with or without anaphylactic shock and are managed along similar lines.

The contribution of chemotherapy to the management of hydatid disease of the liver is growing. Mebendazole and flubendazole, used initially, have been discarded due to poor absorption and low serum and cyst fluid concentration. Albendazole,⁵¹ another benzimidazole derivative and praziquin-

tal,⁵² an isoquinoline derivative are currently most widely used. They achieve high serum and cyst fluid concentrations and are rapidly scolical. On the germinal membrane, however, albendazole is more effective.⁵ A recent WHO trial⁵³ has recommended their use in inoperable cases (primary chemotherapy) and to prevent postoperative recurrences (adjuvant chemotherapy). For primary chemotherapy, albendazole 10 mg/kg/day is the drug of choice. In adjuvant setting either of these can be used. They may be given pre-operatively, to achieve 'sterilization' of the cysts (albendazole 10 mg/kg/day for 4 weeks or praziquin 50 mg/kg/day for 2 weeks) or postoperatively, in cases with cyst rupture before surgery or inadvertent spillage during surgery. Recently, both the drugs used together have been shown to be more effective in *in vitro*⁵⁴ and animal *in vivo* studies.⁵⁵ Their combined use in human subjects is under evaluation. Primary chemotherapy as an alternative to surgery in operable cases is still controversial as no hard data are presently available.

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